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MISSION STATEMENT
The North American Neuro-Ophthalmology Society (NANOS) is dedicated to the achievement of excellence in patient care through the support and promotion of education, communication, research, and the practice of neuro-ophthalmology.

TARGET AUDIENCE
Neurologists, Ophthalmologists, Neuro-Ophthalmologists, physicians and other health care professionals who have a special interest in neuro-ophthalmology, or have fellowship training in neuro-ophthalmology and are members of the North American Neuro-Ophthalmology Society.

POLICY ON COMMERCIAL SUPPORT AND FACULTY DISCLOSURE
The North American Neuro-Ophthalmology Society maintains a policy on the use of commercial support which ensures that all educational activities sponsored by NANOS provide in-depth presentations that are fair, independent, and scientifically rigorous. To this end, all speakers and planners are required to complete a “Disclosure Form”. This information is included in this syllabus (see pages 8 - 11) and/or may be supplemented by announcements by moderators.

DISCLOSURE OF UNLABELED/UNAPPROVED USES
This educational program may include references to the use of products for indications not approved by the FDA. Areas of unapproved uses will be disclosed at each presentation. Opinions expressed with regard to unapproved uses of products are solely those of the faculty and are not endorsed by the North American Neuro-Ophthalmology Society or any other manufacturers of pharmaceuticals.

ACCREDITATION
The North American Neuro-Ophthalmology Society is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

CREDIT DESIGNATION
NANOS designates this live activity for a maximum of 33.5 AMA PRA Category 1 Credit(s)™. Physicians should claim only the credit commensurate with the extent of their participation in the activity.
NANOS CME MISSION STATEMENT

The North American Neuro-Ophthalmology Society (NANOS) is dedicated to the achievement of excellence in patient care through the support and promotion of education, communication, research, and the practice of neuro-ophthalmology.

To this end, the Society sponsors an annual scientific meeting which is its main CME (continuing medical education) activity. Recent advances in the structure and function of the nervous and visual systems as they pertain to neuro-ophthalmology, its pathology, new therapies, and new diagnostic and therapeutic technology comprise the major focus of the annual scientific meeting and its CME content. In addition, the Society supports joint sponsorship with other respected and recognized medical organizations that comply with the ACCME Essential Areas and their Elements.

Members of the Society, physicians, fellows, and resident physicians comprise the meeting participants and target audience for our CME program. They are predominantly neuro-ophthalmologists, neurologists, and ophthalmologists but also include physicians and scientists from other disciplines and specialties.

The annual scientific meeting of NANOS includes the following types of activities: symposium topics of current interest, special lectures, reports of original research presented at both platform and poster sessions, and reports of special committees assigned to evaluate and report to the membership on specific problems and controversial issues.

The NANOS annual scientific meeting features advances and best practices in neuro-ophthalmology, so that as a result attendees can incorporate them into their medical practices. The scope of the meeting includes: enhancing our diagnostic skills by discussion of challenging cases, promoting evidence-based treatments and avoiding ineffective treatments, and using new diagnostic tools and techniques. Participants should learn the results of ongoing basic and clinical research in neuro-ophthalmology. NANOS uses member surveys, program evaluations (both during and after the meeting), meetings of its Scientific Program Committee, Education Committee, and Board of Directors, and other appropriate means to assess the effectiveness whether the program achieved these outcomes and discuss the knowledge gaps that exist in the field of neuro-ophthalmology that should be addressed in future educational activities.

The NANOS Board of Directors, Scientific Program Committee, Education Committee, and CME Committee review and assess the educational gaps and content and participant critiques of the annual NANOS CME program and general membership comments and suggestions to ensure that all educational objectives are achieved.

Adopted by the NANOS CME Subcommittee October 11, 2005

Adopted by the NANOS Board of Directors October 15, 2005

Reviewed and Approved by the NANOS Board of Directors February 10, 2007

Updated by the NANOS CME Subcommittee and NANOS Board of Directors January 9, 2009
FAIRMONT HOTEL VANCOUVER FLOOR PLAN

Conference Floor

Annual NANOS Reception and Banquet – Columbia Ballroom

Poster Session – Columbia Ballroom

Spouse/Guest Hospitality Suite – 900 West “Tea Room” Main Floor

Opening Reception – The Roof (15th Floor)
<table>
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<tr>
<th>Name</th>
<th>Affiliation and Location</th>
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<tr>
<td>Marie Acierno, MD</td>
<td>LSU Health Sciences Center, Baton Rouge, LA</td>
</tr>
<tr>
<td>Laura Balcer, MD, MSCE</td>
<td>University of Pennsylvania, School of Medicine, Philadelphia, PA</td>
</tr>
<tr>
<td>Jason Barton, MD, PhD, FRCP</td>
<td>VGH Eyecare Centre, Vancouver, BC, Canada</td>
</tr>
<tr>
<td>Jeffrey Bennett, MD, PhD</td>
<td>University of Colorado Denver, Aurora, CO</td>
</tr>
<tr>
<td>Beau Bruce, MD</td>
<td>Emory University, Atlanta, GA</td>
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<tr>
<td>Dean Cestari, MD</td>
<td>The Massachusetts Eye and Ear Infirmary, Boston, MA</td>
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<tr>
<td>Joseph G. Chacko, MD</td>
<td>Jones Eye Institute, University of Arkansas for Medical Sciences, Little Rock, AR</td>
</tr>
<tr>
<td>Sophia M. Chung, MD</td>
<td>St. Louis University Eye Institute, St. Louis, MO</td>
</tr>
<tr>
<td>Joseph Corbo, MD, PhD</td>
<td>Washington University School of Medicine, St. Louis, MO</td>
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<tr>
<td>Wayne Cornblath, MD</td>
<td>University of Michigan, Ann Arbor, MI</td>
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<tr>
<td>Fiona Costello, MD, FRCP</td>
<td>Calgary, AB, Canada</td>
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<tr>
<td>Kathleen Digre, MD</td>
<td>John Moran Eye Center, University of Utah, Salt Lake City, UT</td>
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<tr>
<td>Eric Eggenberger, DO, MSepi</td>
<td>Michigan State University, East Lansing, MI</td>
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<tr>
<td>Julie Falardeau, MD, FRCS(C)</td>
<td>Casey Eye Institute, Portland, OR</td>
</tr>
<tr>
<td>Edmond FitzGibbon, MD</td>
<td>National Eye Institute/NIH, Bethesda, MD</td>
</tr>
<tr>
<td>Steven Galetta, MD</td>
<td>University of Pennsylvania, Philadelphia, PA</td>
</tr>
<tr>
<td>Ari Green, MD</td>
<td>University of California, San Francisco, San Francisco, CA</td>
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<td>Jonathan C. Horton, MD, PhD</td>
<td>University of California, San Francisco, San Francisco, CA</td>
</tr>
<tr>
<td>Walter Jay, MD</td>
<td>Loyola University Medical Center, Maywood, IL</td>
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<tr>
<td>Lanning B. Kline, MD</td>
<td>University of Alabama, Birmingham, AL</td>
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<tr>
<td>Byron Lam, MD</td>
<td>Bascom Palmer Eye Institute, University of Miami School of Medicine, Miami, FL</td>
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<tr>
<td>Kiara Landau, MD</td>
<td>University Hospital Zurich, Zurich, Switzerland</td>
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<tr>
<td>Jeanne Le Ber</td>
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<tr>
<td>Andrew Lee, MD</td>
<td>The Methodist Hospital, Houston, TX</td>
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<tr>
<td>Richard Legge, MD</td>
<td>Omaha, NE</td>
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<tr>
<td>Y. Joyce Liao, MD, PhD</td>
<td>Stanford University School of Medicine, Palo Alto, CA</td>
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<td>Grant Liu, MD</td>
<td>University of Pennsylvania, Philadelphia, PA</td>
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<td>Nancy T. Lombardo</td>
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<tr>
<td>Neil Miller, MD</td>
<td>Wilmer Eye Institute, Johns Hopkins Hospital, Baltimore, MD</td>
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<tr>
<td>Mark Morrow, MD</td>
<td>Harbor-UCLA Medical Center, Torrance, CA</td>
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<tr>
<td>Raghur Mudumbai, MD</td>
<td>University of Washington, Seattle WA</td>
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<tr>
<td>Jeffrey Odel, MD</td>
<td>Edward S. Harkness Eye Institute, Columbia Presbyterian Medical Center, New York, NY</td>
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<tr>
<td>Paul H. Phillips, MD</td>
<td>University of Arkansas for Medical Sciences, Arkansas Children's Hospital, Little Rock, AR</td>
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<tr>
<td>Howard Pomeranz, MD, PhD</td>
<td>North Shore Long Island Jewish Health System, Great Neck, NY</td>
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<tr>
<td>Joseph Rizzo, III, MD</td>
<td>Massachusetts Eye and Ear Infirmary, Boston, MA</td>
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<tr>
<td>Janet C. Rucker, MD</td>
<td>Mount Sinai School of Medicine, New York, NY</td>
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<tr>
<td>Peter Savino, MD</td>
<td>Shiley Eye Center, University of California, San Diego, La Jolla, CA</td>
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<tr>
<td>Aseem Sharma, MD</td>
<td>Mallinckrodt Institute of Radiology, Washington University School of Medicine, St. Louis, MO</td>
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<tr>
<td>Robert Shin, MD</td>
<td>University of Maryland, Baltimore, MD</td>
</tr>
<tr>
<td>Richard Spaide, MD</td>
<td>Vitreous Retina Macula Consultants of New York, New York, NY</td>
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<tr>
<td>Susanne Trauettel-Klosinski, MD</td>
<td>University Eye Hospital, Tuebingen, Germany</td>
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<tr>
<td>Gregory P. Van Stavern, MD</td>
<td>Washington University in St. Louis School of Medicine, St. Louis, MO</td>
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<td>Judith Warner, MD</td>
<td>John Moran Eye Center, University of Utah, Salt Lake City, Utah</td>
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<td>Dean Wingerchuk, MD</td>
<td>Mayo Clinic, Scottsdale, AZ</td>
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<tr>
<td>Agnes Wong, MD, PhD, FRCS(C)</td>
<td>University of Toronto, The Hospital for Sick Children, Toronto, ON, Canada</td>
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NANOS would like to thank the following Supporters and Exhibitors for their financial support of these activities:

**2011 Supporters (as of 1-4-11)**

**Teva Neurosciences**
Teva Neurosciences has contributed $25,000

**Biogen Idec**
Biogen Idec has contributed $10,000 and supports the Wednesday breakfast

**Quark Pharmaceuticals**
Quark Pharmaceuticals has contributed $10,000

**2011 Exhibitors (as of 1-4-11)**

**Biogen Idec**

**Diopsys, Inc.**

**Elsevier Canada**

**Haag-Streit USA**

**Heidelberg Engineering**

**NovaVision**

**Richmond Products**
CME ACTIVITIES FACULTY DISCLOSURE STATEMENTS

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Disclosure information for Poster Presentations is listed in this syllabus at the end of each abstract.

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<td>Laura Balcer, MD, MSCE</td>
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<td>Jason Barton, MD, PhD, FRCPC</td>
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<td>Jeffrey Bennett, MD, PhD</td>
<td>EMD Serono, Teva Neuroscience, Biogen-Idec</td>
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<tr>
<td>Susanne Trauzettel-Klosinski, MD</td>
<td>STZ-Biomed</td>
<td>Scientific Consultant; No personal financial gain. Potential earnings will be reinvested in improving the software.</td>
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<td>Gregory Van Stavern, MD</td>
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<td>Dean Wingerchuk, MD</td>
<td>Alexion, Genzyme, Genentech</td>
<td>Research Support</td>
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PROGRAM PLANNER DISCLOSURE STATEMENTS

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Disclosures for members of these groups who are also speakers appear on the previous page.

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<td>Marie Acierno, MD</td>
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<td>Adeela Alizai, MD</td>
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<tr>
<td>Anthony Arnold, MD</td>
<td>Pfizer</td>
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<tr>
<td>Susan Benes, MD</td>
<td>Pfizer, Lundbeck, Ohio Ophthalmologic Society, State of Ohio Board of Optical Dispensers, Various Legal Firms</td>
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<td>Valerie Biousse, MD</td>
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<td>Swaraj Bose, MD</td>
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<td>Preston Calvert, MD</td>
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<td>Thomas Carlow, MD</td>
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<td>Deborah Friedman, MD</td>
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<td>Larry Frohman, MD</td>
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<td>Michael Lee, MD</td>
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<td>Leah Levi, MD, MBBS</td>
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<td>Timothy McCulley, MD</td>
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<td>Luis Mejico, MD</td>
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<td>Nancy Newman, MD</td>
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<td>Anil Patel, MD, FRCS(C), FACS</td>
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# PROGRAM PLANNER DISCLOSURE STATEMENTS, continued

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<td>Lyn Sedwick, MD</td>
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<td>Prem Subramanian, MD, PhD</td>
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<td>Speaker Expense Reimbursement/Contract</td>
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<td>Roger Turbin, MD</td>
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The following individuals are presenting during the Frank B. Walsh Session and/or Platform Presentation Session.

<table>
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<tr>
<th>FULL NAME</th>
<th>FINANCIAL SUPPORT</th>
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<tr>
<td>Marie D. Aciero, MD</td>
<td>None</td>
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<td>Rebekah Ahmed, MBBS</td>
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</tr>
<tr>
<td>Robert Avery, DO, MSCE</td>
<td>Dr. Avery received support from the National Institute of Health (NIH/NEI Loan repayment program; 1T32NS061779-01 Neurologic Clinical Epidemiology Training Program, PI: Laura Balcer, MD). Dr. Shah received support from the National Institute of Allergy and Infectious Diseases (K01 AI73729) and the Robert Wood Johnson Foundation under its Physician Faculty Scholar Program. Dr. Licht is supported by grants from the NINDS (NS-052380), the DANA Foundation and a gift from the June and Steve Wolfson Fund for Neurological Research. Dr. Huh received support from National Institute of Neurological Disorders and Stroke (NS053651).</td>
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<tr>
<td>Valerie Biousse, MD</td>
<td>Research to Prevent Blindness; Nat’l Institute of Health</td>
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<tr>
<td>Celia Chen, MBBS, MPH, FRANZCO</td>
<td>The study received research grant support from the Perpetual Trustee The Lindsay &amp; Heather Payne Medical Research Charitable Foundation.</td>
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<td>Fiona Costello, MD, FRCP</td>
<td>The Multiple Sclerosis Society of Canada</td>
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<td>Marc Dinkin, MD</td>
<td>None</td>
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<td>Mohammad Fouladvand, MD</td>
<td>Study designed and conducted by Chestnut Medical (V3) and we have received consulting fee for conducting study and data analysis.</td>
<td>We have received support and consulting fee for study from Chestnut. Dr. Kim Nelson has stock in Chestnut.</td>
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<tr>
<td>Clare Fraser, MBBS</td>
<td>Dr. Clare Fraser is supported by the Sydney Eye Hospital Alumni Travelling Fellowship.</td>
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<td>Jeffrey Gelland, MD</td>
<td>None</td>
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<td>Jennifer Graves, MD, PhD</td>
<td>Supported in part by NIH K24 EO108116; Dr. Gina Remington has received honoraria from Biogen and Teva. Dr. Peter Calabresi has served on the advisory boards or consulted for Biogen Idec, Genetech, Sereno, Teva, Novartis, Eisai, Vertex, and Amgenmune. Dr. Frohman has been a speaker for or received consulting fees for Biogen Idec, Teva, Athena, and Abbott Laboratories. Dr. Laura Balcer has received honoraria from Biogen-Idec and Bayer. Dr. Steven Galetta has received honoraria from Biogen-Idec and Teva.</td>
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<td>Joyce Ho</td>
<td>Medical Scholars Program (JKH), Burroughs Wellcome Foundation (YJL), Vice Provost Undergraduate Education Faculty Grant (YJL)</td>
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<td>Jonathan Horton, MD, PhD</td>
<td>Supported by NEI</td>
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<td>Thomas Hwang, MD, PhD</td>
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<td>Hong Jiang, MD, PhD</td>
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<td>Randy Kardon, MD, PhD</td>
<td>Department of Veterans Affairs, Rehabilitation, Research and Development Division and Iowa City VA Center of Excellence for the Prevention and Treatment of Visual Loss; Department of Defense (TATRC), Carver Translational Research Grant</td>
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<tr>
<td>Bradley Katz, MD, PhD</td>
<td>Supported by NIH Center for Excellence in Molecular Hematology F30 AG021334 and by NIH DK070947. Dr. Koeing is supported by PHS UL1-R0025764 and C06-R211234 and by an award from the Veterans Administration. Dr. Katz is a Jahnigen Scholar (American Geriatrics Society, New York, NY). Supported by a gift from Mrs Grace Stillwell and by a grant to the Department of Ophthalmology from Research to Prevent Blindness, Inc., New York, NY. The authors thank the members of the Neuro-Ophthalmology Giant Cell Arteritis (ND-GCA) Consortium who donated blood and tissue samples: Moran Eye Center, University of Utah, Salt Lake City, UT; Bradley J Katz, MD, PhD; Curry Koenig, MD; Kathleen B Digre; MD; Judith E A Warner, MD; Bhupendra Patel, MD; Chang Hee Kim, MD; Nick Mamalis, MD; Emory Eye Center, Atlanta, GA; Valerie Biousse, MD; Beau Bruce, MD; Nancy Newman, MD; University of Minnesota, Minneapolis, MN; Michael Lee, MD; Andrew Harrison, MD; Feinstein Institute for Medical Research/North Shore–Long Island Jewish Health System: Howard Pomeranz, MD, PhD; Kellogg Eye Center, University of Michigan, Ann Arbor, MI; Wayne Todd Connablath, MD; Jonathan Trobe, MD; University of Pittsburgh, Pittsburgh, PA; John Charley, MD; Jones Eye Clinic, Sioux City, IA; Jason Jones, MD; Wilmer Eye Institute, Johns Hopkins Hospital, Baltimore, MD; Prem Subramanian, MD; New York Eye and Ear Infirmary, New York, NY; Mark Kupersmith, MD; Jules Stein Eye Institute, University of California, Los Angeles, CA; Lynn K Gordon, MD, PhD; University Hospital Zurich, Zurich, Switzerland: Klara Landau, MD; Marc Toeteberg, MD; Pascal Knecht, MD.</td>
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<td>FULL NAME</td>
<td>FINANCIAL SUPPORT</td>
<td>FINANCIAL INTEREST DETAILS</td>
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| Melissa Ko, MD | Supported by unrestricted grants from research to Prevent Blindness, Inc, NYC, NY, and Lions District 20-Y1. | No financial interest.
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SATURDAY, FEBRUARY 5

12:00 p.m. – 5:00 p.m. Board Meeting

2:00 p.m. – 8:30 p.m. Registration

7:00 p.m. – 8:30 p.m. Opening Reception (all are welcome)

LOCATION

Board Room

Pacific Foyer

The Roof (15th Floor)

SUNDAY, FEBRUARY 6

6:30 a.m. – 5:30 p.m. Registration

6:30 a.m. – 7:45 a.m. Continental Breakfast

6:30 a.m. – 12:30 p.m. Exhibit Hall Open

8:30 a.m. – 10:30 a.m. Spouse/Guest Hospitality Suite

7:45 a.m. – 5:00 p.m. FRANK B. WALSH SESSION [6.75 CME]

Co-Chairs: Sophia M. Chung, MD & Gregory P. Van Stavern, MD

Neuropathologist: Joseph Corbo, MD, PhD

Neuroradiologist: Aseem Sharma, MD

7:45 a.m. – 5:00 p.m. FRANK B. WALSH SESSION [6.75 CME]

LOCATION

Pacific Ballroom

British Room

Vancouver Island Room

900 West “Tea Room”

11:40 a.m. – 1:20 p.m. Lunch on own – Please see local restaurant list for recommendations.

This session is designed to present a wide variety of neuro-ophthalmic cases to an audience of physicians with varying neuroscience backgrounds who have a common intellectual interest in the broad range of conditions that impact the human visual pathways and ocular motor systems.

The format is a clinicopathologic conference. Clinical cases will be presented by neuro-ophthalmologists with comments by a neuroradiologist, neuropathologist and other selected experts. Necropsy, surgical pathology, and neuroimaging will help illuminate clinical points. Cases will be discussed from clinical, anatomic, radiologic and pathologic aspects with emphasis on diagnosis, pathophysiology and management. Audience participation is encouraged.

At the conclusion of this program, participants should be able to: 1) Recognize the varied presentations of neuro-ophthalmic disease; 2) Correlate the anatomic localization and histopathologic appearance with the clinical presentations; 3) Effectively use radiologic procedures in diagnosis; 4) Recognize both the value and limitations of neuropathology; and 5) Discuss newly described diseases and their connection to neuro-ophthalmology.
<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
<th>Location</th>
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<tr>
<td>5:15 p.m. – 5:45 p.m.</td>
<td>Frank B. Walsh Committee Meeting</td>
<td>Tweedsmuir Room</td>
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<td>5:15 p.m. – 6:00 p.m.</td>
<td>Fellowship/Professional Standards Committee Meeting</td>
<td>Garibaldi Room</td>
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<td>5:30 p.m. – 6:30 p.m.</td>
<td>Resident/Student Program and Reception</td>
<td>Board Room</td>
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<td>Evening</td>
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<td>Time</td>
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<td>8:00 a.m. - 8:20 a.m.</td>
<td>Another Case of Guillain-Alajouanine-Garcin Syndrome [also known as Bertolotti-Garcin syndrome, Garcin’s syndrome, Garcin-Guillain syndrome, Hartmann’s syndrome (Friedrich Hartmann), Schmincke tumor-unilateral cranial paralysis syndrome…]</td>
<td>Valerie Biousse, MD</td>
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<td>8:20 a.m. - 8:40 a.m.</td>
<td>A Case of Recurrent Encephalopathy, Seizures and Retinopathy</td>
<td>Ryan Walsh, MD</td>
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<td>8:40 a.m. - 9:00 a.m.</td>
<td>Combing The Globe For Terrorism</td>
<td>Norah Lincoff, MD</td>
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<td>9:00 a.m. - 9:20 a.m.</td>
<td>Not a Benign Tumor</td>
<td>Hong Jiang, MD, PhD</td>
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<td>9:20 a.m. - 9:40 a.m.</td>
<td>Pseudo-Pseudo-Pseudotumor Cerebri</td>
<td>Marc Dinkin, MD</td>
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<td>9:40 a.m. – 10:00 a.m.</td>
<td>Coffee Break</td>
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<th>Time</th>
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<tr>
<td>10:00 a.m. - 10:20 a.m.</td>
<td>When Life Gives You Lymphocytes, Make Limeade</td>
<td>Melissa Ko, MD</td>
<td>27</td>
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<td>10:20 a.m. - 10:40 a.m.</td>
<td>Poor Visual Recovery Following Treatment of Panuveitis in a 62 Year-old Man with Biopsy-Proven Sarcoidosis</td>
<td>Jeffrey Gelfand, MD</td>
<td>29</td>
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<td>10:40 a.m. - 11:00 a.m.</td>
<td>To Biopsy or Not to Biopsy, That is the Question</td>
<td>Michael Yoon, MD</td>
<td>31</td>
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<tr>
<td>11:00 a.m. - 11:20 a.m.</td>
<td>A Case of Net Visual Loss and Gain</td>
<td>Thomas Slamovits, MD</td>
<td>33</td>
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<tr>
<td>11:20 a.m. - 11:40 a.m.</td>
<td>A Bright Spot Causing Darkness</td>
<td>Thomas Hwang, MD, PhD</td>
<td>35</td>
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<td>11:40 a.m. -1:20 p.m.</td>
<td>Lunch on own – see restaurant listing</td>
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</table>
Frank B. Walsh Session [6.75 HRS CME], continued

Session III
Moderators: Sophia M. Chung, MD & Joseph G. Chacko, MD

1:20 p.m. - 1:40 p.m.  What’s in a Name  
Nicholas Volpe, MD

1:40 p.m. - 2:00 p.m.  ‘Tis Nobler in the Mind to Suffer... or to Take Arms Against a Sea of Troubles and by Opposing End Them?  
(Hamlet act 3, scene 1)  
Heather Moss, MD, PhD

2:00 p.m. - 2:20 p.m.  Egg in Your Face  
Padmaja Sudhakar, MD

2:20 p.m. - 2:40 p.m.  Trouble Reading  
Rebecca Stacy, MD, PhD

2:40 p.m. - 3:00 p.m.  Bad Eyes, Bad Walking and Bad Judgement  
Clare Fraser, MBBS

3:00 p.m. – 3:20 p.m.  Coffee Break

Session IV
Moderators: Paul H. Phillips, MD & Julie Falardeau, MD, FRCS(C)

3:20 p.m. - 3:40 p.m.  Tics and Fleas  
Nancy Newman, MD

3:40 p.m. - 4:00 p.m.  Some Hairy Gossyp  
Ivey Thornton, MD

4:00 p.m. - 4:20 p.m.  Non-Functional Sinus Disease  
Julie Shelton, MD

4:20 p.m. - 4:40 p.m.  Twice Bitten, Once Shy  
Robert Shin, MD

4:40 p.m. - 5:00 p.m.  A Runny Nose  
Steven Newman, MD
HISTORY & EXAM:
A 37 yo white man presented with multiple right-sided cranial nerve palsies. PMHx included NIDDM, hypertension and sleep apnea. In 2004, he had a “benign soft mass” removed from the left side of his neck. He was a smoker (12 pack/y) and denied alcohol. Medications included metformin and losartan potassium-hydrochlorothiazide. He worked in a warehouse and used a forklift. Family history was remarkable for diabetes, glaucoma and thyroid disease.

In May 2007, while shooting a gun, he noticed right hearing loss, followed by right tinnitus and paresthesias involving the inside of his right ear. He presented locally with a “right temple mass draining pus”, which resolved spontaneously, and NIDDM was diagnosed. The right hearing loss persisted.

In September 2007, he complained of right facial hypoesthesia and right facial weakness with difficulty closing the right eye. The inside of his right nose and the right side of his gums felt numb. This worsened over a few weeks and he subsequently developed bilateral horizontal diplopia with a right abduction deficit.

By the end of December 2007, he also had occasional difficulty swallowing, change in his voice and he noticed altered taste on the right side of his tongue.

He stopped using the forklift because of episodic unsteadiness, but he was otherwise doing well with no other neurologic or systemic symptoms.

An ENT MD obtained a good quality brain and IAC MRI with contrast in December 2007, which showed only a large posterior fossa arachnoid cyst.

In January 2008, a neurologist documented right V, 1, 2, 3, VI, VII, VIII, IX, and X nerve palsies with otherwise normal neurologic examination. Repeat MRI of the brain, orbits, and skull base in February 2008 showed only the arachnoid cyst. A CT of the skull base with contrast and bone windows was also normal. A lumbar puncture was normal, with negative cytology. Chest X-Ray, ACE, CBC, ESR, RPR, RF, ANA, Lyme were normal or negative.

We first saw the patient in April 2008 with the previously described clinical findings. Review of all the imaging by our neuro-radiologists confirmed that they were normal; a very subtle enhancement of the lateral aspect of the basilar artery was noted, but was not interpreted as abnormal. Numerous repeat imaging studies, blood tests and lumbar punctures performed over the subsequent few months remained normal. The patient’s cranial nerve palsies persisted, in association with mild ataxia…

FINANCIAL DISCLOSURE: Research to Prevent Blindness; National Institute of Health
Another Case of Guillain-Alajouanine-Garcin Syndrome
[also known as Bertolotti-Garcin syndrome, Garcin’s syndrome, Garcin-Guillain syndrome, Hartmann’s syndrome (Friedrich Hartmann), Schmincke tumor-unilateral cranial paralysis syndrome...]

Answer

FINAL DIAGNOSIS:
Primary glioma stage IV, presenting with isolated cranial nerve palsies (Garcin syndrome) and normal neuro-imaging for 17 months.

SUMMARY OF CASE INCLUDING PATHOLOGY:
Unilateral multiple cranial nerve palsies were highly suggestive of a skull base infiltrative process, and repeat extensive evaluations and close follow-up were performed over the following year:

- The long course (11 months between first sign and our evaluation) and the absence of general symptoms or signs made an infection very unlikely. Repeat LP, CBC, ESR, CRP and CT of sinuses were normal.
- Work-up for sarcoidosis and Wegener’s granulomatosis remained negative (repeat ACE, many chest X-Rays, two chest CTs, one lung biopsy, one whole body FDG PET-CT, ANCA, UA). A trial of steroids did not improve the patient’s signs.
- Paraneoplastic panel was negative. A systemic neoplasm was not found. General examinations (including testicles), nasopharynx and larynx endoscopies, remained normal. CT of the chest, abdomen and pelvis, testicle US, and whole body FDG PET-CT (June 2008) were normal.
- Repeat MRIs (May and July 2008) of the brain, skull base and cranial nerves with contrast remained normal except for the arachnoid cyst and the subtle enhancement of the lateral aspect of the basilar artery.
- A CT-Cisternogram performed in June 2008 to rule out a prepontine epidermoid cyst and to check that the cerebellar subarachnoid cyst was not lobulated, was normal.
- Two other lumbar punctures were normal in May and in July 2008 (with negative cytology and flow cytometry).

In July 2008, his balance deteriorated and he stopped working. Repeat neurologic consultation suggested a right cerebellar syndrome, persistent right V1,2,3, VI, VII, VIII cranial nerve palsies and now obvious right IX, X, XI, XII palsies. Repeat MRI was unchanged. A right superior orbital nerve biopsy was performed (and was unremarkable).

In October 2008, he had a transient episode of confusion with generalized weakness and he was admitted to neurology. Repeat brain MRI with contrast showed enlargement and enhancement of the right prepontine VII and VIIIth cranial nerves. There were no parenchymal lesions and no leptomeningeal enhancement (this is the first definitely abnormal MRI, 17 months after onset of symptoms).

Repeat LP again was normal with negative cytology. Repeat paraneoplastic panel and many many blood and CSF tests remained normal or negative. Repeat Chest CT showed small bilateral pulmonary nodules. Lung biopsy was normal. The patient refused a biopsy of the right VII/VIII cranial nerves and he was given steroids without improvement of his cranial nerve palsies. He remained stable until February 2009, at which time he had an episode of GI bleed with severe anemia requiring transfusions. Upper and lower GI endoscopies did not show any mass. His ataxia worsened.

In March 2009, he became confused and unable to walk because of severe cerebellar syndrome. He was intubated because of respiratory distress. Repeat MRI showed worsening of the preponine enhancement noted in October 2008 as well as new areas of T2 and FLAIR hyperintensity extending from the medulla into the pons, right brachium pontis and superiorly into the left cerebral peduncle. A suboccipital craniotomy was performed for cerebellar biopsy. Frozen section revealed a high grade glioma, and this was confirmed on pathologic examination. Cerebellar biopsy showing a densely cellular malignant neoplasm infiltrating the cerebellar parenchyma. The tumor cells have fibrillar eosinophilic cytoplasm and markedly anaplastic nuclei, with multinucleated tumor giant cells and there are foci of necrosis. These morphologic findings are diagnostic for glioblastoma, WHO grade IV.

His clinical status continued to deteriorate. The patient died two weeks later; no autopsy was performed.

Despite a very high clinical suspicion for an infiltrative process, repeat high-quality neuroimaging read by an expert neuro-radiologist remained normal for 17 months. The cranial nerve palsies remained isolated with only mild ataxia for 18 months prior to rapid clinical worsening corresponding to radiologic evidence of brainstem and cerebellar lesions. Garcin, Guillain, and Alajouanine (3 famous French Neurologists in the 1920’s) were correct: chronic isolated multiple unilateral cranial nerve palsies are related to infiltrative neoplasms! Even modern neuro-imaging may be unrevealing in this setting!

KEYWORDS: Cranial Nerve Palsies, Neoplasm, Glioblastoma

REFERENCES:
A Case of Recurrent Encephalopathy, Seizures and Retinopathy

Ryan Walsh, Michael Stewart, Paul Brazis
Mayo Clinic Florida, Jacksonville, FL, United States

HISTORY & EXAM:
A 45 year-old right-handed male was transferred from an outside hospital for further evaluation of “possible encephalitis”. At age 44 he was diagnosed with right hemisphere encephalitis when he presented with seizures, altered mental status, and left hemiparesis. Brain MRI at that time demonstrated right hemispheric edema; brain biopsy showed a non-specific chronic inflammatory infiltrate. A clear etiology was not determined and he was empirically treated with dexamethasone with slow improvement to near baseline. Two weeks prior to his current presentation, he experienced an apparent seizure with speech arrest and disorientation. He presented to his local hospital where an EEG showed diffuse slowing with low amplitude, worse in the left hemisphere. MRI of the brain showed extensive gliosis in the right parietal, temporal, and occipital lobes, as well as bilateral thalami, cerebral peduncles, midbrain, and pons. He was treated with IV dexamethasone and transferred to our hospital for further evaluation.

Past medical history was significant for a “learning disability” and a complex partial seizure disorder diagnosed 20 years earlier and managed with a single anti-epileptic drug. He was diagnosed with macular degeneration at age 43. Family history was unremarkable. He quit smoking 1 year prior to presentation after a 20-pack-year history, and his alcohol use was minimal.

Neurologic examination demonstrated a Glasgow Coma Scale score of 11; eye opening 4, motor response 5 (left arm), and verbal response 1. He was essentially nonverbal and only had occasional ability to follow 1-step commands. He blinked to threat bilaterally. There was right lower facial weakness and flaccid paralysis of the right upper and lower extremities with full strength on the left. Deep tendon reflexes were asymmetrically diminished on the right and he had positive Babinski reflexes bilaterally.

Ophthalmologic examination was remarkable for retinal pigment epithelium loss in a “bull’s eye” pattern bilaterally.

FINANCIAL DISCLOSURE: NONE
A Case of Recurrent Encephalopathy, Seizures and Retinopathy

Answer

FINAL DIAGNOSIS:
α-methyl-acyl-CoA racemase deficiency

SUMMARY OF CASE INCLUDING PATHOLOGY:
This 45 year-old male presented with seizures, recurrent encephalopathy, diffuse neurologic deficits, and bilateral retinopathy. Etiologic considerations are many including a myriad of metabolic, infectious, and degenerative conditions. Multiple investigations were undertaken. MRI of the brain with contrast demonstrated gyral T1 and T2 hyperintensity of the right temporal, parietal and occipital lobes consistent with laminar necrosis; linear enhancement of the left parietal and occipital lobes and diffuse T2 hyperintensity throughout the left hemisphere; and symmetric regions of T2 hyperintensity of the thalamus, midbrain and pons. EEG demonstrated diffuse slowing and several seizures emanating from the left and right central temporal regions. EMG with nerve conductions showed a mild peripheral neuropathy. Previous brain biopsy was consistent with a chronic necrotizing encephalitis with leukoencephalopathy.

Extensive laboratory testing showed evidence of primary hypogonadism. CSF evaluation was unremarkable. Very long chain fatty acid analysis showed an elevated pristanic acid level (51.47 umol/L, normal <2.98) and normal phytanic acid level (0.77, normal <9.88), resulting in a markedly elevated pristanic-to-phytanic acid ratio (67.02, normal <0.39); straight chain very long chain fatty acids were normal. Molecular analysis of the α-methyl-acyl-CoA racemase (AMACR) gene revealed homozygosity for c.154>C mutation.

The patient was diagnosed with AMACR deficiency, a rare autosomal recessive peroxisomal disorder caused by a defect in AMACR, an enzyme necessary for the catabolism of pristanic acid and the synthesis of bile acids. Only 6 cases (4 adults and 2 children) have previously been reported, with all but one demonstrating the c.154>C mutation. Phenotypically, AMACR deficiency is somewhat heterogeneous with the adult-onset form bearing some resemblance to Refsum disease. Pigmented retinopathy has been described in two previous cases. Ours is the first to document the retinal findings with retinal photography. Treatment with dietary restriction of pristanic and phytanic acids has had variable success.

AMACR deficiency is a rare peroxisomal enzyme disorder with a heterogeneous and incompletely defined phenotype. Our patient’s presentation, which included diffuse neurologic deficits (including peripheral neuropathy) and retinopathy, is reminiscent of Refsum disease, however phytanic acid levels were normal. Had pristanic acid levels not been checked, the correct diagnosis likely would have been missed. Ophthalmologic examination was crucial in directing the appropriate evaluation.

KEYWORDS: Retina, Metabolic, Seizure, Retinal Disorders, Hereditary

REFERENCES:
HISTORY & EXAM:
An 83 year old gentleman presented with painful blindness in his left eye in 8/09. He was initially seen by his ophthalmologist with count fingers vision, swelling of his L optic nerve, mild lid swelling and pain. He was diagnosed with NAION. His pain was so severe he necessitated hydrocodone/APAP, propoxyphene and tramadol to control it. A CT scan was read as normal. The patient was placed on oral prednisone by a local neurologist. Prednisone alleviated the pain about 50% allowing lowering of the narcotic medication.

The patient's PMHx was significant for HTN, Afib, type II DM, squamous cell carcinoma of the face, and prostate cancer treated with seed implants in 8/02. His medications included warfarin, quinapril, simvastatin, tamsulosin, diltiazem and pantoprazole.

The patient presented to our institution 2 weeks later with signs of a complete retinal artery and vein occlusion with NLP vision. His periocular headache pain was still excruciating causing anorexia. His lid edema had resolved but he showed 1 mm of proptosis with mild conjunctival injection on the L side. His right eye and field were normal with 20/20 vision despite cataract. His IOP's measured 12 bilaterally.

Laboratory testing showed an ESR of 2, a CRP of 0.85 and a normal CBC, RPR, ACE, ANCA p and n. HA1c was 7.5. MRI imaging revealed complete optic nerve thickening and sheath enhancement. An LP revealed no cells and an elevated protein of 65. The patient went on to severe recurrent iris neovascularization which required repeat injections of Bevacizumab (Avastin) to control.

A temporal artery biopsy was performed which revealed extensive disruption of the elastic lamina interna but no arteritis (day 20 of prednisone). A PET scan performed in 11/09 revealed only an area of hypermetabolism measuring 16mm in the area of the left thyroid cartilage. The thyroid lesion revealed a colloid nodule and lymph node aspirations were negative for disease.

Ten months after his initial presentation the patient reported new painless visual disturbance in his good eye. Follow up imaging and a biopsy were performed.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Optic Nerve Glioblastoma Multiforme

SUMMARY OF CASE INCLUDING PATHOLOGY:
Follow up MRI showed progression of the nerve enhancement and thickening beyond the canal as well as involvement of the chiasm and L optic nerve tract. The optic nerve was now normal in caliber within the orbit. Due to new visual loss and changes in the visual field in the R eye he went on to biopsy.

The biopsy revealed Glioblastoma Multiforme of the optic nerve with extension into the chiasm. Intraoperative photos pre and post treatment were obtained.

Malignant optic glioma of adulthood is an extremely rare optic pathway tumor. Till 2004 only 45 cases of adult malignant optic gliomas had been described. Of these patients 51% were male, 49% females; the mean age at diagnosis was 54 years. The sites of occurrence of malignant optic glioma is usually in the optic chiasm. The malignant optic glioma of adulthood is classified pathologically as either an anaplastic astrocytoma or a glioblastoma multiforme. In adults, malignant optic gliomas are rare and are rapidly fatal. They represent 2.5% of all deaths due to cancer. Survival depends on tumor grade, resectability, and the age of the patient. Mean survival from the time of presentation is usually less than 1 year. Radiation therapy and chemotherapy with temozolomide (alkylating agent) following surgery is still standard of care. Bevacizumab has shown to reduce vasogenic edema in these cases but is still very controversial because of case reports suggesting the risk of catastrophic tumor recurrence.

Malignant glioma of the optic nerve is extremely rare. This tumor originated in the area of the canal where it outgrew it space and likely self infarcted explaining the delay in obvious progression. Intraocular Bevacizumab may also have partially treated the tumor by systemic effect of the drug. The severe pain and CRAO/CRVO certainly suggested tumor as the disease process but once the enhancement of the optic nerve resolved no tissue was felt to be obvious to biopsy. The patient's warfarin, and partial response to prednisone also made surgeons hesitant to biopsy early.

KEYWORDS: Glioblastoma Multiforme, Optic Pathway Glioma, Painful Blindness, Combined Retinal Artery And Vein Occlusion, Avastin

REFERENCES:
HISTORY & EXAM:
A 43-year-old African American woman was evaluated for blurred vision for 2 months in 6/2010.

She has a 10-year history of recurrent colon cancer, which was treated with 3 colon resections and radiation therapy. She had multiple chemotherapies in the past, which was restarted in 5/2009 with combination of Irinotecan and panitumumab. Irinotecan was discontinued in 11/2009, she continued with panitumumab until 2/2010. The patient has been closely followed up by her oncologist. Her extended workups including MRIs, PET scans, bone scans were unremarkable, except an incidental finding of a pituitary mass (about 12 mm) on brain MRI in 5/2009. She was considered to have a small pituitary adenoma and recommended by Neurosurgery to have a follow up Brain MRI in 6 months. The follow up MRI six months later showed a decrease in size of the tumor to 8mm. In 4/2010, the patient began to develop blurry vision OD, followed by headaches, dizziness, extreme thirst, frequent urination and profound fatigue. She was diagnosed as hypopituitarism and diabetes insipidus secondary to the pituitary tumor. 2 months later, she had blurry vision OU. Her vision dropped from 20/20 OU to 20/60 OD and 20/40 OS over one month, and had temporal visual field depression of left eye and diffuse depression of visual field in right eye. Brain and orbital MRI showed the pituitary tumor had doubled in size with suprasellar extension and severe chasmal compression, extension into the third ventricle. A procedure was performed.

FINANCIAL DISCLOSURE: NONE
Not a Benign Tumor
Answer

FINAL DIAGNOSIS:
Metastatic colon cancer to the pituitary gland.

SUMMARY OF CASE INCLUDING PATHOLOGY:
Emergent transsphenoidal decompression of the pituitary mass was performed in 7/2010. Pathology showed metastatic colon carcinoma in the pituitary gland. The carcinoma stained strongly for CDX2 (nuclear immunostain) and Villin and was positive when stained with cytokeratin 20 and negative for cytokeratin 7. The patient received radiation therapy right after the transsphenoidal decompression, but vision worsened post radiation. Repeat MRI two weeks later showed regrowth of the mass in pituitary. She was placed on high dose steroids and panitumumab was restarted. Her vision gradually improved. Her repeated MRI brain in 9/2010 showed interval shrinkage of the suprasellar component of the mass. Her most recent follow-up in 10/2010 demonstrated 20/25 OU and near normal visual fields.

Pituitary metastatic cancer occurs rarely in cancer patients, the most common metastatic cancers are from lung and breast.\(^1\)\(^2\)\(^3\) To our knowledge, there is only one other case of metastatic colon cancer to the pituitary gland reported.\(^4\) However, when a patient with known cancer develops diabetes insipidus and has radiographic evidence of a pituitary mass, the diagnosis of metastasis should be suspected. Given longer survival rates in patients with colon carcinoma, the frequency of metastases to rare sites may become more frequent in the future. In retrospect, panitumumab \(^5\) (an anti-epidermal growth factor receptor antagonist) probably helped to control the growth of the pituitary metastasis until it was discontinued in 2/2010, and after radiation therapy in 9/2010. While panitumumab has not been reported to have activity on metastatic colon cancer to pituitary, our case suggests panitumumab has therapeutic activity on metastatic cancer in central nervous system.

A pituitary incidentaloma in a colon cancer patient first decreased and then increased in size causing severe vision loss. Surgical resection showed colon cancer metastasis to the pituitary gland, which is seldom reported with colon cancer. The initial “spontaneous” improvement was in retrospective probably secondary to panitumumab treatment which has not been reported to have effect on colon cancer metastasis to brain. The case lends panitumumab as a novel therapy for metastatic colon cancer to brain.

KEYWORDS: Metastatic Colon Cancer, Pituitary Gland, Vectibix

REFERENCES:
HISTORY & EXAM:
A 27 year old man with migraines since childhood developed white-outs of vision lasting seconds with changes in position, worsening headaches, trouble focusing and subjective blind spots. He was diagnosed with migraine and treated with sumatriptan. He developed blurry vision and was found to have papilledema. Acuities were 20/50 OD and 20/25 OS. There were superonasal defects OD>OS. Brain MRI was negative. MRV was performed and showed mild transverse sinus stenosis without thrombosis. Lumbar puncture showed clear contents but an increased opening pressure of 40 cm of water. He was diagnosed with idiopathic intracranial hypertension (IIH) and started on acetazolamide 250 mg BID. Ten days later, he developed acute, severe decline in vision OU. Visual acuity fell to CF OD and 20/30 OS. The acetazolamide dose was doubled. A lumbar-peritoneal shunt was placed but a few days later, he developed confusion, an excruciating headache and vomiting. CT venogram showed superior sagittal and right transverse sinus thrombosis. He was started on anticoagulation with heparin to warfarin. Shunt pressure was 12 cm of water. He then presented to our service. On exam, best-corrected VA was 20/600 OD and 20/50 OS. Perimetry showed severe field loss OU with only relative temporal sparing OS. There was an RAPD OD and optic atrophy OD>OS. Blood tests were drawn. Four months later, a diagnostic procedure was performed.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Increased intracranial pressure secondary to dural arteriovenous malformations which worsened with the secondary development of venous sinus thrombosis.

SUMMARY OF CASE INCLUDING PATHOLOGY:
Blood tests for hypercoagulability showed a mutation (one copy of 4G allele) in the plasminogen activator inhibitor-1 (PAI-1) gene. Conventional angiogram was performed and showed a dural arteriovenous fistula (AVF) with arterial supply from the bilateral middle meningeal arteries, the right superficial temporal artery and the anterior falx artery off the left ophthalmic artery. Venous drainage was into the superior sagittal sinus. The first three feeders were closed with catheter-based embolization and the fourth was clipped. The patient remained on warfarin and has been without headaches or new visual changes since.

The case serves as a reminder that dural AVFs should be considered in cases of IIH and otherwise unexplained venous sinus thrombosis (VST). It is unique in that it demonstrates a radiological presentation of VST that followed treatment for what appeared to be IIH with an initially normal venogram. This was further compounded by the discovery of an extensive dural AVF that was the likely cause of the increased ICP and eventually the thrombosis.

Dehydration associated with acetazolamide in a patient with an AVF and a genetic propensity toward hypercoagulability, may have contributed to the development of the venous sinus thrombosis and ensuing worsening of his clinical course.

The original presentation fulfilled the criteria for IIH, defined as elevated ICP with normal MRI and lumbar puncture contents. The original MRV showed only transverse sinus narrowing commonly seen in IIH, underlining the fact that this modality may miss dural AVMs. These factors led to the first wrong diagnosis of IIH. As he progressed, CTV led to a second incomplete diagnosis of VST, but the primary abnormality was only seen with conventional angiogram.

KEYWORDS: Idiopathic Intracranial Hypertension, Venous Sinus Thrombosis, Dural Arteriovenous Fistula, Hypercoagulable State, Papilledema

REFERENCES:
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When Life Gives You Lymphocytes, Make Limeade

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HISTORY & EXAM:
A 62 year-old woman with hypercholesterolemia presented with acute onset horizontal diplopia on awakening. Ophthalmic examination in the ED demonstrated: VA 20/30 OU and a left abduction deficit of 14 prism dipters. Head CT showed small vessel ischemic disease. MRI brain revealed non-specific lesions in the periventricular white matter. She was thought to have an ischemic left sixth nerve palsy, instructed to continue on aspirin and discharged home.

At her initial neuro-ophthalmic consultation a week later, her diplopia had worsened. Examination revealed: VA: 20/20 OU with a complete left abduction deficit. Extensive blood work was ordered.

Four days later, the patient presented to the ED with left facial weakness that developed over one day. Examination showed a left lower motor neuron facial palsy. During her hospital admission: the lumbar puncture showed: CSF WBC 310, a protein of 101, and a glucose of 51. CSF electrophoresis was suspicious for a faint monoclonal band. Cytology revealed abundant atypical lymphocytes with plasmacytoid characteristics and mitotic figures; the sample was not viable for flow cytometry. Studies for syphilis, cryptococcus, HSV, and bacterial growth were negative. Repeat MRI brain showed no significant changes. MRA of the brain was unremarkable. The hospitalist treated the patient with one dose of IV solumedrol. The evening prior to discharge, the ELISA and Western Blot IgM for Borrelia burgdorferi returned positive with a negative IgG. She was treated with IV ceftriaxone and discharged home on twenty-one days of outpatient IV antibiotic.

Repeat lumbar puncture performed 3 days later demonstrated 520 WBC, a protein of 81, and a glucose of 61. The cytology was unchanged. Flow cytometry showed 84% T-cells with normal CD4/CD8, 11% B-cells with lambda excess and B lymphocyte monoclonal expansion possibly consistent with non-Hodgkin's lymphoma. A procedure was performed.

FINANCIAL DISCLOSURE: Supported by unrestricted grants from research to Prevent Blindness, Inc, NYC, NY, and Lions District 20-Y1.
FINAL DIAGNOSIS:
Lyme Neuroborreliosis

SUMMARY OF CASE INCLUDING PATHOLOGY:
Initial Brain MRI imaging demonstrated non-enhancing foci of FLAIR hyperintensity in the subcortical and deep white matter, and the left frontal lobe.

A comparison of post contrast T1-weighted MRI brain images taken on initial presentation (A) and two weeks later (B) demonstrated no significant interval change.

Giemsa stain of CSF from the second spinal tap at 200x. An abundance of small and atypical lymphocytes were noted.

Multiple magnifications of the preceding sample with Papanicoulaio stain at 500x(A &B), Giemsa stain at 1000x (C) and Wright Giemsa stain at 1000x(D). The specimen was highly cellular with abundant small lymphocytes and plasmacytoid cells. Bi-nucleated cell forms (A & C) and mitotic figures were seen. The presence of immunoblasts (D) was suspicious for a lymphoproliferative disorder.

Histopathological examination of bone marrow with H +E stain at 500x with normal cell morphology.

Protein electrophoresis of the second CSF sample (A) and bone marrow (B). Immunoglobulin heavy chain PCR (A) demonstrated a monoclonal band of approximately 240 bp with primers for the Framework 2 (FR2) region. Analysis of the bone marrow (B) one week later showed a diminished band of comparable size.

Hematology-oncology performed a bone marrow biopsy which showed normal morphology and a faint monoclonal band on electrophoresis. CT imaging was negative for primary malignancy. The patient was followed with serial examinations and lumbar punctures. The post-antibiotic CSF demonstrated WBC 39, protein of 37, and glucose of 67, with small lymphocytes, monocytes and smudge cells.

The patient gradually improved after antibiotics were initiated. Five months after initial presentation, the neuro-ophthalmic exam showed resolution of the sixth and seventh nerve palsies. CSF analysis at that time demonstrated WBC 0, a protein of 31, and a glucose of 65.

In its initial stages, Lyme neuroborreliosis can be challenging to distinguish from neoplastic etiologies such as non-Hodgkin's lymphoma due to an aggressive inflammatory response. Our patient was seropositive for Lyme disease but also had CSF demonstrating monoclonal expansion that was highly suspicious for lymphoma. This led to an extensive oncological workup. Previous reports suggest a possible molecular mimicry between Lyme disease and lymphoma, indicating a common etiopathological relationship.

KEYWORDS: Lyme Neuroborreliosis, Lymphoma, Diplopia, Sixth Cranial Nerve Palsy, Facial Weakness

REFERENCES:
HISTORY & EXAM:
A 62 year-old African-American man presented with progressive falling and confusion. Lumbar puncture revealed meningitis (75 WBC, 0 RBC, initially neutrophilic, then lymphocytic predominance), with low glucose, elevated protein, >5 OCBs and IgG index 0.7. Lung and hilar lymph node biopsies demonstrated non-caseating granulomatous inflammation consistent with sarcoidosis. A ventriculoperitoneal shunt was placed for worsening hydrocephalus. Over the next 4 years, he developed lupus pernio, panuveitis and worsening interstitial lung disease. Treatment was initiated with pulse steroids, followed by high dose oral steroids and methotrexate, and most recently with mycophenolate mofetil, hydroxychloroquine and low dose oral steroids.

In early 2010, the patient developed visual blurring in the left eye. He was found to have panuveitis OS and treated with steroid eye drops. He subsequently received an intravitreal steroid injection. Three weeks later, the uveitis had improved, but he was 20/200 OS. IOP was 8. A dilated fundus examination showed a sharp disc with no other retinal abnormalities to explain the visual loss. However, a new afferent pupillary defect was noted in the left eye. Full-field pattern reversal VEPs revealed a P100 latency delay OS. MRI of the orbits (performed without contrast due to chronic kidney disease) revealed FLAIR hyperintensity of the prechiasmatic left optic nerve with associated nerve enlargement.

The patient was treated for presumed sarcoid optic neuritis with IV solumedrol x 3 days, but unfortunately had no improvement in vision. Neurological exam remained stable. He received infliximab 5 mg/kg for treatment of refractory sarcoid optic neuropathy in the context of worsening pulmonary sarcoidosis. On reexamination 1 week later, he had no light perception OS.

Two weeks later, he reported new painless visual blurring OD, accompanied by worsening gait instability. A dilated fundus examination (see photos) showed retinal vasculitis OD with sheathing, focal areas of whitening, retinitis, and cystoid macular edema OD. The left eye showed scattered intraretinal hemorrhages anterior to the equator and a small region of retinal whitening in the nasal periphery only (not visible in fundus photos).

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:

1) Progressive Outer Retinal Necrosis OU due to varicella zoster virus infection, with associated VZV meningitis.
2) Likely infectious optic neuropathy OS due to varicella zoster virus, masquerading initially as sarcoid optic neuropathy.

To our knowledge, this is the first reported case of infectious progressive outer retinal necrosis in sarcoidosis. This association invites caution about how best to evaluate optic neuropathy in patients with known inflammatory disease and reinforces the importance of ruling out infectious causes of optic neuropathy in the presence of atypical clinical features of optic neuritis.

SUMMARY OF CASE INCLUDING PATHOLOGY:

An anterior chamber paracentesis was performed; the VZV PCR was positive. Cerebrospinal fluid examination revealed 130 WBC (47% N, 28%L, 25%M), glucose 100, total protein 264 and a positive VZV PCR. Repeat MRI of the orbit showed extension of the FLAIR hyperintensity into the left optic tract (with persistence of the FLAIR hyperintensity in the left prechiasmic optic nerve and chiasm). There were no new parenchymal brain lesions. MRI of the spine was unremarkable. HIV was negative.

He was treated with IV ganciclovir and foscarnet, followed by intravitreal ganciclovir and foscarnet, and ultimately with an intravitreal ganciclovir implant, pars plana vitrectomy, silicone oil to prevent retinal detachment and laser retinopexy to create a prophylactic barrier around the macula.

For management of immunosuppression, he received one dose of IV solumedrol 1 gram, and then was rapidly tapered to 20 mg prednisone/day. Mycophenolate mofetil was immediately stopped, and infliximab was discontinued. On discharge to a skilled nursing facility 3 weeks later, he had an island of macula capable of 20/400 vision OD, and the retinopathy appeared quiescent. The visual acuity remained NLP OS.

The afferent pupillary defect immediately pointed towards the additional process of an optic neuropathy in addition to panuveitis, which was supported by full-field VEP and MRI. At that time, a dilated retinal examination did not suggest retinitis or vasculitis, and his neurological exam was stable.

Our presumption was that this patient with known sarcoidosis had developed sarcoid optic neuritis, and we offered aggressive treatment to try to preserve vision in that eye (and to reduce the risk of recurrent panuveitis in the other eye). Only after he lost vision in the previously unaffected eye, and continued to worsen from the optic neuropathy, did the suggestion of an infectious optic neuropathy and diagnosis of infectious progressive outer retinal necrosis become readily apparent.

KEYWORDS: Sarcoidosis, Optic Neuropathy, Infection, Meningitis, Steroids

REFERENCES:

To Biopsy or Not to Biopsy, That Is the Question
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HISTORY & EXAM:
A 16 year-old female honor student presented for evaluation of vertigo, headache, mild aphasia, and blurred vision. Eight weeks prior to presentation, she experienced 3 – 4 days of intense vertigo, which waxed and waned. Her bifrontal headache initially fluctuated, but by the time of presentation was persistent. Her parents felt she understood language normally but was unable to remember words. Visual symptoms were vaguely characterized as blurriness with intermittent diplopia. She occasionally stumbled while walking, which she attributed to difficulty seeing.

On ophthalmic examination, visual acuity was 20/20 OU with no RAPD. EOM showed trace limitation of abduction OS. External and anterior segment examinations were normal. DFE revealed moderate bilateral disc edema.

There was a right homonymous hemianopia and left inferior quadrantanopia on Humphrey visual field testing. There was peripheral constriction within remaining quadrant.

Neurologic examination revealed normal sensation and strength in the face and extremities. Cerebellar function, deep tendon reflexes, and gait were normal. Her speech was fluid with poor naming of low frequency objects. Comprehension was moderate (slow to do three-step commands). Reading was poor, and she was able to write simple sentences.

On MRI, there were two large areas of extensive FLAIR hyperintensity in the left temporal and occipital lobes, extending into the splenium of the corpus collosun and within the cuneous of the right occipital lobe. There was peripheral enhancement of the lesions with linear reduced diffusion along the leading central edges, and T2 intensity of the central portions of the lesions. Spinal MRI was normal.

LP, performed at the referring hospital, showed normal protein (53), glucose (36), WBC (1), and RBC (1). Opening pressure was not recorded. Cytology was not performed. IgG index was normal. Oligoclonal bands were absent. Other unremarkable CSF studies included: HSV and CMV, myelin basic protein, JC virus DNA, and AFB smear. Unremarkable serologic studies included: WBC, Hemoglobin, Hematocrit, Platelets, Ferritin, ACE, ANA, SS-A/SS-B, dsDNA antibody, HIV, RPR, RF, very long chain fatty acids, Lyme and NMO antibodies.

In the midst of much disagreement among services, a diagnostic procedure was performed.

FINANCIAL DISCLOSURE: NONE
To Biopsy or Not to Biopsy, That Is the Question
Answer

FINAL DIAGNOSIS:
Tumefactive demyelinating lesion

SUMMARY OF CASE INCLUDING PATHOLOGY:
Uncomplicated stereotactic guided biopsy of the right occipital lesion was consistent with demyelinating disease. There was a dense infiltrate of macrophages associated with interspersed reactive hypertrophic astrocytes. There were a few scattered small lymphocytes in a perivascular distribution. Luxol fast blue/PAS immunohistochemical staining showed a diffuse, extensive loss of myelin, while neurofilament stains demonstrated relative preservation of with rare swollen axons. There were no malignant cells or evidence of lymphomatous infiltration.

Tumefactive demyelinating lesions are rare in the pediatric population. Causative diseases include multiple sclerosis, acute disseminated encephalomyelitis, neuromyelitis optica, and myelinoclastic diffuse sclerosis (Schilder’s disease). Radiographic appearance of these lesions can mimic tumors or infections, making the diagnosis on imaging challenging. Typically, on MRI there are ill-defined borders, contrast enhancement with an “open ring” appearance, mild mass effect, and dilated vascular structures within the lesion. Diffusion weighted imaging and MR spectroscopy have been suggested and may prove useful in differentiating these lesions.

Elevated intracranial pressure (ICP) in patients with multiple sclerosis has been described by Newman et al. In their series, three patients with non-tumefactive MS had documented elevated ICP with papilledema, although the etiology of this was unclear. Most published reports on tumefactive demyelinating lesions have not described the ophthalmoscopic appearance of the optic discs. The mechanism in tumefactive MS is likely similar to cases without tumefaction, although one might speculate an additional mass effect.

Following biopsy, methylprednisolone 1g IV daily x 5 days was initiated. Over the next several days, she slowly improved with stable ophthalmologic examination. Two months later, follow-up revealed acuity of 20/20 OU. Her visual field showed slight improved with normalization of the left superior quadrant, and some improvement in the left inferior quadrant. Disc edema resolved completely without atrophy.

This case presented many challenges, including the difficult decision whether to perform a brain biopsy or treat empirically with steroids. Repeat LP with cytology was deemed too dangerous for risk of herniation and steroid could limit the diagnostic yield of biopsy.

KEYWORDS: Papilledema, Demyelinating Disease, Retrochiasmal

REFERENCES:
A Case of Net Visual Loss and Gain

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HISTORY & EXAM:
A 68 year old computer programmer became aware of sudden visual loss in both eyes in January. She initially assumed that this was related to her anemia (her hemoglobin levels were then around 10g/dl). Over several weeks, her vision deteriorated further and then stabilized, without further progression. Initially she was able to read, but that became increasingly more difficult and she also had more difficulty recognizing faces. She denied any pain and had no systemic or neurologic complaints. She underwent a negative work up for anemia and her hemoglobin on its own improved to 11.7g/dl. Medical history was unremarkable. Review of systems was negative for jaw claudication, scalp tenderness, arthralgia, or myalgia. She was on no medications except for multivitamins.

Examination revealed an alert lady oriented x 3 with spectacle corrected visual acuity of 20/125 OD and 20/100 OS, improvable with pinhole to 20/100- OD. There was a 2+ left afferent pupillary defect and subjective redness and brightness desaturation on the left. Ishihara color vision was absent bilaterally. Eye movements were full with a suggestion of a small exotropia. Corneal sensation, orbital and slit lamp findings were unremarkable with mild nuclear sclerotic cataracts. Intraocular pressures were normal. Dilated funduscopy revealed normal discs, maculae and vessels in both eyes. Her neurologic examination revealed quite brisk reflexes with unsustained ankle clonus and an equivocal upgoing toe on the left.

Humphrey 24-2 threshold fields revealed superior paracentral defects in both eyes. There was no significant OCT evidence of retinal nerve fiber layer loss in either eye.

FINANCIAL DISCLOSURE: NONE
A Case of Net Visual Loss and Gain

Answer

FINAL DIAGNOSIS:
Paraneoplastic optic neuropathy secondary to non-secretory pancreatic (islet cell) neuroendocrine tumor.

SUMMARY OF CASE INCLUDING PATHOLOGY:
A workup revealed slight anemia; blood work was otherwise negative. An initial evaluation was negative for tumor or autoimmune abnormalities. Brain and orbital MRI scan were normal. An electoretinogram was normal, and visual evoked potentials revealed marked prolongation in P 100 latency in both eyes. Tests for Leber’s hereditary optic neuropathy and neuromyelitis optica were negative. To further evaluate the unexplained bilateral optic neuropathy, blood was sent for serological evaluation. Western blot analysis of an extract of pig optic nerve identified an antibody reactive with the optic nerve.

Further evaluation included repeat normal brain and orbit MRI and normal total spine MRI and CSF studies. Paraneoplastic markers, including CRMP-5 (CV2 by Athena Diagnostics) were all negative.

A PET scan showed multiple bilobar hepatic lesions with mild increased uptake. An MRI cholangiogram (MRCP) identified multiple rim-enhancing hepatic lesions, consistent with metastatic disease and a 2.5 x 2.4 cm² area of parenchymal prominence in the tail of the pancreas, suspicious for a mass. A liver metastasis biopsy revealed a low grade, well differentiated neuroendocrine neoplasm, with tumor cells positive for synaptophysin, chromogranin and CD56 while negative for hepar, CD34, and MCEA. Reticulin was negative supporting the diagnosis of a low-grade neuroendocrine tumor. Combining the radiographic and clinical findings, the patient was given a diagnosis of a pancreatic neuroendocrine tumor, non-functional (ie, non-hormone secreting) metastatic to the liver.

After right hepatic transarterial embolization the patient was treated with IVIg and subsequently with octreotide. Following treatment there was fairly prompt, gradual, sustained improvement in vision to the 20/30 level with return of ability to drive and use a computer. Visual field and VER improvement was noted; OCT showed development of some bitemporal RNFL loss.

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KEYWORDS: Paraneoplastic Optic Neuropathy, Paraneoplastic Syndromes, Complication of Cancer

REFERENCES:
A Bright Spot Causing Darkness
Thomas Hwang
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HISTORY & EXAM:
A 20 year-old Hispanic male presented to a comprehensive ophthalmologist with new headache worse lying down and blurry vision for 2 weeks. His vision was 20/20 bilaterally. The anterior segment exam was unremarkable. The fundus examination revealed bilateral optic disc edema. An MRI was interpreted as showing a thrombus in the vein of Galen and MRV confirmed an associated blockage.

An inpatient internist performed a spinal tap but did not record an opening pressure. He was treated with warfarin, and his headache and papilledema resolved over 2-3 months. Five months later, he returned to the same ophthalmologist complaining of vision loss worse on the left worsening over three weeks. He denied headache, transient visual obscurations, or tinnitus. His vision was 20/30 OD and 20/200 OS. His visual fields showed bilateral predominantly cecocentral vision loss.

Repeat MRI showed an enlargement in the lesion.

Spinal tap revealed an opening pressure of greater than 550 mm H$_2$O. He was referred for neuro-ophthalmology evaluation with the question of whether his vision loss stemmed from intracranial hypertension. His examination revealed symmetric slightly sluggish pupils with no afferent papillary defect.

The extraocular motility was normal, the ocular alignment was orthotropic, and the anterior segment exam was unremarkable. The fundus revealed normal macula, vessels, and periphery but moderately pale nerves bilaterally with minimal optic nerve swelling.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Metastatic melanoma causing venous thrombosis and paraneoplastic retinopathy.

SUMMARY OF CASE INCLUDING PATHOLOGY:
The MRI lesion was intrinsically bright on T1. This finding limited the differential to acute blood, fat, or melanin. Review of the MRI showed a smaller second lesion in the right frontal area making this a multi-focal process.

CSF cytology showed cells that contained melanin but were not otherwise identifiable. Dermatologic examination revealed a giant nevus on his back and flank.

Full field ERG showed normal photopic and scotopic responses, but multi-focal ERG showed a clear maculopathy that matched the visual field defect.

A craniotomy was performed which revealed black pigment on the meningeal surface. The pathology showed malignant melanoma based on HMB45, Melan A, and S-100 staining.

A retinal auto-antibody panel revealed a positive antibody against enolase consistent with a paraneoplastic retinopathy.

The patient was treated with temozolomide and external beam radiation. He unfortunately continued to worsen with rapid progression of leptomeningeal involvement as well new lesions in his spinal cord. He is currently enrolling in a ipilimumab clinical trial for immune therapy of metastatic melanoma.

Initial presentation with papilledema, intracranial hypertension, and response to warfarin was consistent with a cerebral venous sinus thrombosis; therefore the MRI lesion was mistakenly identified as a sinus thrombosis. In retrospect, extrinsic compression by melanoma caused venous stasis and likely secondary thrombosis, which explains the initial clinical improvement on anti-coagulation. Second presentation with intracranial hypertension and cecocentral visual loss but without papilledema raised suspicion for another process confirmed by multi-focal ERG, retinal auto-antibodies, and brain biopsy.

KEYWORDS: Paraneoplastic, Cerebral Venous Sinus, Binocular Vision Loss, Multi-Focal Electrotoretinogram, MRI/MRV

REFERENCES:
HISTORY & EXAM:
A 76-year old man presented in April of 2008 with ptosis and impaired eye movements. Five years earlier (2003) he had symptoms of muscle pain and arthralgia with increased ESR and was diagnosed with polymyalgia rheumatica. He had a negative temporal artery biopsy at that time. One year later he presented with persistent PMR symptoms, episodes of “episcleritis” that responded to topical steroids, abrupt left eye vision loss with a normal fundus appearance and an abduction deficit. Presumed posterior ischemic optic neuropathy was diagnosed and with a negative MRI scan, he was diagnosed with temporal arteritis without a biopsy. High dose steroids improved his eye movements although vision remained poor. Two years later he was treated with high dose steroids because of increased ESR, arthralgias with muscle pain. He was then treated with low dose prednisone for two years. Five months prior to this presentation he had a normal orbital and eye movement examination but maintained light perception vision in his left eye.

Currently, while hospitalized for a gastrointestinal illness, he noticed the abrupt onset of ptosis. He denied jaw claudication or worsening of myalgias and arthralgias. There was no eye pain or redness and no tenderness over the temporal arteries. Several days later he noticed a mild frontal headache and some numbness on his forehead. Medications included 10 mg of prednisone. His visual acuity was 20/20-2 in the right and light perception in the left eye. An APD, 3mm of exophthalmos and a severe omni directional limitation of eye movements were present in the left eye. There was also hypoesthesia over the cranial nerve V1 distribution on the left and lacrimal sac dacryocoeles. There were no abnormal findings in the right eye. The left optic nerve was pale. CT and MRI scan were performed.

FINANCIAL DISCLOSURE: Support from Research to Prevent Blindness
**What's in a Name**

**Answer**

**FINAL DIAGNOSIS:**
Necrotizing granulomatous inflammation of the orbit and sclera without systemic disease

**SUMMARY OF CASE INCLUDING PATHOLOGY:**
MRI demonstrated an intraconal mass which demonstrated heterogeneous enhancement and appeared to extend to the optic canal. A biopsy revealed an extensive necrotizing process with palisading histiocytes and multinucleate giant cells with an inflammatory infiltrate, including neutrophils, lymphocytes, and plasma cells. Vascular structures within the inflammatory field were involved. Special stains performed for bacteria, acid-fast bacilli and fungi were all negative.

The patient was placed on 50 mg prednisone, tapered and initially azathioprine was added. Work up for ANCA associated granulomatosis revealed negative P ANCA, C-ANCA, chest x-ray and urinalysis. He was treated successfully (improved motility and MRI) for two years with low dose steroids and Mycophenolate. Over one month he developed recurrent bouts of scleritis and given the poor vision, and pain as well as reluctance to use high dose steroids, an enucleation was performed. A necrotizing granulomatous inflammatory process was found, with negative cultures and no vasculitis.

ANCA-associated granulomatous vasculitis (AAGV-formerly Wegener's Granulomatosis) is an autoimmune disease characterized by necrotizing granulomatous inflammation commonly affecting upper and lower respiratory tracts, lungs, kidneys, eyes and orbit. Ocular manifestations occur in up to 50% of patients, with orbital inflammation being the most common presentation (1,2). To diagnose AAGV, a patient must have at least 2 of the 4 following criteria: 1) granulomatous inflammation; 2) abnormal CXR; 3) nasal/oral inflammation; and 4) abnormal urinary sediments (3). Diagnostic workup is useful when AAGV is clinically and/or pathologically suspected. Antineutrophil cytoplasmic antibody (ANCA) is the diagnostic that has been reported to have a high sensitivity up to 85-90% in widespread AAGV and a specificity of 99% (4). Our patient's findings placed AAGV high on our differential diagnosis although + ANCA never developed nor any systemic manifestations. As well the enucleation specimen did not show significant vasculitis further confounding the "name" of this disease.

The difficulty stemmed from the overlapping profiles of temporal arteritis and granulomatous orbital inflammation and the question as to why there were no other systemic manifestations, and a negative ANCA, as well as his resistance to treatment. We wonder whether this could have been an ANCA associated-like necrotizing granulomatosis over the 7 year presentation or were both diseases present? In addition, initial concerns were of an opportunistic infection when the orbital apex syndrome first presented.

**KEYWORDS:** Scleritis, Orbital Inflammation, Diplopia, Orbital Mass, Ischemic Optic Neuropathy

**REFERENCES:**

HISTORY & EXAM:
A 41 year-old african-american woman developed a shadow over her left eye in early May. She denied pain. Visual acuity was 20/20 OD and 20/25+2 OS. Central static perimetry showed moderate nasal, superior and inferior constriction in the left eye. There was 360-degrees of left optic nerve head elevation. Right eye examination was normal. A diagnosis of optic neuritis was felt most likely. MRI report was consistent with this.

By early June left eye vision had deteriorated to 20/400. The left optic nerve remained swollen. Neuro-ophthalmic and neurologic examinations were otherwise normal. She was admitted to the hospital for IV steroids and further workup.

She had no previous medical or ocular history. She was not taking any medications. She worked in an office at the county jail and did not have exposure to inmates. PPD had been negative. There was no history of toxin exposure. There was no family history of ophthalmic, neurologic or autoimmune disease.

MR imaging was reviewed. It showed perineural enhancement in both orbital apices, as well as multiple superficial, nodular, enhancing lesions of the brain and upper spinal cord (see figures). MR angiography of the brain, CTs of the chest, abdomen and pelvis, and gallium scan were normal.

Vitamin D was low. ESR was 34. Blood work was otherwise unremarkable including CBC, BMP, LFTs, TSH, ACE, C-ANCA, P-ACNA, anti DS-DNA and SSA/B. Serum infectious studies were negative including quantiFERON TB gold, Lyme, HIV, toxoplasma and cystercercosis

Spinal fluid analysis showed normal glucose, elevated protein (105 mg/dL), and a lymphocytic pleocytosis (89 wbc/µL, 85% lymphocytes, 9% monocytes, 6% neutrophils). There were no oligoclonal bands. JC virus was negative. AFB smear and culture were negative. Bacterial and fungal cultures were negative.

A procedure was performed. When that failed to reveal the diagnosis, a second procedure was performed.

FINANCIAL DISCLOSURE: NONE
‘Tis Nobler in the Mind to Suffer... Or to Take Arms Against a Sea of Troubles and by Opposing End Them?

Answer

FINAL DIAGNOSIS:
Neuro-sarcoidosis

SUMMARY OF CASE INCLUDING PATHOLOGY:
Biopsy of left orbital fat showed chronic inflammation and normal adipose tissue (see figures). Flow cytometry did not identify a monoclonal population.

A superficial cervical spinal cord lesion was biopsied. Microscopic examination showed coalesced granulomata with central caseous necrosis surrounded by chronic inflammation. Gram, GMS and acid fast stains did not reveal organisms (see figures). Acid fast culture was negative. Bacterial culture was negative. PCR for Mycobacterium TB was negative. There were no features typical for neoplasia. CSF angiotensin converting enzyme was elevated. Within 5 days of receiving IV steroids left eye vision improved to 20/30. Optic nerve edema resolved and MRI demonstrated decreasing size of the lesions (see figure), which are typical for granulomatous disease (1). In mid-July she developed a shadow in the right eye. Visual acuity was 20/20 with each eye and automated visual fields were normal. She was treated with steroids, then four drug antimycobacterial therapy without improvement in subjective symptoms. Antimycobacterial agents were stopped after 2 weeks due to side effects. Off all therapy, her clinical examination and imaging have remained stable.

Both tuberculosis and sarcoidosis are known as mimickers with diagnosis relying on a high degree of clinical suspicion coupled with, often elusive, confirmatory laboratory or pathological evidence. In this case the lack of demonstration of infectious organisms using multiple modalities (serology, csf and tissue culture, tissue stains and tissue PCR), elevated CSF ACE, which is reported to have greater than 90% specificity for sarcoidosis (2), response to steroids and lack of worsening off antimicrobial therapy led to a diagnosis of sarcoidosis. While necrotizing granulomata are considered classic for mycobacterial disease, they have been reported in sarcoidosis (3-5) and therefore do not dissuade us from this diagnosis.

T.B. or not T.B.? When pathology shows caseating granulomata, the burden of proof for a diagnosis other than tuberculosis is on the physician. In this case circumstantial evidence is plentiful. Exoneration requires proof of an alternative diagnosis. How does one prove a diagnosis of exclusion (i.e. sarcoidosis)? Dilemmas regarding empiric therapy are coupled to the diagnostic dilemma. When is antimycobacterial therapy indicated in a patient who tolerates it poorly? When is monotherapy with steroids appropriate?

KEYWORDS: Optic Neuropathy, Meningitis, Brain Lesions, Granulomatous Disease

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Egg in Your Face
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HISTORY & EXAM:
A previously healthy 8-year-old girl presented with 2 days of confusion, headache, and vomiting. Lumbar puncture was acellular with a normal glucose but a protein of 81 mg. No opening pressure was recorded. She was diagnosed with viral meningitis and not treated. She had lingering headache.

One month later she presented with renewed confusion, vomiting, ataxia, left upper extremity weakness and dysarthria. MRI showed high T2/FLAIR signal in the cerebrospinal fluid spaces with thickening of the tectal plate and superior vermis and mild ventriculomegaly. There was diffuse brain and spinal cord leptomeningeal thickening and enhancement (Fig.1A-D). Electroencephalography showed non-convulsive status epilepticus originating from the right frontotemporal region and requiring 3 antiepileptic drugs for control. Visual acuity was 20/70 OD, 20/50 OS with normal pupils and mild bilateral optic disc edema. Lumbar puncture showed an opening pressure of 36 cm H2O, 3 white cells (9 lymphocytes and 91 histiocytes), normal glucose, protein 72mg/dL and was negative for cryptococcal antigen, standard viral panel, and cytology. A right frontotemporal brain biopsy showed leptomeningeal fibrosis with macrophages and lymphocytes. She received a course of intravenous steroids.

Subsequent eye exam showed visual acuity of 20/25 OD, 20/40 OS, with a left APD and persistent mild bilateral disc edema. Two months later she presented with vomiting and lethargy. MRI showed increasing ventriculomegaly and pressure monitoring revealed pressures spiking to 54 cm H2O. She underwent right ventriculoperitoneal shunting. Lumbar punctures continued to show elevated protein (187, 1817) but remained negative for infection and neoplasm. Despite the shunt and another course of intravenous steroids, her mental status continued to decline. Leptomeningeal enhancement became more widespread.

A procedure was performed.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Primary Leptomeningeal Oligodendroglioma WHO II

SUMMARY OF CASE INCLUDING PATHOLOGY:
A right temporal lobe meningeal and parenchymal biopsy showed neoplastic oligodendroglial cells in the leptomeninges. The adjacent cerebral cortex was not involved except for a few cells in one or two regions. The cells were positive for synaptophysin, but negative for GFAP, vimentin neurofilament, chromogranin, CD3, CD45, and EMA. The MIB-1 proliferation index was 15-20%. The pathologic diagnosis was primary leptomeningeal oligodendroglioma WHO Grade II.

She underwent chemotherapy consisting of 6 cycles of weekly carboplatin. After two doses of chemotherapy she developed worsening headache, ataxia, and slurred speech. Imaging revealed tonsillar descent and early syrinx with a dilated fourth ventricle. The fourth ventricle was connected by a catheter to the shunt. Leptomeningeal signal abnormalities lessened on MRI three months later. She has lingering fatigue and rare seizures but no neurologic deficits except visual acuities of 20/30 OU and flat, mildly pale optic discs.

Primary leptomeningeal oligodendroglioma is a very rare and aggressive malignancy of the central nervous system that develops from the heterotopic nests of oligodendrocytes within the leptomeninges. Very few cases have been reported with most cases being diagnosed at autopsy.

Cauterization artifact in the first biopsy had apparently led to mistaking oligodendroglial cells for lymphocytes. The lack of a positive cytology (and high protein) led to the mistaken diagnosis of meningitis, but is also typical of this tumor.

The chronic mild bilateral optic disc edema with APD led to the mistaken impression of chronic optic neuritis, particularly given the working diagnosis of meningitis. The difficulty of assessing visual function in this young, ill, and therefore uncooperative patient further complicated the distinction between papilledema (the correct diagnosis) and optic neuritis.

Diagnosing a primary leptomeningeal malignancy is hard. It is rare. Lumbar punctures typically do not harvest the malignant cells. Biopsy may capture only the surrounding inflammatory response. In this setting, differentiating optic neuritis from papilledema is also hard. The diffuse meningeal thickening with enhancement, high protein, and periependymal high signal were clues of a primary leptomeningeal malignancy and suggested papilledema rather than optic neuritis. In such cases, early and multiple brain/meningeal biopsies may be critical.

KEYWORDS: Leptomeningeal Tumor, Oligodendroglioma, Papilledema, Seizures, Leptomeningeal Fibrosis

REFERENCES:
HISTORY & EXAM:
A 60 year old female artist reported difficulty reading, specifically in scanning newspapers and word comprehension. These problems had developed and gradually progressed over four weeks. Her prior ophthalmological history included 20 years of recurrent granulomatous iritis in both eyes which had been treated with some success with topical prednisolone and mydriatics. Her medical history was significant for chronic variable immune deficiency (CVID), diagnosed 23 years ago, with associated hepatitis and dermatitis. Her CVID was treated with IVIG and hydroxychloroquine. She took no other medications. On examination, visual acuity was 20/30 OU with normal color vision and pupillary responses. Rare cell with keratic precipitates were seen in both eyes and the intraocular pressures and posterior segments were normal. Visual fields showed a right homonymous hemianopsia. Neurological examination revealed no defects in speech, motor function, sensation, or gait, but the patient had difficulty writing and with word-finding.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Progressive multifocal leukoencephalopathy in the setting of CVID

SUMMARY OF CASE INCLUDING PATHOLOGY:
An MRI revealed lesions in the left posterior temporal and occipital regions which were minimally enhancing but bright on T2/FLAIR. DWI showed mild hyperintensity with variable ADC changes, indicating T2 shine through.

A lumbar puncture returned cerebrospinal fluid (CSF) with glucose of 43 mg/dl, normal protein, no red cells, and 1 white cell. Tests for VZV, Cryptococcus, syphilis, CMV, EBV, Enterovirus, and polyomavirus JC DNA were negative. IgH gene rearrangement PCR was also negative. After a few days in the hospital, the patient’s cognitive state declined and she became unresponsive. Imaging showed no acute infarcts and EEG suggested seizure activity. Given the lack of CSF findings, a brain biopsy was performed which showed foamy macrophages and gliosis. No vasculitis or granulomas were seen. Luxol fast blue stain revealed severe demyelination, but neurofilament stain showed intact neuronal processes. The SV40 stain was positive within individual cells. The patient was treated with anti-epileptic medication and mefloquine added to her regimen for CVID.

The patient’s cognitive difficulties and subsequent decline in the setting of an immunocompromised state raised the possibility of lymphoma, infection, granulomatous process, or PML as the etiology of her symptoms. The patient’s PCR for JC DNA was negative. However, the sensitivity of JC PCR can be as low as 58% and therefore a negative result should not exclude the diagnosis in a patient with suspicious symptoms. PML associated with CVID has rarely been reported.

KEYWORDS: progressive multifocal leukoencephalopathy, common variable immune deficiency, homonymous hemianopsia, demyelination

REFERENCES:
Bad Eyes, Bad Walking and Bad Judgement
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HISTORY \& EXAM:
A 20-year-old engineering student presents with deterioration of vision in his left eye, down to 6/18 initially diagnosed as optic neuropathy. On further review there was a pale disc, attenuated vessels and an abnormal ERG, thus he was diagnosed with a retinal dystrophy. One year later he presents with deterioration of vision in the other eye, and is now light perception in left and 6/12 in right. He is sent to a tertiary referral centre for further work-up. There is progressive pallor of the optic discs and vessel changes in both eyes.

He was diagnosed with combined variable immunodeficiency (CVID) at age 7, for which he has a limited IgM and IgA immunoglobulin preparation. His sister has the same condition. He has been previously treated for atypical mycobacterium following exposure to an infected family member, including ethambutol and isoniazid, all treatment had finished prior to onset of symptoms. He does not smoke and rarely drinks alcohol. He has used ecstasy, cocaine, ketamine and amphetamines but not in recent months.

An MRI and electro-diagnostics are performed. Between his review appointments, he has crashed his car (twice) and been refused entry to pubs for being “too drunk”. His mother reports a “difficult mood at home”. He is agitated, and clinic staff report that he seems intoxicated. On examination he has acuity of 6/18 N8 right and no light perception left. He scores 12/13 on Ishihara colour testing in the right eye. Intraocular pressures are normal. There are occasional anterior chamber cells bilaterally but the vitreous is quiet. The optic discs are pale with vessel attenuation and sheathing, left more affected than right, there is no retinal pigment epithelial clumping. Neurological examination reveals a cerebellar ataxia, dysarthria and choreiform movements. Tone and reflexes are increased with pyramidal signs. Power and sensation of the limbs are normal. A psychiatric examination concludes that he is manic.

FINANCIAL DISCLOSURE: Dr. Clare Fraser is supported by the Sydney Eye Hospital Alumni Travelling Fellowship.
Bad Eyes, Bad Walking and Bad Judgement

Answer

FINAL DIAGNOSIS:
Autoimmune retinopathy and encephalopathy

SUMMARY OF CASE INCLUDING PATHOLOGY:
This young man has a progressive asymmetrical retinopathy, with electrodiagnostic features including: an electronegative ERG, rod dysfunction, reduced cone function and VEP reduction. Fluorescein angiogram and optic coherence tomography are non-contributory. He subsequently develops cerebellar and psychiatric disturbances. He has a white cell count of $2.6 \times 10^3$, with CD4 191, CD8 216 and B-cells 11. The main differential diagnosis includes infectious, autoimmune or paraneoplastic pathologies.

Given his background of combined variable immunodeficiency a full infectious work-up is performed including lumbar puncture. In addition blood tests for autoimmune and paraneoplastic antibodies are requested. An MRI of the brain and spinal cord as well as a full body CT and PET scan are performed. The lumbar puncture shows a mildly reactive lymphocytosis but otherwise all tests are normal. Genetic testing for SCA and Lebers are normal.

Serial electrodiagnostic investigations show progressive deterioration of his ERG. This is matched with deterioration in mobility and speech, as well as an increase in choreiform movements. His mania is relatively well controlled with input from our psychiatric colleagues. A trial of IV methylprednisolone results in no subjective improvement, and his ERG recordings continued to deteriorate after treatment, before a period of stabilisation.

His sister who also has CVID developed an autoimmune thrombocytopenia purpura, which failed to respond to prednisolone, but did respond to rituximab. The immunology team was reluctant to try this treatment until viral encephalopathy had been completely excluded. A retinal biopsy of the blind eye is performed which shows mild ganglion cell and cone loss and no inflammatory infiltrate. A brain biopsy shows a perivascular infiltrate of CD3+ T cells and microglial nodules, concluding that immune mediated encephalitis is most likely.

He is commenced on IV gammaglobulin, and then rituximab.

The initial misdiagnosis was of optic neuropathy, and then unilateral retinal dystrophy. The difficulty with the case is how to distinguish between autoimmunity and an indolent viral infection in the setting of immuno-compromise. The next dilemma is how to treat autoimmunity is a patient who is already immuno-deficient.

KEYWORDS: Vision Loss, Electrophysiology, Autoimmune Diseases, Neurologic Disorder, Retinal Disorder

REFERENCES: NONE
HISTORY & EXAM: A 61-year-old Caucasian woman has tic douloureux and horizontal binocular diplopia.

Her past history includes squamous cell skin cancer removal below the right eye in 2003.

On February 12, 2008, local anesthetic for basal cell carcinoma resection anterior to her right tragus results in severe brief right facial shooting pain. By late February, her right upper lip has become numb and over the next two months, the numbness spreads to involve the right cheek, with pain and tingling of her right cheek and upper right teeth. Ophthalmology and dental examinations were normal. Over the next 7 months she is seen by 5 neurologists who note decreased sensation in the right V1 and V2 distributions and diagnose her with atypical trigeminal neuralgia, possibly related to multiple sclerosis. Four MRIs are normal with the exception of a few nonspecific white matter T-2 bright lesions. She is treated with trials of gabapentin, acyclovir, pregabalin, oxcarbazepine, baclofen, duloxetine, valproic acid, amitriptyline and acupuncture treatments without relief. Her pain increases, requiring narcotics.

In November 2008, she develops mild right facial weakness. MRI and MRA on November 20, 2008 are normal, but a loop of her right superior cerebellar artery contacts the trigeminal nerve at the root entry zone. In January 2009, she notes an enlarged right cervical lymph node. On January 28, 2009 she undergoes microvascular decompression with placement of pads around the right cranial nerve V and along right VII and VIII. Her right facial weakness is more pronounced after surgery. Her cervical lymph node painlessly enlarges. On February 17, 2009, she develops horizontal binocular diplopia, worse with gaze right.

Neuro-ophthalmology examination on March 11, 2009 shows a 3 cm nontender right cervical/ submandibular lymph node, mild right facial palsy, 50% right abduction, and decreased facial sensation in the right V1 and V2 distribution.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Perineural spread of squamous cell carcinoma of the face AND Hodgkin’s lymphoma.

SUMMARY OF CASE INCLUDING PATHOLOGY:
MRI on March 11, 2009 shows enhancement of the right trigeminal nerve in Meckel’s cave, the foramen ovale, the foramen rotundum and the infraorbital foramen, with likely involvement of the cavernous sinus.

Review of MRI from November 20, 2008 shows thickening of the right trigeminal nerve.

March 20, 2009: biopsy of right cervical lymph node shows architectural distortion due to the large numbers of small non-caseating granulomas; scattered throughout were enlarged atypical cells with large ovoid nuclei, often multiple, with enlarged nucleoli, typical of Reed Sternberg cells, immunoreactive for CD30 and CD20 and negative for CD15. Pathology was felt to be classic for Hodgkin lymphoma.

Work-up/staging with whole body PET, bone marrow biopsy, and CSF analysis X 2 unremarkable, except for hypermetabolic activity in the right neck and upper center chest lymph nodes and spleen.

Treated with ABVD (Adriamycin (doxorubicin), bleomycin, vinblastine and dacarbazine) chemotherapy beginning April 29, 2009 with no improvement in her facial pain or abduction deficit.

June 8, 2009: her right facial palsy requires a tarsorrhaphy. Repeat PET on June 22, 2009 shows resolution of the metabolic hyperactivity. By September 2009, her right facial pain has increased.

October 5, 2009: followup neuro-ophthalmology examination with complete right abduction deficit and decreased sensation in right V1, V2, V3.

October 12, 2009 MRI unchanged.

December 11, 2009: infraorbital nerve biopsy with grossly abnormal infraorbital nerve and pathology diagnostic of neurotrophic spread of moderately differentiated squamous cell carcinoma.

Treated with radiation therapy with poor response.

The diagnosis of trigeminal neuralgia should never be made when there is loss of facial sensation, and clinicians should always be wary of “normal” neuroimaging. This case breaks the “law of medical parsimony” (Occam’s Razor) by having two unrelated cancers simultaneously. The MRI appearance was highly suggestive of perineural spread of squamous cell carcinoma all along, and Hodgkin lymphoma only exceptionally involves these cranial nerves. The first diagnosis delayed further biopsy and correct diagnosis.

KEYWORDS: Trigeminal Neuralgia, Perineural Spread, Squamous Cell Carcinoma, Hodgin Disease, Sixth Nerve Palsy

REFERENCES:
HISTORY & EXAM:
A 13 year-old, left-handed female student first noted a change in vision that occurred while she was playing clarinet one week prior to presentation to our Neuro-Ophthalmology service. The change in vision was perceived as a “whiteness” over the right half of a sheet of music, similar to "lying on a beach and looking toward the sun with your eyes closed". An ophthalmological examination that day was “normal”. Her vision improved that day, though she could only see normally out of the upper region of her right eye. She denied headaches or any systemic symptoms. An MRI, MRA, carotid ultrasound, echocardiogram, bubble study, hypercoagulability work up and visual evoked potentials (all performed at Children’s Hospital) were “normal”. Her past medical history was significant for a spontaneously closed ventricular septal defect, an atrial septal defect that was surgically repaired at age 16 months, and a mitral valve repair at age 3 years. She has a history of seizure-like activity that began 3 years ago; sleep-deprived EEG showed abnormal activity, with "seizure-like spikes".

Her Neuro-Ophthalmological examination revealed best corrected visual acuity of 20/10 OU without dyschromatopsia or pupillary abnormality. Goldmann visual field testing showed complete loss of field inferiorly below approximately 10 degrees from fixation (Fig. 1). Her visual field was full OS. There was subtle swelling of the nerve fiber layer superior to the macula OD in an area consistent with her inferior visual field defect; no emboli were identified. Based upon our concern about a possible embolic source from the heart, the medical team obtained an echocardiogram that was normal. Her medical team then agreed to start on Coumadin. Two months later, she experienced another episode of visual field loss in the right eye (INR: 1.7), and a Goldmann visual field showed further visual field loss (Fig. 2). Funduscopy showed subtle retinal swelling superiorly within posterior pole; no emboli were identified. CT/CTA (upper chest to head) and repeat echocardiogram were normal. Color Doppler imaging of the retrobulbar segment of the central retinal artery was normal. She was found to be Factor V Leiden positive. Her Coumadin dose was increased and a daily aspirin was added.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Recurrent (presumably) fibrinous retinal emboli OD, secondary to endocarditis from infected retained foreign material from her prior cardiac surgery.

SUMMARY OF CASE INCLUDING PATHOLOGY:
Neuro-Ophthalmology strongly suspected cardiac embolization. Multiple meetings of the medical staff and cardiovascular surgeon led to a scrutiny of her cardiac status that did not uncover any clear abnormality. Thus, a decision was made to observe clinically. Continued input from Neuro-Ophthalmology regarding our interpretation of the visual events (i.e. they were embolic events, almost certainly from the heart) led to a consensus to recommend open heart surgery.
Cardiac exploration revealed: “thrombus versus vegetation”, with acellular fibromyxoid tissue, on the mitral valve; surface fibrin thrombus on the posterior leaflet of the mitral valve; retained foreign body material, including calcified cotton Gossypium hirsutum pledgets, at the base of the mitral valve. This foreign material was positive for Gram-positive cocci in pairs and in chains. Cultures (pledgets and blood) were negative cultures. Rare inflammatory cells were found in these abnormal regions. She was given prolonged course of intravenous antibiotics for endocarditis. She has not had any other embolic events or any new cardiac complications over the past 2 years of her follow-up.

In light of the repeated unrevealing echocardiogram and essentially negative work up, it was difficult to convince the medical team that her visual loss was likely the result of an embolus. The heart was an obvious source for emboli, but her stable clinical course and negative non-invasive cardiac studies were factors that led the cardiac surgeon and medical team to discourage cardiac surgery but to recommend Coumadin and aspirin therapy. However, the team was readily available for multi-disciplinary discussions, which ultimately led them to concur with the need for open heart surgery.

KEYWORDS: Transient Monocular Vision Loss, Cardiac Emboli

REFERENCES: NONE
Non-Functional Sinus Disease

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HISTORY & EXAM:
A 6 year old girl presented to our clinic with a one week history of vision loss, worse in the right eye. She had a two month history of progressively worsening headaches. About a month prior she had a CT scan which reportedly showed sinusitis. She was treated with a ten day course of amoxicillin without relief. Within the past month she had intermittent vomiting and fever. Her grandmother noticed she was having new difficulty reading, and had her see an optometrist. He found vision loss in both eyes, and wondered about a non-physiologic disorder, but referred her urgently for further evaluation. Her past medical history was not significant. She took loratidine 5 mg daily and acetaminophen for the headaches. She lived at home with both parents and attended kindergarten. Review of systems was positive for fatigue, skin rash, temple pain, allergy symptoms, and disruptive snoring. On examination, the right eye had no light perception vision, and the left eye was 20/100. She had a 2.7 log unit right relative afferent pupillary defect. Color vision was absent in the right eye, and she saw 4/13 color plates in the left eye. Motility was full bilaterally, and she had no proptosis, ptosis, or anisocoria. She had a 25 prism diopter right exotropia. Dilated fundus exam revealed mild pallor of the right optic nerve. Goldmann perimetry showed generalized depression and a central scotoma in the left eye (unable to perform on right eye because of no light perception vision). MR scans revealed sinus abnormalities. A diagnostic procedure was performed.

FINANCIAL DISCLOSURE: NONE
Non-Functional Sinus Disease
Answer

FINAL DIAGNOSIS:
Langerhan’s Cell Histiocytosis of the skull base, extending into both orbital apices, causing compressive optic neuropathies, right greater than left.

SUMMARY OF CASE INCLUDING PATHOLOGY:
The differential diagnosis initially included compressive optic neuropathy or optic neuritis associated with a recent viral infection. Upon close inspection, MRI brain and orbits showed a moderately enhancing heterogenous mass centered within the sphenoid sinus, with extension into the ethmoids and anterior cranial fossa with some destruction of the cribiform plates, and posteriorly into the pituitary fossa. It also extended to the apices of both orbits, with extraconal extension medially displacing both medial rectus muscles. The mass lesion looked most like a parameningeal rhabdomyosarcoma, but the radiologic differential diagnosis included metastatic neuroblastoma, an undifferentiated sinus tumor, lymphoma, Ewing’s sarcoma, or Langerhan’s cell histiocytosis.

Biopsy was performed which showed a mixed inflammatory infiltrate, including numerous eosinophils and histiocytes with eosinophilic to clear cytoplasm, and grooved nuclei that stained positive for S100 and CD1a.

She was diagnosed with Langerhan’s cell histiocytosis (LCH) and underwent resection of the tumor and chemotherapy. Her vision has improved to 4/200 right eye and 20/20 left eye, with residual visual field constriction on the right only. She has been free of recurrence for two years.

One month before presentation the patient was diagnosed and treated for sinusitis based on a CT. Weeks later she saw an optometrist who suspected functional vision loss. Imaging revealed a mass, considered most likely rhabdomyosarcoma. Radiologic features uncommon for LCH included its solitary nature, extension into the medial orbital walls (rather than lateral), and its extraskeletal soft tissue location. The patient did not display periorbital edema or proptosis, the most common clinical signs of LCH.

KEYWORDS: Langerhans Cell Histiocytosis, Orbital Neoplasm, Optic Neuropathy, Sinus Disease

REFERENCES:
HISTORY & EXAM:
In October 2009, a previously healthy 57-year-old woman developed fatigue, confusion and headache, associated with a right inferior quadrantanopsia. Brain MRI revealed multiple enhancing brain lesions with minimal mass effect, including left frontal and left parieto-occipital lesions. Initial diagnostic considerations included acute multi-focal demyelination, sarcoid, and malignancy.

The left parieto-occipital lesion was biopsied, revealing mononuclear infiltrates with numerous T cells (CD3+), histiocytes (CD68+), and a few scattered B cells (CD20+). There was no cytological evidence of malignancy, and axonal preservation was evident, consistent with demyelination. TAF, GMS, AFB and FITE stains did not demonstrate any microorganisms. In addition, HSV 1 and 2 immunohistochemistry, EBV in-situ hybridization and HIV serologies were negative.

Histopathology (October 2009): H&E 60x showing reactive perivascular astrocytosis and macrophages (left), CD68 60x immunostain for histiocytes (center), NF 180x showing relative preservation of axons (right)

A presumptive diagnosis of demyelinating disease (acute disseminated encephalomyelitis versus tumefactive multiple sclerosis) was made, and the patient was treated with a course of intravenous corticosteroids. Though her right inferior quadrantanopsia persisted, she clinically improved and was discharged, only to be re-admitted weeks later with increasing right-sided weakness and dysarthria. Repeat brain MRI showed decreased enhancement of her left frontal lesion but a marked increase in the size of the left parieto-occipital lesion with vasogenic edema and transtentorial herniation.

The patient refused both a lumbar puncture and a second brain biopsy so close to the first. With additional intravenous corticosteroid treatments and the addition of rituximab infusions, the patient’s clinical course once again improved, remaining stable for the next 5 months, with an improvement in the brain MRI appearance.

In April 2010, the patient was again hospitalized with worsening symptoms of right-sided hemiparesis and dysarthria.

FINANCIAL DISCLOSURE: NONE
FINAL DIAGNOSIS:
Primary central nervous system lymphoma preceded by “sentinel” demyelination.

SUMMARY OF CASE INCLUDING PATHOLOGY:
Despite the initial brain biopsy results consistent with demyelination, it was felt that the MRI appearance of the brain lesions (their density and nodular pattern of enhancement) strongly suggested the possibility of lymphoma. PET-CT scan showed increased metabolism within the brain lesions, with no evidence of extracranial malignancy.

Concerned that the initial brain biopsy had been misleading, we sought and obtained permission for a second brain biopsy from the patient’s family.

This repeat brain biopsy of the left parietal lesion, six months after the first biopsy, produced markedly different histopathologic results – large, perivascular malignant lymphoid cells uniformly positive for CD20 with a smaller number of CD3+ mature T cells – consistent with diffuse large B-cell lymphoma.

Histopathology (May 2010): H&E 180x showing large malignant perivascular lymphoid cells (left), CD20 180x pan B-cell immunostain (right).

Despite treatment with high-dose methotrexate and leucovorin, she continued to clinically deteriorate, was transferred to hospice care and, several days later, expired.

Primary central nervous system lymphomas (PCNSL) are rare neoplasms that account for less than 1% of all brain tumors. Though more common in the setting of immunosuppression, the incidence of PCNSL has increased in the last three decades among immunocompetent individuals.

It has been reported that some patients with PCNSL may initially present with steroid-responsive, demyelinating “sentinel” lesions characterized by a predominance of T cell infiltrates and few B cells. These demyelinating brain lesions may be histologically indistinguishable from those seen in multiple sclerosis (MS). Because both MS and PCNSL may present with contrast-enhancing white matter lesions and relapsing and remitting symptoms that improve with steroid therapy, this can lead to diagnostic confusion.

In this case, the patient’s clinical course and MRI appearance of her brain lesions (suggestive of lymphoma) was at odds with the results of the initial brain biopsy (consistent with demyelination). An awareness of the existence of “sentinel” demyelination preceding primary CNS lymphoma may allow clinicians to consider repeating a brain biopsy earlier in the clinical course, possibly resulting in an improved outcome.

KEYWORDS: Demyelination, Central Nervous System Demyelination

REFERENCES:
A Runny Nose
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HISTORY & EXAM:
This 78 year old gentleman was referred from the emergency room with a 3-4 week history of his left eye drooping and progressive decreased vision. His previous ophthalmic history was remarkable only for early cataract formation and refractive error. He had been checked four months earlier and was found to be 20/40 and 20/20. On examination, visual acuity was 20/25 OD and hand motion OS. Visual fields demonstrated a central scotoma and inferior altitudinal defect OS. Palpebral fissures were 8 and 11 with an upper lid range of 16 and 4 and severe ptosis. Hertels with a base of 122 were 21 and 23. Pupils were 4 mm OU with a > 1.8 log unit left afferent pupillary defect. Motility was fully intact OD but there was complete ophthalmoplegia OS. Slit lamp examination revealed 1+ nuclear sclerosis. Applanation tensions were 13 and 10. Funduscopic examination showed disc elevation OS with mild hyperemia.

Four months earlier, the patient had presented to his physician complaining of a runny nose on the left side. He was seen by an otolaryngologist and three months prior to our evaluation he had undergone resection of turbinate and cauterization of the nose. Patient denied any recent weight loss, change in appetite, fevers, chills, sweats, or pain on chewing. A CT and an MRI scan were obtained.

FINANCIAL DISCLOSURE: NONE