



NANOS

Patient

Brochure

Optic Nerve Sheath Meningioma

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Optic Nerve Sheath Meningioma

General description:

Optic nerve sheath meningioma is a benign (not malignant) tumor which involves the covering of the optic nerve. Meningiomas (along with gliomas and pituitary tumor) are the most common tumors inside the skull. These are slow growing, leading to problems when they interfere with normal function. Because the covering of the optic nerve is actually an extension of the dura, (covering of the brain) occasionally (approximately 1%) meningiomas may involve and surround the optic nerve. These have been reported in all age groups, but are most common in women in their thirties.

Anatomy: The optic nerve is responsible for sending information from the eye back to the brain where seeing actually takes place. In order for seeing to take place, the optic nerve needs to be able to function normally.

Physiology: When the optic nerve is squeezed, the blood supply and ability for the chemical messages and signals to travel through the nerve may be impaired. This may cause vision loss. When the tumor affects the covering of the optic nerve, it can actually mechanically squeeze the nerve or it may interfere with the blood supplied normally from small vessels from the optic nerve sheath. Either way, this will cause gradual loss of optic nerve function usually presenting with progressive visual loss.

Symptoms: Often, patients are unaware there is a problem until their eye doctor tests their vision. Sometimes patients may become aware that things look darker, dimmer, or more blurred out of one eye than the other. Because these tumors grow very slowly, in most cases, change occurs over a long period of time, usually months and sometimes even years, although occasionally, patients may become suddenly aware of a visual problem, particularly if they get something in the opposite eye. Less commonly the growing mass within the orbit can produce other symptoms including a slight bulge of the eye and limitation in eye movements often causing problems with double vision. In rare situations, vision may worsen when the patient tries to look to the side.

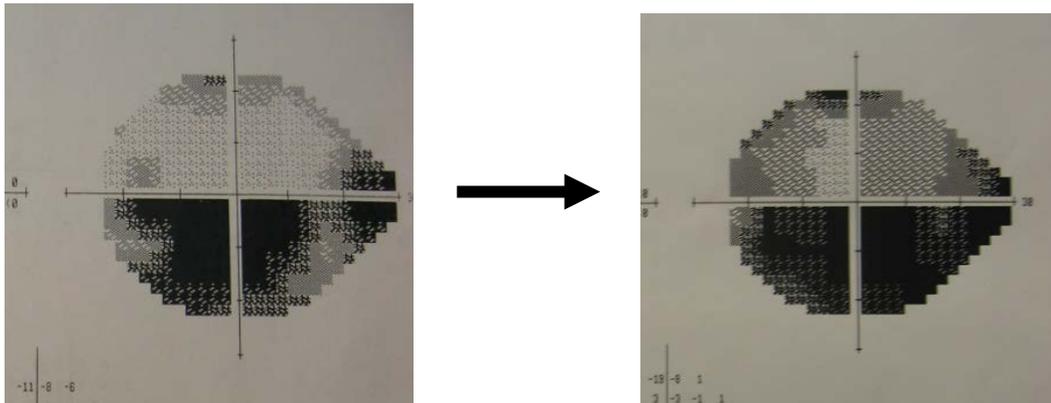


Figure A. Progression of the loss of side vision (black areas) on the visual field of the left eye over 3 years.

Signs: Other than mild bulge of the globe the most common signs relate to how the eye is working (vision and visual fields) and the appearance of the back of the eye. Vision is measured by testing on a vision chart. This is usually reduced although in some cases it may be only mildly affected. The doctor may notice that there is a less amount of light coming into the eye that is affected by swinging a bright light from one eye to another (afferent pupillary defect). The back of the eye often shows changes including swelling of the beginning of the nerve (optic disc) or

relative thinning of the optic nerve (disc pallor). On some occasions one can also see abnormal blood vessels on the disc surface (optociliary shunt vessels).

Diagnosis: The diagnosis is suspected by the symptoms and signs as listed above but confirmed by imaging studies, particularly an MRI scan or a CT scan (which often shows calcification). Although other lesions may affect the optic nerve sheath, these are much less common and usually are distinguishable from classical optic nerve sheath meningiomas. In very rare cases, a biopsy may be required to rule out other conditions.

Prognosis: As we like to tell patients, the good news about meningiomas is they grow SLOWLY. The bad news about meningiomas is they GROW slowly. Most meningiomas, if untreated, will progress, usually over a period of years to even decades.

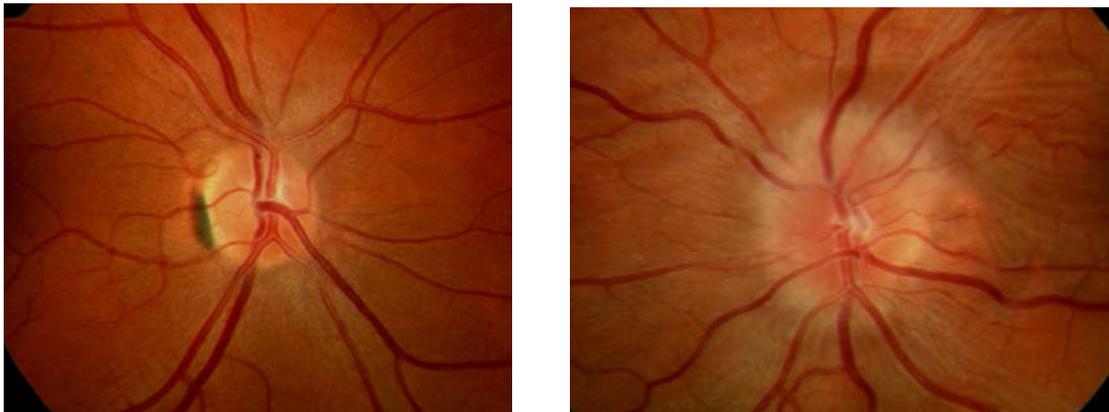
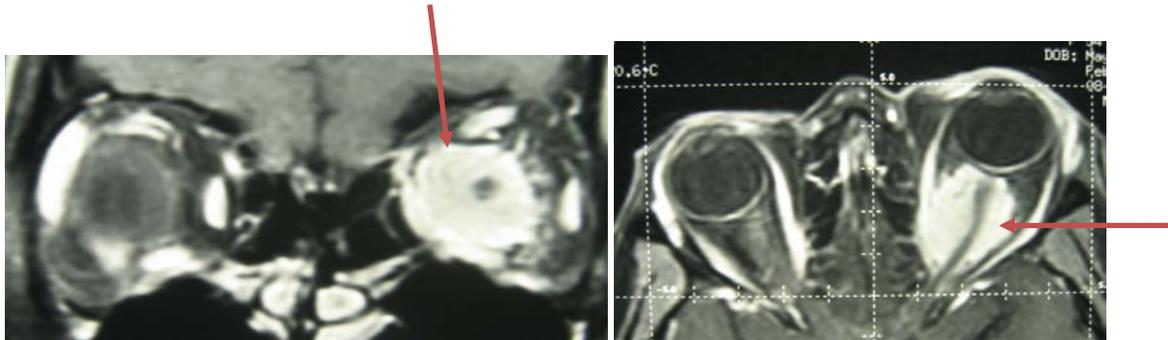


Figure B shows a normal right optic nerve on the left and a swollen left optic nerve with blurred edges on the right.



Figures C, D show the optic nerve on the left eye surrounded by the meningioma on MRI.

Treatment: Meningiomas that compress the optic nerve from one side (usually seen when they arise from the bone around the optic canal) can theoretically be surgically removed. Optic nerve sheath meningiomas that surround the optic nerve (most common) usually cannot be removed without worsening optic nerve function (often with total loss of vision in one eye). Although never studied in a controlled fashion, there is good evidence that radiation therapy can not only slow the growth of meningiomas, but actually can improve vision. In most cases, even when the vision is still fairly good, radiation therapy given in small doses over usually 20 days is usually recommended. Not all patients undergoing fractionated radiation therapy get better and some may continue to progress in spite of treatment. Other forms of radiation such as Gamma Knife or

Cyber Knife that is unfractionated (single dose) is likely to actually worsen the vision and will not have the same effect as fractionated radiation therapy.

Follow up: It will be important to follow the status of optic nerve function with repeat visual acuity measures, visual field measurements, and anatomic measurements with OCT. Repeat imaging is usually less necessary unless something else is going on.

Close follow-up may be provided by the patient's ophthalmologist to monitor visual acuity and visual fields. Also, the patient may also obtain evaluations from radiation oncology and/or neurosurgery, depending on the extent of the growth and the chosen therapies.

Frequently Asked Questions

1. *Will this meningioma affect the other eye?*

In retrospective series meningiomas that involve the optic nerve sheath rarely if ever extend to involve the opposite side. It is possible that optic nerve sheath meningiomas can involve the bone where the two optic canals enter the skull. These are, however, unusual. In uncommon cases patients may have bilateral optic nerve sheath meningiomas.

2. *Can this tumor cause other problems?*

Unlike other types of cranial tumors, optic nerve sheath meningiomas seldom, if ever, extend to cause other problems. Meningiomas that affect the base of the skull intracranially can cause problems with eye movements and result in double vision but this is unlikely to be associated with an optic nerve sheath meningioma.

3. *Are there other treatments available?*

While surgery has been attempted in patients with optic nerve sheath meningiomas, the majority of these will lead to worsening of vision. In the past, surgery was reserved for patients that had lost complete optic nerve function and vision in that eye. Attempts at medical therapy have been employed in the past although at this stage we have no proven medical treatments.