

Meningiomas

Conley B. Call MD, Keith Carter MD, Richard Allen MD, PhD

August 30, 2011

Chief Complaint: “Bulging left eye”

History of Present Illness: A 36 year old female presented to the Oculoplastics Service stating that her left eye was “bulging” and appeared swollen. She first noticed this approximately one month prior to her visit and felt that her symptoms were gradually worsening. She had experienced occasional headaches but denied any vision changes, eye pain or diplopia.

Past Ocular History: Unremarkable

Past Medical/Surgical History: Tonsillectomy

Social History: Unremarkable

Family History: Undefined cancer in maternal grandmother and paternal grandfather

Allergies: No Known Drug Allergies

Review of Systems: Unremarkable

OCULAR EXAMINATION:

Uncorrected Visual Acuity:

- Right eye (OD) 20/20-2
- Left eye (OS) 20/20-3

Confrontation visual fields: Full OU

Pupils: <0.3 log unit afferent pupillary defect OS

Ocular motility and alignment: Full OU. Orthophoric.

Intraocular pressure (IOP): 17 mmHg OD, 21mmHg OS

Exophthalmometry: 16mm OD, 22mm OS (base 93mm). See Figure 1.

Slit lamp exam: Anterior segments normal OU

Dilated fundus examination: See Figure 2.



Figure 1. Left eye proptosis

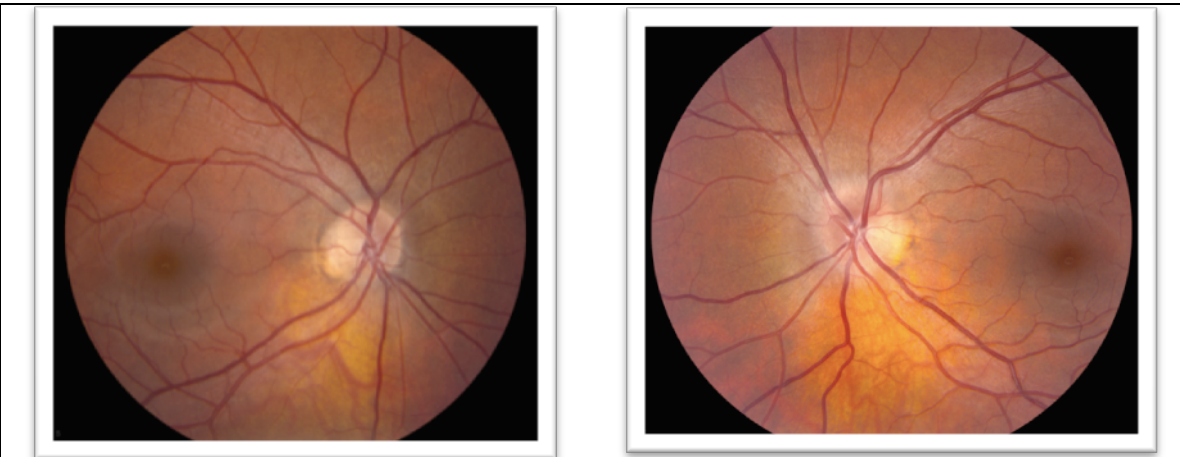


Figure 2. Optic nerve photos. Note the disc edema on the left.

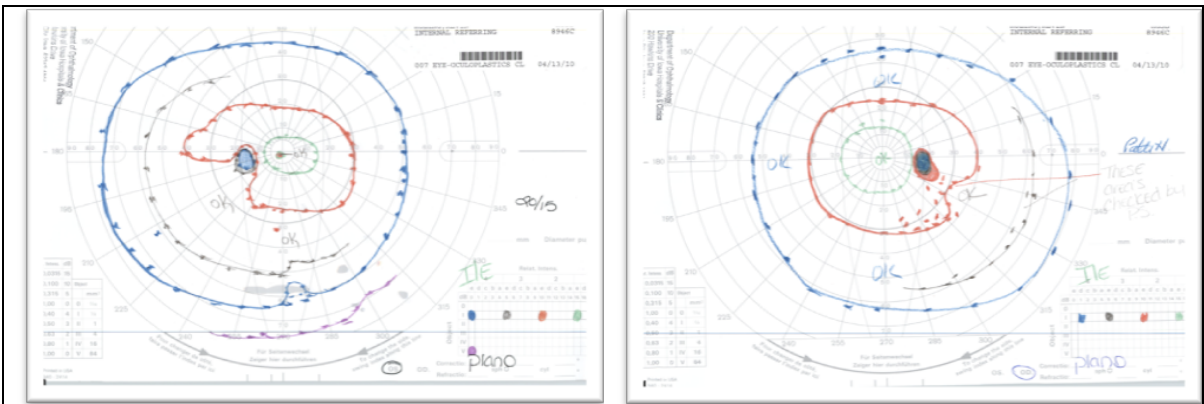


Figure 3. Goldmann Visual Fields (GVF). Note the enlarged blind spot OS>OD



Figure 4. CT scan (axial and coronal views). Note the hyperostosis of the left sphenoid wing

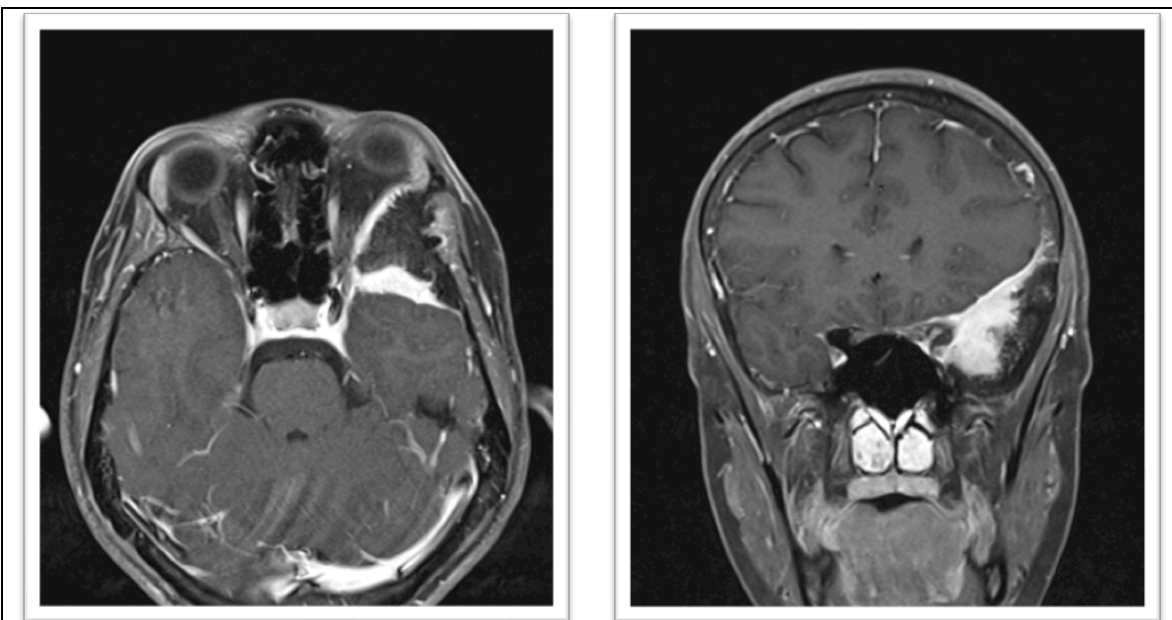


Figure 5. MRI (T1 sequence). Note again the prominent involvement of the left sphenoid wing.

CLINICAL COURSE

The patient’s findings were consistent with a left **sphenoid wing meningioma**. She underwent a left pterional craniotomy with sphenoid wing meningioma resection and decompression of the left optic nerve. Histopathological analysis confirmed a chordoid meningioma (WHO grade II) with subtotal resection.

On her one-week post operative exam, the patient complained of a left upper lid droop and the sensation that “her blind spot” was “a little more noticeable.”

One-Week Post-Op Exam		
	Right	Left
Visual acuity	20/15	20/50
Motility	Full	-0.5 abduction deficit
Exophthalmometry (base 102)	15	20

- + Left upper lid ptosis
- + 0.3 log RAPD OS
- + left cranial nerve V1 hypesthesia

The patient’s post-operative findings were noted and she was given additional time for further recovery. On her one-month post operative exam she felt that her vision had remained stable since the surgery, but continued to be bothered by “grey spots” in her vision inferotemporally.

One-Month Post-Op Exam		
	Right	Left
Visual acuity	20/15	20/50+2
Motility	Full	Full
Exophthalmometry (base 91)	15	19

Left upper lid ptosis, RAPD, and V1 hypesthesia remained stable.

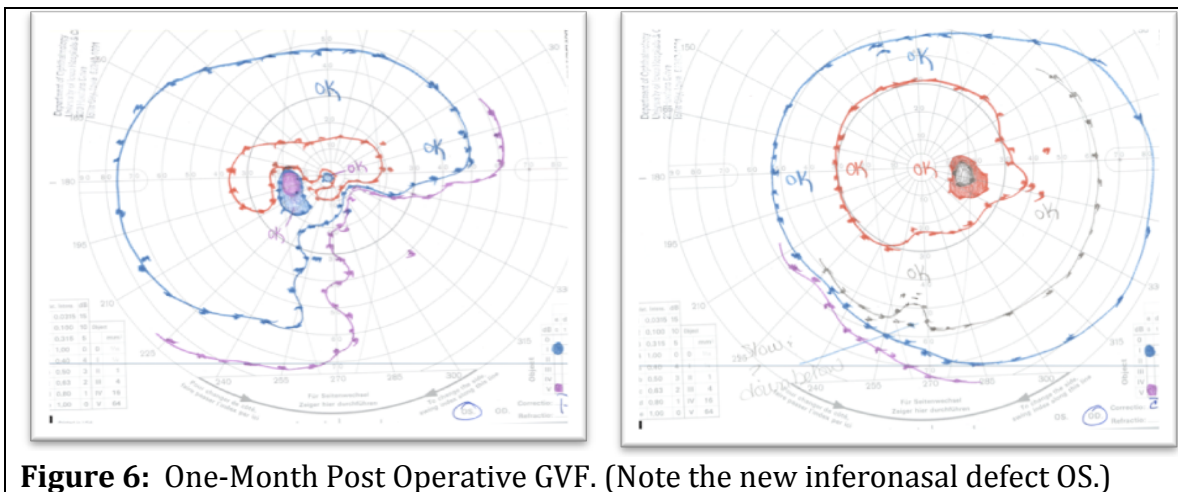


Figure 6: One-Month Post Operative GVF. (Note the new inferonasal defect OS.)

In summary, the patient was stable one-month after a left craniotomy for removal of the sphenoid wing meningioma with optic nerve decompression. She developed left upper lid ptosis, cranial nerve V₁ hypesthesia and inferonasal visual field defect. The prognosis for improvement of these new post-operative findings was uncertain. Radiation therapy is planned for treatment of the residual tumor.

DIFFERENTIAL DIAGNOSIS: Unilateral Proptosis

- Orbital Tumor
 - Primary (i.e. cavernous hemangioma, optic nerve sheath meningioma)
 - Secondary (i.e. intracranial meningioma, sinus tumors)
 - Metastatic (i.e. breast, lung, prostate)
- Vascular (i.e. carotid cavernous fistula)
- Infectious (i.e. cellulitis)
- Inflammatory (i.e. thyroid eye disease, sarcoidosis, myositis)

DISCUSSION:

Meningiomas are tumors arising from the menigothelial cells of the arachnoid layer of the meninges. They represent approximately 4% of all intraorbital tumors and 20% of all intracranial tumors. Orbital lesions can be primary lesions (i.e. optic nerve) or secondary lesions (i.e. extension from adjacent structures).

Classification of meningiomas is based on World Health Organization (WHO) Guidelines*. For most nervous system tumors the WHO grade is really an estimate of malignancy. Grading for meningiomas reflects the likelihood of recurrence and aggressive behavior. Classification currently includes 15 named subtypes:

- Menigothelial
- Fibrous (fibroblastic)
- Transitional (mixed)
- Psammomatous
- Angiomatous
- Microcystic
- Secretory
- Lymphoplasmacyte-rich
- Metaplastic
- Clear cell
- Chordoid
- Atypical
- Papillary
- Rhabdoid
- Anaplastic meningioma

* The WHO Classification of Tumors of the Nervous System. J Neuropathol Exp Neurol, Vol 61, March 2002.

Primary Orbital Meningiomas

Primary orbital meningiomas (i.e. optic nerve sheath meningiomas) are derived from meningotheelial cells found in the orbit. They account for approximately 2% of orbital lesions, and approximately 5% are bilateral. Common presenting signs/symptoms include painless, progressive visual loss and the classic triad of:

- Optic atrophy
- Visual loss
- Retinociliary collateral vessels.

These lesions tend to grow circumferentially around the optic nerve (see Figure 7) and are difficult to resect without nerve compromise. Histologically they tend to be of the meningotheelial and transitional pathological subtypes. The prognosis for survival is good with essentially no mortality. However, the prognosis for vision is guarded with many cases leading to progressive visual loss.

Treatment

Treatment options vary with close follow-up and periodic imaging often advocated for 20/40 vision or better. Surgical options are limited and biopsies are becoming more uncommon in part because of associated morbidity. Optic nerve sheath fenestrations to decompress the nerve are of historical interest only and can lead to massive orbital invasion of tumor. En bloc resection may be considered in cases of poor vision. Finally, radiation treatment is considered when there is evidence of progressive optic nerve compromise (visual acuity, visual field) or definite tumor enlargement in non-diabetic patient. It is delivered via 3-dimensional stereotactic techniques in order to minimize side effects.

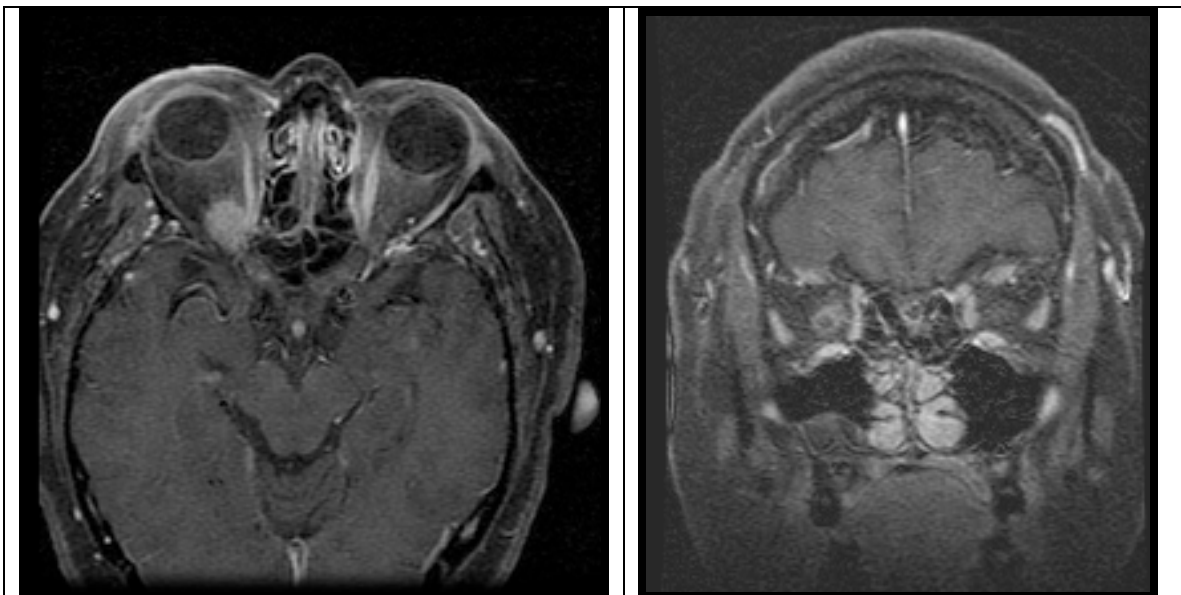


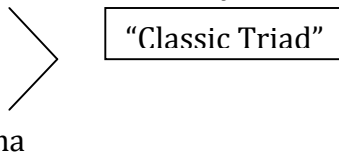
Figure 7: Optic nerve sheath meningioma. Axial (a) and coronal cuts (b). Note the circumferential growth (white arrow) around the right optic nerve visualized on the coronal cut.

Orbital Meningioma – Secondary lesions

Secondary orbital lesions constitute approximately 2% of all orbital lesions and extend into the orbit from various locations. They may spread from the following locations:

- Sphenoid wing (as illustrated in the case above)
- Clinoid
- Planum sphenoidale
- Frontoparietal area
- Olfactory groove

Presenting signs/symptoms of secondary lesions include the following:

- Proptosis
 - Vision loss
 - Ocular paresis
 - Optic nerve edema
- 
- “Classic Triad”

Treatment

Treatment options vary by individual case and include surgery, radiation, and close observation. Surgery is considered for any patient with a symptomatic or growing tumor. The goal is either complete resection or to stabilize the tumor or symptoms. Surgery employs combined neurosurgical and oculoplastic participation and often includes decompression of the optic canal. Nevertheless, these tumors are difficult to manage surgically because of their close proximity to vital structures such as the optic nerve and the internal carotid artery. Diffuse involvement of the skull base often increases surgical complexity. Debate continues as to whether surgery leads to better results than the natural history of the disease, and reported surgical outcomes vary. Visual function testing may demonstrate results ranging from mild improvement to continued deterioration. Exophthalmometry readings also vary, but the majority of patients show stable to improved measurements. Surgical follow up includes monitoring with yearly imaging studies.

As suggested above, various factors are reviewed when contemplating radiation treatment. These include the extent of surgical resection, tumor grade and histological subtype. Radiation is usually reserved for tumors that are rapidly progressive despite surgery or are histologically atypical. Shrivastava et. al. do not recommend post-op radiation for all patients with residual tumor but do advocate repeat surgery for recurrent tumors followed by post-operative radiation. The patient in our case received post-operative radiation based on the tumor grade and histological subtype.

Summary

<p>Epidemiology</p> <ul style="list-style-type: none"> • Mean age at presentation: 45 years old • Female (59%) > Male (41%) • Association with NF-2 • 4% of intraorbital and 20% of intracranial tumors • Can be primary or secondary tumors • Histopathologic subtype: <ul style="list-style-type: none"> ○ Meningothelial (49%) ○ Transitional (43%) ○ Most WHO grade I 	<p>Signs</p> <ul style="list-style-type: none"> • Primary lesions <ul style="list-style-type: none"> ○ Proptosis ○ Optic atrophy ○ Retinociliary collateral vessels • Secondary lesions <ul style="list-style-type: none"> ○ Proptosis ○ Ocular paresis ○ Optic nerve edema
<p>Symptoms</p> <ul style="list-style-type: none"> • Primary lesions <ul style="list-style-type: none"> ○ Painless, progressive vision loss • Secondary lesions <ul style="list-style-type: none"> ○ Painless, progressive vision loss 	<p>Treatment</p> <ul style="list-style-type: none"> • Primary lesions <ul style="list-style-type: none"> ○ Observation ○ Stereotactic radiation ○ Surgical resection considered in certain situations (see text) • Secondary lesions <ul style="list-style-type: none"> ○ Observation ○ Surgery +/- radiation in certain patients (see text)

References

- Cannon PS, et al. The Surgical Management and Outcomes for Spheno-orbital Meningiomas: A 7-year Review of Multi-disciplinary Practice. Orbit 28(6), 371-376, 2009.
- Deepali Jain, MD et al. Intraorbital Meningiomas A Pathologic Review Using Current World Health Organization Criteria. Arch Pathol Lab Med. 2010 May;134(5):766-70.
- Dutton JJ. Optic Nerve Sheath Meningiomas. Surv Ophthalmol. 1992 Nov-Dec;37(3):167-83. Review.
- Eddleman CS, Liu JK. Optic nerve sheath meningioma: current diagnosis and treatment. Neurosurg Focus. 2007;23(5):E4. Review
- Kleihues P, et al. The WHO Classification of Tumors of the Nervous System. J Neuropathol Exp Neurol. 2002;61:215-225
- Mourits MP, van der Sprinkel JW. Orbital meningioma, the Utrecht experience. Orbit. 2001;20:25-33
- Shrivastava RK, et al. Sphenoorbital meningiomas: surgical limitations and lessons learned in their long-term management. J. Neurosurg. 2005; 103: 491-97
- Turbin RE, Pokorny K. Diagnosis and Treatment of Orbital Optic Nerve Sheath Meningioma. Cancer Control. 2004 Sep-Oct;11(5):334-41. Review.

Suggested citation format: Callk CB, Carter KD, Allen R. Meningiomas. EyeRounds.org. August 30, 2011. Available from: <http://EyeRounds.org/cases/137-meningiomas.htm>.