Abstract: Meningiomas are tumors arising from the meningo-endothelial cells of arachnoid layer of meninges. Optic nerve sheath meningiomas comprise 2 of orbital tumors. We are reporting a rare case of recurrent optic nerve sheath meningioma. Histopathological examination showed meningothelial type of meningioma.

Keyword: Recurrent optic nerve sheath meningioma, orbital tumor, meningo endothelial meningioma.

Introduction: In 1964, Felix Plater first described meningioma at an autopsy. In 1938, Harvey Cushing introduced them as a separate category of exoparenchymal tumors. Meningiomas arise from arachnoid cap cells and are usually attached to the dura. These tumors may arise from any location where meninges exists.

Case report:
A 55yr old female presented with history of forward protrusion of left eye since 4yrs, insidious in onset, gradually progressive, no h/o of pain or postural variation. It is associated with diminishment of vision in the left eye since 4 yrs, insidious onset, gradually progressive. Past history revealed h/o similar complaints of forward protrusion and diminishment of vision in the left eye 13 yrs back for which she was operated 10 yrs back. Discharge details showed a diagnosis of left optic nerve sheath meningioma for which transcranial orbitomy and excision of tumor was done. Following surgery proptosis had improved. Vision remained poor but some useful vision was retained. Family history was unremarkable.

<table>
<thead>
<tr>
<th>On Examination</th>
<th>Right Eye</th>
<th>Left Eye</th>
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<tbody>
<tr>
<td>Uncorrected Visual Acuity</td>
<td>6/24</td>
<td>6/24</td>
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<tr>
<td>Best Corrected Visual Acuity</td>
<td>6/24</td>
<td>6/24</td>
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<tr>
<th>Head posture and facial symmetry maintained.</th>
<th>Right eye</th>
<th>Left eye</th>
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<tr>
<td>Eye position</td>
<td>Orthophoria</td>
<td>Axial Proptosis</td>
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<tr>
<td>Iris</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Erythema</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
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Recurrent Optic Nerve Sheath Meningioma - a case report
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Exophthalmometry(Hertel’s) – Right Eye 17mm, Left Eye 23mm

Base 110mm) Intra ocular tension by applanation tonometry- Right
Eye 14 mm Hg, Left Eye 16 mm Hg. In upaze both eyes- 16 mm Hg. Colour vision and fields – Right Eye normal. Left eye couldn’t be
tested because of poor vision.

Fundus: Right Eye- normal.

Left Eye-media hazy due to lens changes. Disc was mildly
hyperaemic, there was blurring of disc margins. Cup was
obliterated. Optic-ciliary shunts were present in the inferior part of
the disc margin. Peripapillary nerve layer edema was noted with
choroidal folds present around the disc. Foveal
reflex was absent. Dermatological examination- no evidence of
neurocutaneous markers. Systemic examination- within normal
limits.

Biochemical investigations: RBS, blood urea, serum creatinine
and serum electrolytes were within normal limits. Coagulation
profile-within normal limits Hb%-11 gm/dl.

CT Orbit & Brain:

MRI:

Left Orbit- evidence of heterogenous mixed density lesion
measuring 4.00 x 2.44 x 2.08 cm involving left globe engulfing optic
nerve sheath complex with calcification within the lesion with
evidence of post operative changes within the frontal bone. Right Orbit- normal.

Histopathological slide

DISCUSSION

Meningiomas are tumors arising from the meningotheelial 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).1 Classification of
meningiomas is based on World Health Organisation(WHO):
Guidelines2,3. For most nervous system tumors the WHO grade is
really an estimate of malignancy. Grading for meningiomas reflects
the likelihood of recurrence and aggressive behaviour.

classification currently includes 15 named subtypes:

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

PRIMARY ORBITAL MENINGOMAS

Primary orbital meningomas (i.e. optic nerve sheath meningiomas) are derived from meningotheelial cells found
in the orbit. They account for approximately 2% of the
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
- Opto-ciliaryshunt vessels.

These lesions tend to grow circumferentially around the
optic nerve 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 loss4,5. 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 nondiabetic patient.
It is delivered via 3-dimensional stereotactic techniques in
order to minimize side effects6.

SECONDARY ORBITAL MENINGOMAS

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
- Clinoid
- Planumshpenoidale
- Frontoparietal area
- Optic nerve
- Optic atrophy
- Optic paresis
- Optic nerve edema

Presenting signs/ symptoms of secondary lesions include
the following:
- Proptosis
- Vision loss
- Ocular paresis
- Optic nerve edema

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. 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.

PROGNOSIS

Meningiomas are thought to be a slow-growing but progressive tumor. The majority of recurrences are thought to be due to residual tumor in the operative bed, which happen because of fear of causing severe functional deficits, where complete resection is attempted. Meningiomas of all locations have a recurrence rates ranging from 10 to 23%. Orbital meningiomas can invade the cavernous sinus, the dura of the sellar turcica, the lateral part of the sphenoid body, the annulus of Zinn, the ptengomaxillary fossa, and the lateral pharyngeal space. Recurrence rates of sphenoid ridge meningioma are much higher than those located in other regions, ranging from 25 to 50%. Recurrent sphenoorbital meningiomas tend to be large infiltrating tumors that have a propensity for invading vital neurological structures.

CONCLUSION

The location of orbital meningiomas poses a particular challenge to the surgeon. As such, residual tumor may be left after resection, resulting in higher recurrence rates than for meningiomas located in other regions. Fortunately, meningiomas are typically slow growing benign tumors that can be followed clinically and radiographically for recurrences. Should they recur, treatment options remain. Radiation therapy has been shown to be effective in controlling local tumor growth. Additional resection remains an option. A complete resection is still the most desired result, as it may be curative. Current advances in peri- and operative management - specifically preoperative endovascular embolization, intraoperative computerassisted image guidance, and refined surgical approaches - assist the surgeon in achieving optimum outcomes.

REFERENCES
