Case Report

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A Rare Case of Severe Proptosis Secondary to Orbitosphenoid Meningioma

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Abstract

Orbitosphenoid meningioma/Meningioma-en-plaque is a rare type of meningioma comprise 2-9% of all meningiomas, characterized by sheet likegrowth and bone hyperostosis. Total surgical resection is difficult and hence tumors have a high recurrence rates. We report here a case of orbitosphenoid meningiomaWHO classification Grade-IwithMicrocystic variant. The patient was a 65-year-old female presented to us with right eye severe proptosis, no perception of light, Relative Afferent Pupillary Defect and optic atrophy. Histology showed Meningioma, Imaging modalities revealed mass on either side of greater wing of sphenoid, which is extending intraorbitally on right side occupying the extraconal and intraconal compartments, another component of the lesion is seen extraaxially along right temporal convexity with a enhancing dural tail suggestive of right sphenoid wing meningioma. The tumour was removed by Right frontotemporal craniotomy, lateral orbitotomy and near total decompression of the mass. Operative findings showed advanced disease almost encroaching upon the cavernous sinus but not infiltrated.

Keywords: Proptosis; Orbitosphenoid meningioma; Craniotomy and orbitotomy.

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Introduction

Orbitosphenoidmeningiomas arise from arachnoid cap cells (not dura) that are associated with the arachnoid granulations and follow neural structures through their foramina.[1] These are usually secondary lesions of the orbit arising from the sphenoid. This is in comparison to the rarer primary meningioma of the orbit that arise from the optic nerve sheath and then spread to the sphenoid bone producing hyperostosis.[2] Meningiomas may occur anywhere where arachnoid cells are found (between brain and skull, within ventricles and along spinal cord). Ectopic meningioma may arise within the bone of the skull (primary intraosseousmeningiomas)[3], and others occur in the subcutaneous tissues with no attachment to the skull.

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Case Report

A 65 year old female patient presented to us with history of gradual progressive forward protrusion of right eye(RE) since 4 & ½ years, gradual progressive diminution of visionfor 6 months followed by complete loss of vision since 1 year. She had also noticed mass at the medial aspect of the right upper eyelid since 1 year (figure 1(a)), dull ache in the temporal regionon and off since one year. There was no other significant history.

On ocular examinationno perception of light in RE and 6/12, N-10 with best corrected visual acuity of 6/6, N6 in left eye(LE). Right eyeball is protruded, deviated outwards and downwards on primary position of gaze, lower scleral show 10 mm, distance between lateral orbital margin and apex

of cornea-28 mm, periorbital edema was present. Conjunctival chemosis was present, whereas cornea, anterior chamber, iris was normal, butthere was relative afferent pupillary defect in RE and round regular reactive pupil in LE, Lens was grey in both eyes. On palpation there was a presence of mass between orbital margins and eyeball, which is firm to hard in consistency,nontender, no palpable pulses were felt, finger cannot be insinuated between orbital margins/eyeball, proptosis doesn't increase in size on bending forward or coughing, on auscultation no bruit heard.Ocular fundus examination revealed primary optic atrophy of the RE, whereas LE fundus was normal. Extra ocular movements were restricted in all cardinal positions of the gaze in RE and normal in LE.Perimetry of LE showed normal visual fields.

All routine investigations were normal.

Biopsy of the lesion was suggestive of Meningioma WHO grade I of microcysticvariant (figure 2).

MRI Brain with Orbit was also suggestive of Right sphenoid wingmeningioma (figure 3 (a to f))

As a team work Neurosurgeon, Oncosurgeon and ophthalmologists discussed about the case in detail. Visual prognosis was explained to the patient and proceeded with Right frontotemporal craniotomy (various modifications to this basic approach can be used depending on the surgeon's preference),[4,5] lateral orbitotomy and near total decompression of the tumor under General anesthesia.

Patient was positioned in supine in a Mayfield head holder with neck extended. At induction first generation cephalosporin was injected.Bicoronal incision was made just anterior to the tragus extending to the contralateral superior temporal line, temporalis was reflected.Burr holes were drilledand frontotemporalbone flap was lifted up. Mass was popping out from the orbit (figure 4(a)), encasing upon the rectus muscles. Initially intracranial extension of the mass was removed, then the orbital mass removed carefully. Tumor was soft to firm, moderately vascular, relatively good plane all around. Thickhyperostosed Sphenoid bone was dissected by piece meal. The defect was filled with temporalis fascia, andfrontotemporal bone flapreplaced back and wound was closed in layers.

Surgery was uneventful, post-operatively patient recovered well (figure 1(b)) and CT scan of orbit post operatively (figure 3 (g & h)) showed gross removal of mass. Patient had minimal scalp edema which subsided after few days. Patient had right eye ptosis in view of involvement of levator muscle. On follow-up of 6 months patient is doing well without fresh deficits. Final diagnosis was made based on MRI and operative findings asOrbitosphenoid Meningioma.

Discussion:

Orbitosphenoidmeningiomas is also called Meningioma-en-plaque is a rare type of meningioma comprise 2-9% of all meningiomas,[6] (Incidence of other subtypes of Meningiomas by location are given in table 1 below) characterized by sheet like growth, bone hyperostosis, usually slow growing, circumscribed(non-infiltrating), benign lesions andmay be asymptomatic. May be multiple in up to 8% of cases,[7] this finding is more common in neurofibromatosis.

Orbitosphenoidmeningiomas were described as meningioma en plaque and classification scheme for sphenoid wing meningiomas first proposed by Cushing and Eisenhardt in 1938 and described as:

Inner or clinoidal

Middle or alar

Outer or pterionalmeningiomas

Orbitosphenoid wing meningiomas may originate from all of the subtype described by Cushing, but usually are associated with the enplaque (or hyperostosing) meningiomas of the lateral sphenoidwing.[8] They will have extension into orbit and often involve the cavernous sinus. These lesions can involve the orbital apex with associated hyperostosis compressing the contents of the orbit and the optic nerve,[9] all these findings were present in our case. Furthermore, these lesions can extend into the anterior, middle, infratemporal/ zygomatic fossae as well as the paranasal sinuses. Orbitosphenoid wing meningiomas as with most meningiomas are more common in females, the most common presenting complaint is proptosis and retro orbital pain, all these seen in our patient. Patient may also present with optic neuropathy, which is most common in recurrent cases, decreased facial sensation in the maxillary and ophthalmic division of the trigeminal nerve due to cavernous sinus involvement, (sensations were intact in our patient, as cavernous sinus was not involved) and diplopia. Visual field deficits tend to occur in cases with optic nerve compression or encasement. In our

case it was advanced disease with no perception of light.These lesions can have extensive interosseous involvement without dural involvement resulting in proptosis and optic nerve compression. Changes can occur in the optic disc and surrounding fundus like optic disc edema and optociliary veins in the setting of progressive visual loss, in our patient fundus changes were advanced, as it was showing optic atrophy.

Table 1: Frequency of meningiomas by location.

Cushing and Eisenhardt (1938)		(2003) Meta-analysis	
Parasagittal/ Falcine	23%	25%	Parasagittal/ Falcine
Convexity	17%	19%	Convexity
Sphenoidal ridge	17%	17%	Sphenoidal ridge
Olfactory groove	9%	8%	Olfactory groove
Suprasellar	9%	9%	Suprasellar
Cerebellar chamber	7%	4%	Posterior fossa
Tentorial	6%	3%	Tentorial
Spinal	6%	3%	Spinal
Intraventricular	2%	1%	Intraventricular
Meckel's cave	2%	4%	Meckel's cave
Foramen magnum	0.3%	1%	Foramen magnum
Orbital/optic nerve sheath	0.3%	1%	Orbital/optic nerve sheath

(**Note:** This table is from text book of Meningiomas by Joung H. Lee, page no. 6)

Figure 1 (a, b): preoperative and postoperative pictures of the patient.

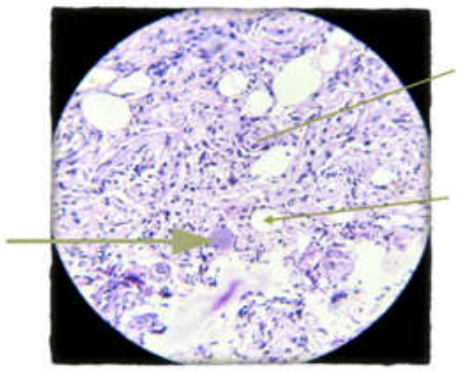

Figure 2: Histopathology tumor cells composed of oval to spindle cells arranged in sheets, bundles and focal areas showing whirling (straight line), microcystic spaces (small arrow) and psammoma bodies (big arrow).

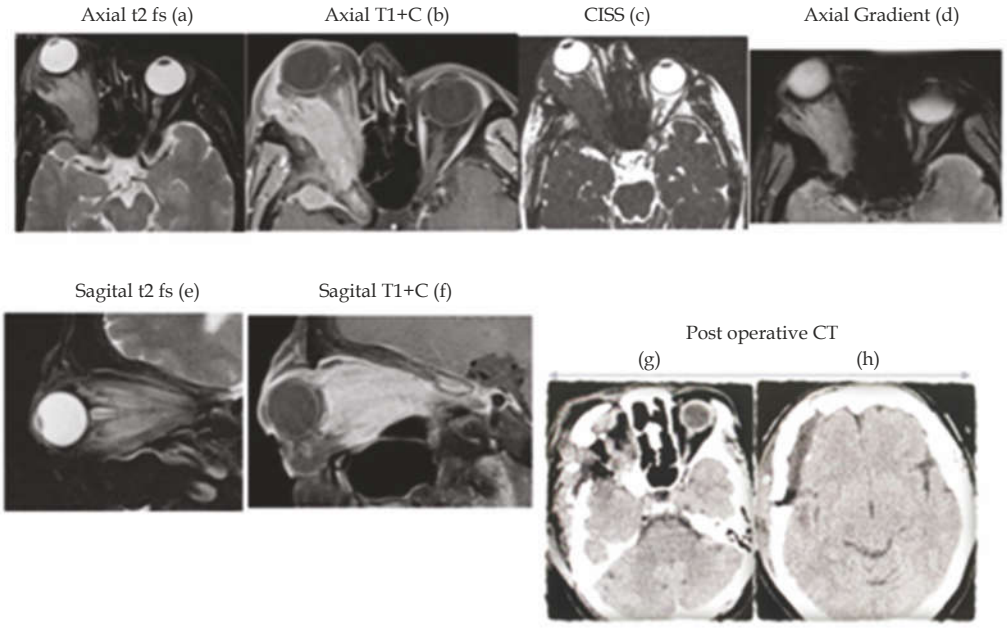

Figure 3(a-to-h): MRI Brain with Orbit (a-to-f), (b) showing dural tail enhancement within the cranium, (d) Gradient scan showing hyperostosis of the right sphenoid wing. (c) CISS (constructive interference steady state) to see tumor vascularity. (e) Shows mass is arising from sphenoidwing, not from the optic nerve. CT Orbit (g, h): post operatively.

Intra-operative (a) Closing with bone flap (b)

Figure 4: Intraoperative photos- tumor mass (small arrow), sphenoid bone hyperostosis (bigarrow).

Conclusion

With this case report we describe chronological occurrence of symptoms, and its sequelae. Diagnosed a rare case of orbitosphenoid meningiomas, intervened at right time before it infiltrated the cavernous sinus and optic chiasma. So we could save the left eye vision for normal functioning of life and prevented the neurological complications to develop because of mass effect intracranially.

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