

Unilateral proptosis veiling aggressive tumour**Rajwinder Kaur¹, Mandeep Kaur¹, Kulbir Kaur², Harijot Singh³**¹Department of Ophthalmology, Gian Sagar Medical College and Hospital, Ram Nagar, Rajpura, District Patiala, Punjab 140601, India²Department of Surgery, Gian Sagar Medical College and Hospital, Banur, Patiala, Punjab 140601, India³Department of Pediatrics, Gian Sagar Medical College and Hospital, Banur, Patiala, Punjab 140601, India***Corresponding author**

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Abstract: Out of all the brain tumours, meningiomas account for 15% of the total share and Sphenoid ridge is the third commonest site, though in children, only 4.2-15% of intracranial meningiomas involve the sphenoid wing. A case of 11 years old male with h/o unilateral proptosis finally fell into diagnosis of sphenoid wing meningiomas. Child underwent neurosurgery, post op images revealed infiltrative tumour. Aggressive meningiomas should be treated with combined approach including surgery, radiotherapy and chemotherapy.**Keywords:** Cavernous sinus, proptosis, psammoma bodies, hemangiopericytoma.

INTRODUCTION:

Shining light on Sphenoid wing meningioma also known as ridge meningioma are most common basal meningioma which are usually more commonly seen in women and are rarely seen in children as and when they occur in children they rather present aggressively, hence along with surgical management they usually require chemotherapy and radiotherapy[1-3]. Early in their development, symptoms are often masked and as these “little devil” grows thus presents with symptoms including decreased visual acuity, proptosis, visual field defects, afferent pupillary defects and loss of facial sensation[6]. Careful evaluation of evolving clinical findings is the key to the early detection necessary to preserve neurological function. We report a 11 year old with proptosis right eye diagnosed with sphenoid wing meningioma on MRI.

CASE REPORT:

A 11 year old male presented with history of proptosis right eye. H/O proptosis right eye which was gradual in onset and progressing slowly, there was no history of pain, redness, local trauma, head injury, headache. His general health status was good. No features of thyroid disorders were observed. There was no positive ocular family history (Fig-1).

On Examination:

Visual acuity was 6/6 in both eyes and intraocular pressure OD=12, OS=11 and normal colour vision. Restricted elevation and adduction of ocular movements in right eye, rest ocular movements were normal (Fig-2). Both pupils were reactive, and no afferent papillary defect was noted. Biomicroscopy showed normal anterior segment with normal

intraocular lens and normal fundus both eyes. Dilated fundus exam was unremarkable.

X-ray orbit showed hyperostosis of sphenoid wing right side (Fig-3).

CECT head revealed large, intensely and heterogeneously enhancing extraaxial lesion 8.8*6.6*5.2 cm in the right temporoparietal lesion involving sphenoid wing and squamous temporal bones on right side suggestive of meningioma, multiple dense calcifications seen in the lesion. Superiomedially lesion extended upto level of centrum semiovale, compressing and displacing basal ganglion, thalamus and right lateral ventricle towards left side with midline shift of 9mm towards left side. Inferiorly lesion is bulging into right orbit compressing superior rectus causing proptosis (Fig-4).

On post contrast images the lesion shows intense heterogenous enhancement with multiple non enhancing necrotic areas within. Prominent feeding vessel is seen along the superior aspect of the lesion.

Patient was referred to neurosurgery department for surgical management where excision of tumour was done by open craniotomy approach, due to local invasion of tumour, surgery was followed by course of chemotherapy and radiotherapy.

Histopathology of excised specimen revealed cellular tumour with interspersed hemangiopericytoma, tumour cells were pleomorphic with hyperchromatic nuclei, small nucleoli and moderate cytoplasm with indistinct cell borders. Focally bone infiltration noted.

Mitosis is 18 -20/hpf. Features of high grade hemangiopericytoma WHO GRADE 3.

Post-operative CECT HEAD, show local local bony infiltration and aggressive behavior of this tumour(Fig-5,6).



Fig-1: Proptosis right eye



Fig-2: Restricted elevation and adduction of ocular movements in right eye



Fig-3: X-ray orbit showed hyperostosis of sphenoid wing right side.

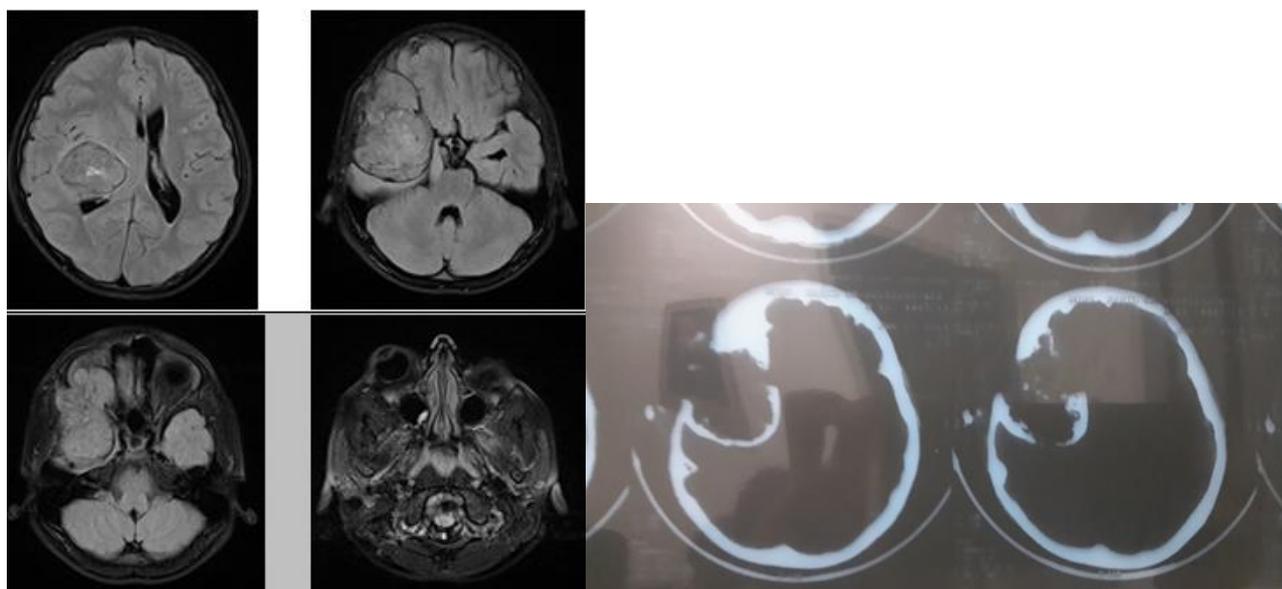


Fig-5: Pre Operative and Post Operative CT Scan Head



Fig-6: Post OP Showing Upward Restriction

DISCUSSION:

Above case description demonstrates the occurrence of otherwise rare sphenoid wing meningioma in this age group and usually present with ocular symptoms. Tumors found in the external third of the sphenoid are of two types: en-plaque and globoid meningiomas. En plaque meningiomas characteristically lead to slowly increasing proptosis with the eye angled downward. Much of this is due to reactive orbital hyperostosis. With invasion of the tumor into the orbit, diplopia is common. Patients with globoid meningiomas often present only with signs of increased intracranial pressure. This leads to various other symptoms including headache and a swollen optic disc. The differential diagnosis for sphenoid wing meningioma includes other types of tumors such as optic nerve sheath meningioma, cranial osteosarcoma, metastases, and also sarcoidosis. Sphenoid wing Tumors growing in the inner wing (clinoidal) most often cause direct damage to the optic nerve leading especially to a decrease in visual acuity, progressive

loss of color vision, defects in the field of vision (especially cecentral), and an afferent pupillary defect. If the tumor continues to grow and push on the optic nerve, all vision will be lost in that eye as the nerve atrophies. Proptosis, or anterior displacement of the eye which was how patient presented to us, and palpebral swelling may also occur when the tumor impinges on the cavernous sinus by blocking venous return and leading to congestion. Damage to cranial nerves in the cavernous sinus leads to diplopia. Cranial nerve VI is often the first affected, leading to diplopia with lateral gaze. If cranial nerve V-1 is damaged, the patient will have pain and altered sensation over the front and top of the head. Horner's syndrome may occur if nearby sympathetic fibers are involved[10,11,8]. 'Early diagnosis,early and aggressive treatment' is the key to manage this tumour.MRI and CT scan are the tools for diagnosis along with complete ocular and neurological workup. Treatment of sphenoid wing meningiomas often depends on the location and size of the tumor. Gamma knife radiation and microscopic

surgery are common options. Their encapsulated, slow growth makes meningiomas good targets for radiosurgery[4,5]. Higher grade tumors have been shown to correspond with higher recurrences. Depending on the grade and extent of resection, from less than 1 in 10 to over two-thirds of tumors will recur after surgical excision. Follow-up clinical exams, as well as neuroimaging, can aid in detecting recurrences. As many meningiomas have receptors for progesterone, progesterone blockers are being investigated. Two other drugs that are being studied for use are hydroxyurea and interferon alpha-2b.

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