Secondary orbital meningioma in an adolescent female- A rare case report

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ABSTRACT

Meningiomas are benign intracranial neoplasms that account for 18% of all intracranial neoplasms. (1) Ectopic meningiomas of the skull are however rare and account for 1 % of all meningiomas- they may be primary or secondary. (2)

We present a case of a 12 year old female child with a secondary orbital meningioma, with symptoms of progressive swelling and proptosis. The lesion was successfully excised and the patient has been free of symptoms since past 4 months. Review of relevant literature has been discussed subsequently.

Key words: Orbital Meningioma; Meningiothelial; Adolescent.

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INTRODUCTION

Meningioma are the tumour that arises from central nervous system meninges. Meningioma are neoplasms derived from arachnoidal cells. Meningiomas accounts for approximately 18% of all primary intracranial neoplasms and 25% of intraspinal neoplasms. Common sites of the ectopic meningioma are skull bones, paranasal sinuses , nose ,orbit, scalp, middle ear , neck

CASE REPORT

A 12year old female patient presented to the ENT OPD with history of right sided swelling over the superior orbital rim and proptosis for the past 12 months, which was gradually progressive in nature. There was no history of diminution of vision, epiphora, diplopia, headache, previous surgery or trauma to the region. On Examination, there was a 3 x 2 cm, bony hard swelling palpable in the medial aspect of the superior orbital rim, pushing the left eye inferolaterally. The swelling was non pulsatile and did not increase in size on coughing.Visual acuity and extra ocular movements were normal. A diagnostic nasal endoscopy did not reveal any bulge in the lateral wall of the nose.

We proceeded to evaluate the patient with a Contrast enhanced Computed tomography (CE-CT) scan of the nose and orbit. Scan showed a 2.8 x 1.5 x 1.5cm heterogenous soft tissue mass with specks of calcification, occupying the superiomedial quadrant of the right orbit displacing the eye inferolaterally. The lesion was in close proximity to the superior oblique

muscle. The bones adjacent to the lesion that is orbital process of the frontal bone, medial wall of the orbit and lamina papyreacea of the ethmoid bone showed loss of diploic space and hyperostosis.
The patient was planned for an excision of the mass through a Lynch Howarth incision under general anaesthesia.Skin, subcutaneous tissue and orbicularis oculi muscle were retracted downwards. Orbital septum was opened .Lesion was found to breach the orbital periostium with minimal adhesion to the superior oblique muscle. Mass was shelved off from the superior oblique muscle and delivered in toto. Mass was firm in consistency. The underlying bone in the superomedial aspect of the orbit was drilled out.

Histopathological examination of the tumor with Hematoxylin and Eosin stain showed sheets of tumour cells arranged in a syncytial pattern, composed of round to ovoid nuclei, with nuclear grooving and intranuclear inclusions. No increase in mitotic figure/ psammamatous calcification was seen. Tumour cells were positive for progesterone receptor and S- 100. This was suggestive of a meningothelial meningioma- WHO grade I.

The patient had an uneventful post-operative recovery and at the end of 1 month follow up her proptosis had completely regressed. Patient has been in follow up now since past 4 months without any symptoms.

Figure 1:Presentation of patientwithswelling in the medial aspect of the superior orbital rim, pushing the left eye inferolaterally.

Figure 2:Intraoperative photograph showing the mass after retraction of orbicularis oculi

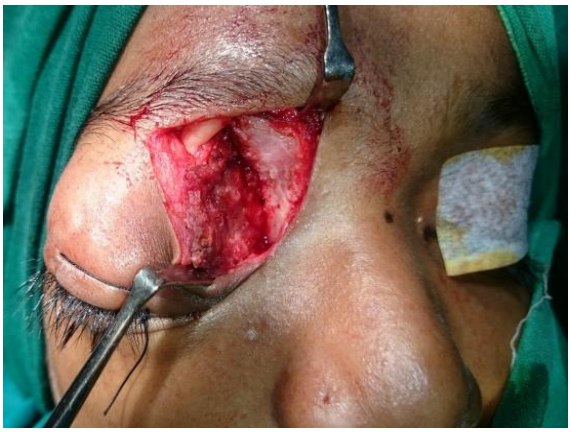


Figure 3:Post-operative photograph of mass

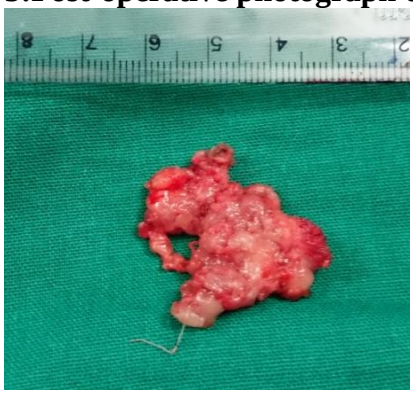
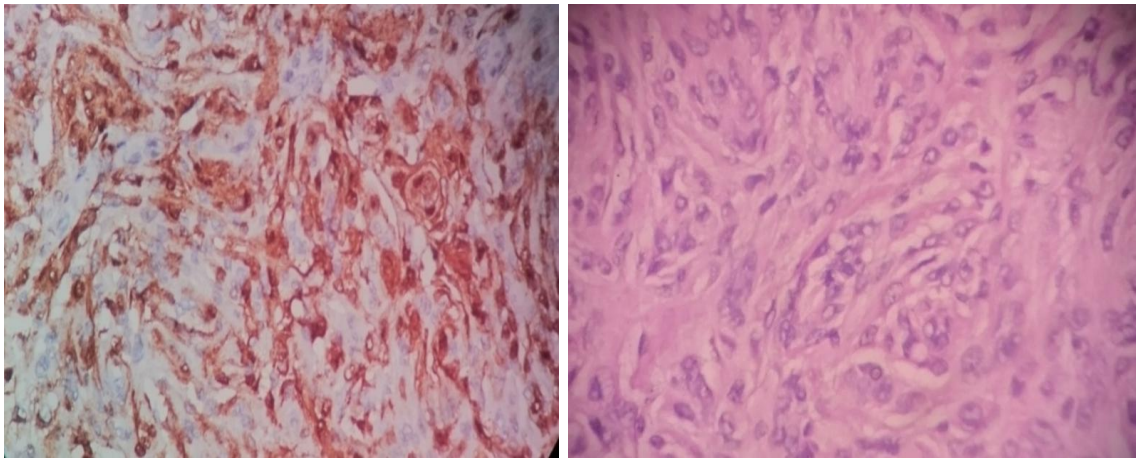


Figure 4:Hematoxylin and Eosin stain showingsheets of tumour cells arranged in a syncytial pattern, composed of round to ovoid nuclei. (10 X)



DISCUSSION

Virchow is credited for describing the pathological features of meningioma, although it was Cushing in 1922 who introduced the term meningioma. (3)Meningiomas of the orbit account for 3 to 9% of all orbital tumors, and can be divided into primary (0.4 to 2% of intracranial meningiomas) and secondary types.(4,5) Primary type arises from the optic nerve sheath, while secondary may arise from outside the orbital confines, usually the inner and outer aspect of the sphenoid wing. Like intracranial meingiomas, their orbital counterparts too have a female predilection, ranging from 73-84%. (5)

Factors predisposing to the development of orbital meningiomas include neurofibromatosis- especially in the pediatric age group, and exposure to radiation. (6,7)Meningiomas occurring in children are uncommon and account for only 1.5 to 2.3% of all intracranial neoplasms.(8)These are associated with poorer prognosis as compared to adults.

The usual presentation of orbital meningiomas include unilateral visual loss and progressive painless exophthalmos. Other features include optic disc changes, headache, nausea, vomiting and diplopia.

The World Health Organization (WHO), in its revised classification has described 15 subtypes of meningiomas, based on which meningiomas have been graded into 3 types- grade 3 being most aggressive. Our case is that of a meningothelial meningioma- belonging to grade 1 or benign type. Pathologically these appear as solid lobulated masses or sheets of meningothelial cells. Their cell membrane is not well defined and mitotic activity is low.

Secondary orbital meningiomas present with bone involvement in the form of hyperostosis, incidence of which varies from 25 to 40%.(3,9) Various theories for hyperostosis have been proposed including vascular disturbance in bone due to tumor, osseous reaction to tumor, preceeding trauma and osteoblast stimulation by adjacent normal bone due to tumor

reaction.(10,11,12,13) Presence of hyperostosis is clinically important because of the need for complete resection of the hyperostotic bone to prevent recurrence.

Radiologically the presence of calcification on a computed tomogram is highly suggestive of a meningioma. Homogenous enhancement of the mass is seen on administration of a contrast medium. This will be associated with the presence of hyperostosis or bony erosion in the case of secondary orbital meningioma.

The treatment options for orbital meningiomas can be surgical, endovascular embolization and stereotactic radiosurgery. Of these surgery is the treatment of choice for secondary orbital meningiomas. Surgical access may be gained by either a lower lid, subciliary, superior lid crease or an eyebrow incision.

Meningiomas have an overall recurrence rate of 10 to 23%, with the major cause being residual tumor in the operative site. (14)

CONCLUSION

Secondary meningiothelialmeningiomas in pediatric age group are very rare. Bony involvement is known to occur, especially in the secondary tumors. Surgical modality of treatment with removal of involved bone is the treatment of choice. Recurrences especially in the pediatric group have known to occur- but since meningiomas are benign and slow growing masses, they can be followed up clinically and radiologically for recurrences. Current advances in treatments, including surgical approaches have assisted surgeons to achieve optimum outcomes.

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