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Orbital Tumours: An Overview of Surgical Management and Histopathology

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Abstract
Introduction: *Orbital tumours represent a diverse spectrum in their histopathological nature. Vast arrays of lesions occur in the orbit and merit a systematic work up. The management options depend on the location of the tumour, radiological features and the probable pathology of the lesion.*
Materials and Methods: *Orbital tumours presented to a tertiary care centre from January 2010 to December 2016 were evaluated retrospectively. There were a total of 28 patients. Hospital records, Outpatient Department Registers, and Records from Department of Radiology were the sources of data collection. Presumptive pre-operative diagnosis was based on Computed Tomography and Magnetic Resonance Imaging findings. Different surgical approaches employed include Superior orbitotomy, Lateral orbitotomy, and Pterional craniotomy depending upon the location of tumour.*
Results: *The presenting complaints in most cases were proptosis (89%).Other symptoms were decreased vision (53.5 %), diplopia (28%), pain (1.2%) and lacrimation (0.8%). Histopathological findings showed 82% benign tumours and 18% malignant tumours. Schwannoma was the commonest (26%) among benign tumours. Visual acuity improved in 72% of cases and remained the same as preoperative in 28%. A recurrence rate of 14.29% is present in our series. Adjuvant therapy was started in the malignant lesions.*
Conclusion: *The best strategy for managing orbital tumours is largely determined by the location, size and extends of the lesion. Careful evaluation with history and physical examination along with modern imaging studies provide invaluable information regarding the possible origin of an orbital lesion. Definitive surgical treatment remains the mainstay of therapy for the majority of orbital tumours.*
Keywords: *Orbital tumour, proptosis, orbitotomy.*

Introduction

Orbit is a complexly organized group of neural, vascular, muscular, ligamentous, and osseous structures[1]. The bony orbit is a conical structure with the apex pointing posteriorly. The walls of the orbit are formed by seven bones namely frontal, zygomatic, sphenoid, lacrimal, ethmoid, palatine and the maxilla. The nerves and vessels entering and exiting the orbit pass through the optic canal and superior orbital fissure, which are partially surrounded by an annular tendon, from which the rectus muscles arise. The soft tissue

structures of the orbit are contained in the orbit which include the globe, the extra ocular muscles, the optic nerve–sheath complex, the lacrimal apparatus, and various vascular and nerve structures.

Tumours that involve the orbit can be classified into two major groups: primary tumours of the orbit and tumours with other sites of origin that extend into the orbit[2]. They may be further classified as benign and malignant.

Orbit and its contents are visualised using computed tomography and magnetic resonance imaging. Bony anatomy and its related pathologies, foreign bodies and calcified lesions are delineated with CT whereas MR imaging has pivotal role to play revealing finer details of neurovascular as well as soft tissue structures in the globe. Special sequences often used are short tau inversion recovery sequences (STIR) suppressing the signal from fat. CT is superior to MRI in surgical planning because of its ability to show bony anatomy, but MRI is preferred when optic nerve involvement by the tumour or disease process must be examined. Once a thoughtful sequential diagnostic workup has been completed, the location and extent of the pathologic process must be defined[3].The points that should be taken into account are whether the tumour is benign or malignant, its relationship to the muscle cone and optic nerve, laterality with respect to optic nerve, the bony integrity of orbit, whether optic canal is widened or hyperostotic, and finally whether lesion is destructive or not.

Definitive surgical treatment is possible for the majority of symptomatic orbital tumours, extensive lesions involving the medial part of the orbit and anterior skull base are still challenging. Detailed knowledge of the microanatomy of the orbit is necessary and would allow surgeons to overcome the pitfalls of intraorbital surgery[4].

Material and Methods

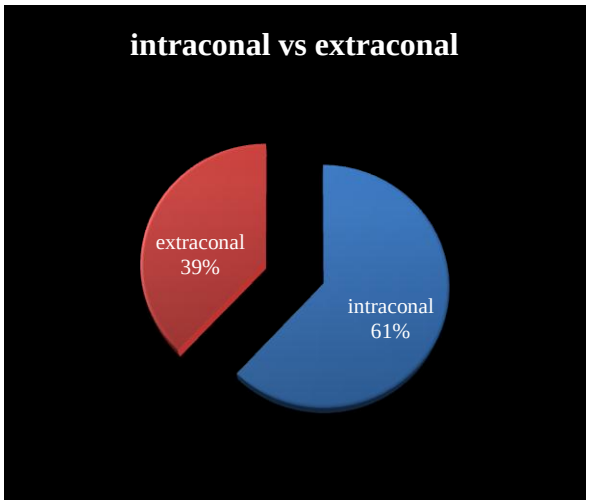
Retrospective analysis of orbital tumours operated between January 2010 and December 2016 in a tertiary care centre was done. The major entities studied were site of lesion, age, sex, the presenting symptoms, imaging characteristics, operative approaches, and final histopathology .Post operative outcomes evaluated in terms of resolution of proptosis, improvement in visual acuity. Data acquisition was done from outpatient clinic, hospital records, and operative notes. Patients were pre operatively imaged with CT and and MRI. Follow up was done 3 months after the surgery and yearly thereafter.

Results

Total number of cases was 28.

Site: 17 cases were intraconal and 11 cases were extraconal

Fig1: Site of lesion



Age: There were 22 adults and 6 children

Fig 2: Age distribution

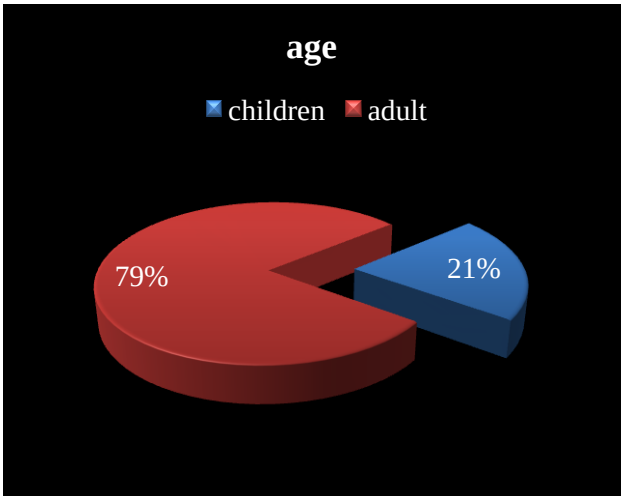
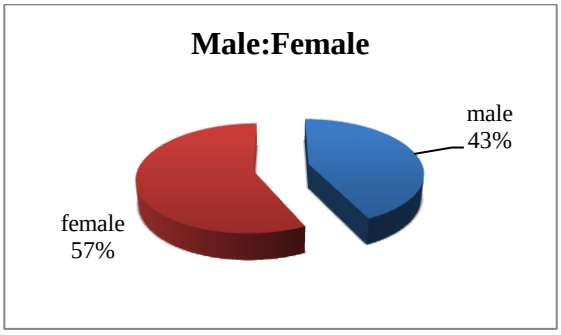


Table 1: Age wise distribution of orbital tumours

Age in years	Number	Percentage
0-9	3	10.71
10-19	3	10.71
20-29	1	3.57
30-39	5	17.86
40-49	7	25.00
50-59	3	10.71
60 and above	6	21.42

Sex: Among the total number of patients 12 were male and 16 were female

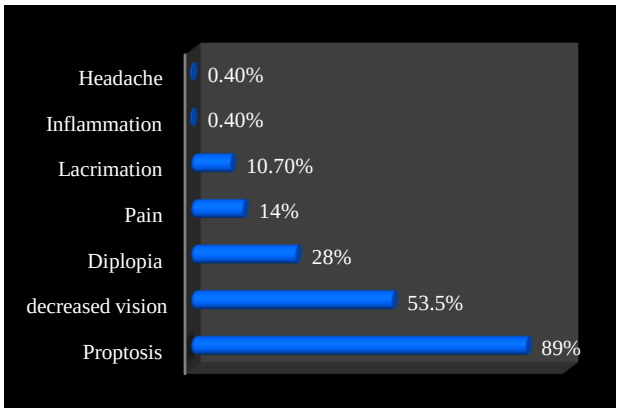
Fig 3: Sex



Clinical symptoms

Proptosis was the major symptom that brought patients to the outpatient department. Many were noted by the patient's relatives, some were by patient himself or herself. The second most common complaint noted was decreased vision. Less noted were pain, lacrimation, and headache.

Fig 4: Clinical symptoms



Surgical approach

Table 2: Operative approach

Operative Approach	No of cases
Lateral orbitotomy	13
Superior orbitotomy	11
Pterional approach and excision	04
Total	28

Histopathology

23 cases were benign and 5 cases were malignant

Fig 5: Benign Vs Malignant lesions

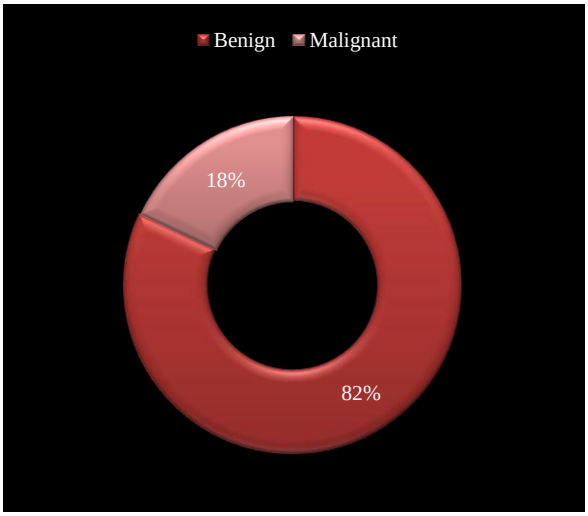
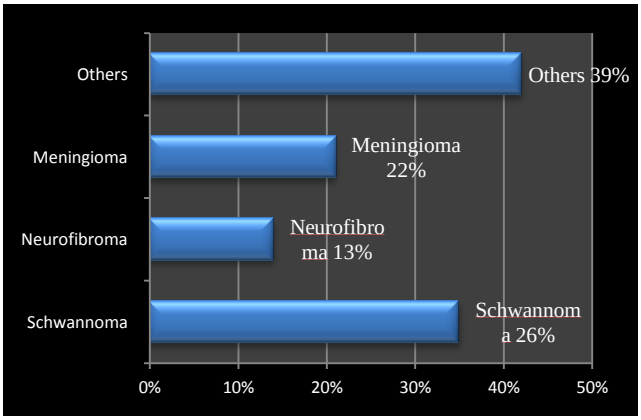


Fig 6: Benign tumours



23 cases were benign, among which Schwannoma was the first consisting of 6 cases (26%), Meningioma second 5 cases (22%) and Neurofibroma as third with 3 cases (13%). The rest mentioned as others were individual case of diverse pathology constituting a share of 9 cases (39%)

Table 3: Histopathology of benign tumours

Schwannoma	6
Neurofibroma	3
Meningioma	5
Venous hemangioma	2
Chronic inflammatory lesion	2
Benign lymphoproliferative lesion (pseudolymphoma)	1
Basal cell adenoma of lacrimal gland	1
Myofibroma	1
Mucocele	1
Dermoid cyst	1
Total	23

Fig 7: Malignant lesions

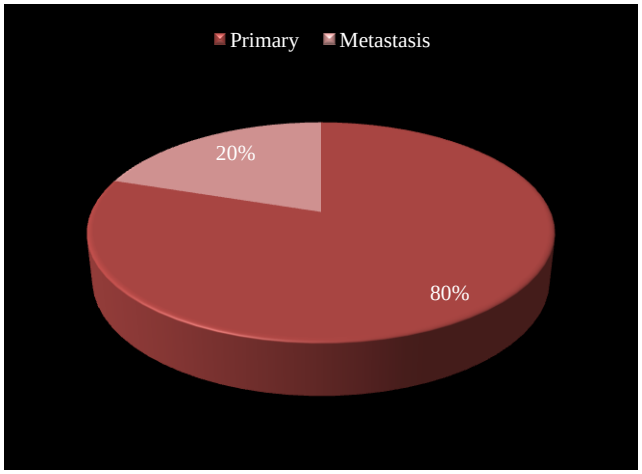


Table 4: Histopathology of malignant tumours

Histopathology	No of cases
Orbital plasmacytoma	1
NHL, low grade ,CD 20 +	1
Malignant small round cell tumour	1
Pleomorphic sarcoma	1
Malignant melanoma	1
Total	5

Table 5: Recurrence

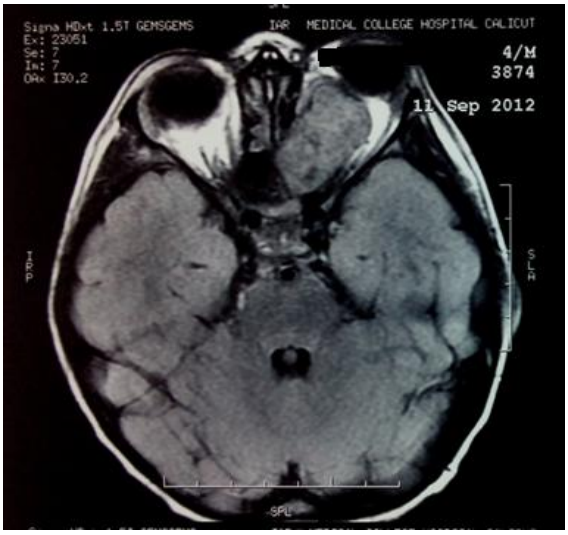
Histopathology of lesion	Number
Meningioma	2
Basal cell adenoma	1
Undifferentiated pleomorphic sarcoma	1
Total	4

A recurrence rate of 14.29 % was noted in our series.

In our series two rare cases worth mention are myopericytoma and Undifferentiated pleomorphic sarcoma (Malignant Fibrous histiocytoma). Myopericytoma which is in the same spectrum as myofibroma is a benign spindle cell neoplasm with smooth muscle differentiation, tumour cells positive for actin and desmin, MIB-1 labelling index low. Undifferentiated pleomorphic sarcoma (Malignant Fibrous histiocytoma) is a soft tissue sarcoma, composed of undifferentiated mesenchymal tumour cells with Fibrinohistiocytic morphology without definite differentiation. Common in parotid, scalp, neck, anterior skull base and is rare in orbit.

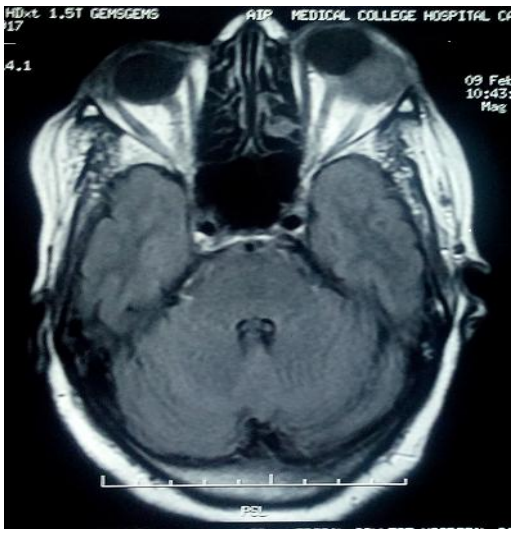
Follow up was done after 3 months and yearly thereafter. Proptosis reverted in 25 cases (89%).Visual acuity improved in 72% and remained stable in 28%.Malignant tumours were sent for adjuvant therapy. In our series pre operative CT and MR images were taken for all lesions. Proper pre operative planning done based on the data obtained by imaging. This was hugely helpful in planning appropriate surgical approaches.

Fig 8: Pre-operative image 1



Well defined intra orbital retrobulbar lesion on left side causing proptosis, isointense to muscle on T1, hypointense on T2,causing compression on optic nerve and superior ophthalmic vein. Rhabdomyosarcoma was considered as possibility with vascular malformation as differential diagnosis. Post operatively histopathology revealed myopericytoma a variant of myofibroma

Fig 8: Pre-operative image 2



Heterogeneously enhancing lesion in left orbit with expansion and thinning of bony walls possibly lacrimal gland tumour, turned out to be basal cell adenoma of lacrimal gland

Discussion

Orbital tumours can be broadly classified as two types. Primary tumours of the orbit and tumours that originate from adjacent structures and extending into the orbit. Depending upon the location they may be intraconal or extraconal. Based on histopathology tumours may be benign or malignant. Cavernous hemangioma, benign nerve sheath tumours, optic nerve sheath meningioma,lacrimal gland tumour, dermoid cyst, inflammatory pseudotumour and granuloma are some of the benign tumours. The commonest orbital lesion with significant intracranial extension is meningioma.[5] Malignant tumours may be primary or metastatic. Metastatic tumour was the commonest tumour in a large series of 414 orbital tumours reported by Maroon et al.[6]

Orbital tumours may produce symptoms and signs by acting as mass lesions, producing proptosis and limitation of eye movements,or by their effect on the optic nerve, the oculomotor nerve, the orbital branches of the ophthalmic division of the trigeminal ,the intraorbital vessels and rarely the nerve supply to the sphincter and dilator muscles of the iris.[7] Pain, lacrimation are the other common features. Pupillary abnormalities are rarepresentations due to isolated involvement of sympathetic and parasympathetic pathways.

Three syndromes may be associated with compression of the optic nerve due to orbital tumours. First is progressive visual olss associated with swelling of the optic disc. Second one is unilateral transient visual loss only in certain directions of gaze.[8] Third is optic atrophy and opticociliary shunt veins.

The earliest reports of surgery for orbital lesions involved approaches directed through the lateral wall of the orbit . Lateral orbitotomy was introduced by Kronlein in 1888. Since then several approaches were introduced.[9] Surgical approaches to orbit depend upon the site,size and extend of the tumour. When selecting orbital approaches, one should avoid crossing the plane of the optic nerve. Therefore, orbital pathology lateral to the optic nerve is accessed via lateral orbitotomies, and medial pathology is accessed via medial orbitotomies.[10]

Different surgical approaches employed briefly are[11]

Choice of operative approach for orbital tumours
Subfrontal approach with superior orbitotomy
• Superiorly and medially placed small and medium sized tumours • Intraconal tumours medial to optic nerve
Fronto-orbitozygomatic craniotomy
• Superiorly and medially placed large size tumours • All tumours with involvement of the apex
Fronto-orbitozygomatico-temporal craniotomy
• All tumours with middle fossa extension • All tumours with infratemporal extension
Lateral orbitotomy
• Laterally placed extraconal tumours • Intraconal tumours lateral or inferior to the optic nerve

Intraconal lesions presented to us outnumbered extraconal lesions. Median age at presentation was 40 years with predeliction for patients above 40 years. Clear female preponderance was noted in our study. Proptosis and decreased vision were the most noted symptoms that brought the patient to outpatient clinic. Most lesions were excised employing a lateral orbitotomy approach, superior orbitotomy ranked as second. Lesions like sphenoid wing meningioma that extended to the orbit was excised with a pterional approach. Majority of lesions in our study were benign. Of the malignant lesions one was metastasis from malignant melanoma. We got one case of Pleomorphic sarcoma which is a rare lesion in orbit. Proptosis reverted in majority of cases which were benign in histopathology. We had a recurrence rate of 14.29 %. Post operatively adjuvant therapy started in malignant lesions.

Conclusion

Strategy for managing orbital tumours is largely determined by the nature of the lesion. Definitive surgical treatment can be offered to symptomatic

orbital tumours. Surgical approach can be tailored according to the location of tumour. The transcranial approach is useful for the resection of medially situated lesions. A lateral orbitotomy is useful for lesions in the lacrimal fossa and lateral orbit. Tumours reveals an array of different histopathology. Gross total resection achieves a cure in case of of benign lesions. Proper adjuvant therapy based on histopathology is advocated in malignant tumours.

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