University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2017, Vol. 3(4)

RARE ORBITAL TUMOURS-A REPORT OF TWO CASES

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Abstract : Tumours of the orbit are rare diseases in ophthalmic pathology accounting for 3.5 - 4percentage of cases(1). They are a great challenge to the ophthalmologist since they present with avariety of signs and symptoms. We herein report the clinical, radiological and histopathologica features of Myoepithelioma of lacrimal gland in a forty five year old woman. The second patient was aunique case of primary Ewings sarcoma in an eighteen months old male child in whom diagnosis was confirmed using immuno histochemical staining. Both the cases have been reported due to their rare occurrence with no known incidence in literature.

Keyword :orbital tumours, lacrimal gland, Myoepithelioma, Ewings, immuno histochemical

INTRODUCTION

Though there are more than 1500 types of tumours which can occur inthe orbit, they are considered to be rare ophthalmic diseases[2]. Myoepithelioma of the lacrimal gland is an extremely rare monomorphicadenoma with very few reported cases[3] Ewing's sarcoma, a small round cell tumour is an extremely malignantneoplasm that primarily occurs in the long bones and pelvis while orbitaloccurrence is very rare[4].

CASE 1

A forty five year old female presented with swelling above the left eye forone year which had progressively increased in size to attain the present size(fig.1).History of defective vision and deviation of left eye was present sincechildhood. Examination showed fullness in the supraorbital region causing downward displacement of the globe with restriction of elevation. Theswelling was diffuse, firm and non-tender. Vision in the right eye was 6/18with pinhole 6/9 and in the left eye was 3/60 not improving with pinhole.Fundus examination was normal. Magnetic resonance imaging showed awell-defined heterogenous, lobulated lesion in the superior region with predominant solid component causing splaying of superior rectus andindentation of the globe (fig. 2,3). Under local anaesthesia, lateralorbitotomy with mass excision was done. Gross examination showed anencapsulated solid mass of about 28x19mm (fig.4). On histopathological examination, neoplasm composed of round, oval and spindle shaped cells with vesicular nuclei and basophilic nucleoli (fig.5) was seen and diagnosis of Myoepithelioma of lacrimal gland was made which was confirmed by immunohistochemical staining with actin (fig.6). Postoperatively, there wasno residual lesion with normal position of globe and complete restoration ofextraocular movements. The vision in the left eye was 6/60 with pinhole improving to 6/36 (fig.7).

CASE 2

An eighteen months old child presented with complaints of swelling in the left lower lid for the past twenty days. The lesion had rapidly increased insize to attain the present size (fig.8). On examination, there was a diffuses welling in the left infraorbital region causing upward displacement of globe.Insinuation of finger between the mass and floor was not possible. There was restriction of elevation, depression and abduction. Conjunctival chemosis was present with prominent vessels in the inferotemporal aspect. Fundus examination was normal. B-Scan imaging showed a lesion withfibrous septations and low internal reflectivity (fig.9). CT orbit showed an ill-defined, homogenous mass measuring 35.9x25.7mm in the infraorbital region with posterior extension (fig.10,11). There was erosion of maxilla and zygoma forming the floor of orbit on the lateral aspect along with lateral wall formed by zygoma. Based on these findings a differential diagnosis of Neuroblastoma, Leukemia and Ewing's sarcoma was made. Systemic workup which included complete blood count, peripheral smear, renal and liver function test, ultrasound abdomen and CT chest was normal. X-ray of the lower limbs did not show evidence of primary lesion (fig.12,13).Incisional biopsy was done which showed multiple small round cells with scanty cytoplasm, hyperchromatic nuclei and prominent nucleoli (fig.14).Immunohistochemical staining was positive for CD99 (fig.15) and negativefor neuron specific enolase confirming the diagnosis of Ewings/Primitive Neuroectodermal Tumour. Patient was started on chemotherapy.

DISCUSSION:

MYOEPITHELIOMA

These are benign tumours of the lacrimal and salivary glands.Myoepithelioma is a monomorphic adenoma with pure proliferation ofspindle cells. They are slow growing and do not have malignant potential.Few cases exist in lacrimal literature and all have behaved clinically andmorphologically like pleomorphic adenoma but without malignant potential.The tumours can be spindle, plasmacytoid or combined cells. CT imagingshows a well circumscribed lesion which is well encapsulated and thereforecan be removed in Toto through a lateral orbitotomy. They usually have anexcellent prognosis. These tumours are commonly found in the salivaryglands but lacrimal gland presentation is very rare with no recordedincidence in literature[5].

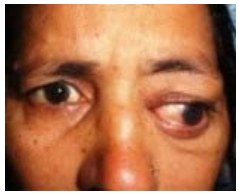

Fig 1

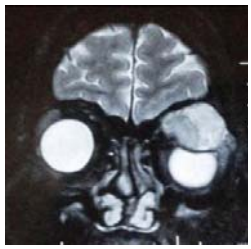

Fig 2

EWINGS SARCOMA

This is a primary malignancy of long bones with cells arising from theneuroectoderm. It arises from the medullary centres of long bones ofextremities and the pelvic girdle. It is considered to be part of primitiveneuroectodermal tumours. Primary tumours of skull occur in 2% of caseswith maxilla and mandible affected most commonly. But incidence ofprimary orbital involvement is not known. Most common presentation in orbitis metastasis from primary site.

Clinical features are due to the mass effect of the lesion encroaching onthe orbit. Proptosis, globe displacement, restriction of ocular motility or diplopia may occur if the tumour is anteriorly placed. Posterior tumours areassociated with optic nerve involvement. Pain is characteristically absent. CT imaging shows an enhancing, hyperdense, moth-eaten soft tissue lesioncausing erosion, destruction and calcification of bone. Histopathology showsa very cellular tumour with few fibrous strands. The cells are regular incontour and are round or oval in shape. Immunohistochemical staining willbe positive for CD99 but negative for neuron specific enolase and S-100[6].

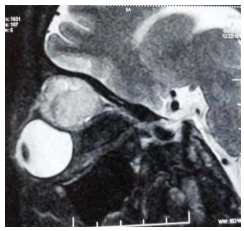

Fig 3

Ewing's sarcoma is considered a systemic disease there fore chemotherapy has become the first line of management replacing surgical excision which was used previously. The drugs commonly used are vincristine, doxorubicin and cyclophosphamide at 3 weekly interval for a maximum of 42 weeks followed by radiotherapy at a dose of 45Gy unitsover 5 weeks. Excision done after a course of induction chemotherapy isalso effective. The 5% survival rate has improved to 80% with modern treatment which involves a combination of chemotherapy, surgery and radiotherapy.

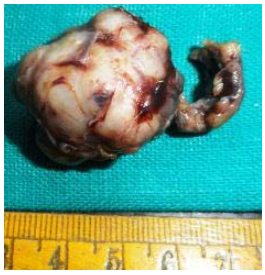

Fig 4

CONCLUSION:

Orbital tumours are always a great diagnostic challenge. Cooperation withpathologist, radiologist, oncologist and neurosurgeon is necessary forprompt diagnosis and treatment.

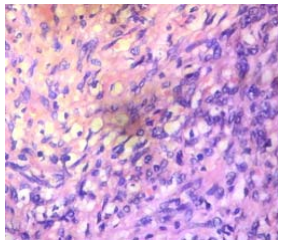

Fig 5

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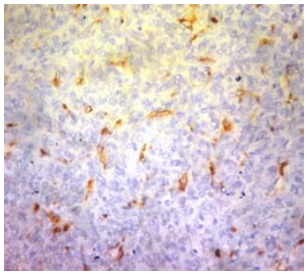


Fig 6

Fig 7

Fig 8

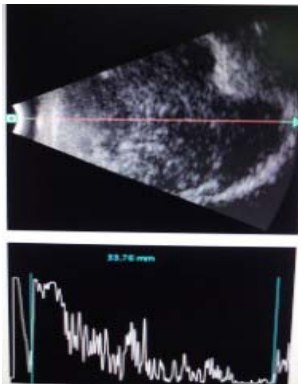


Fig 9

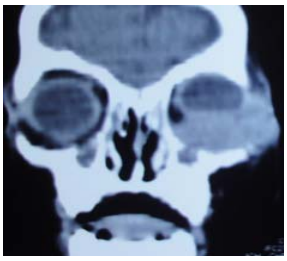
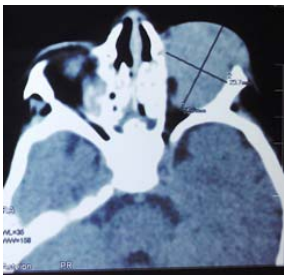


Fig 10



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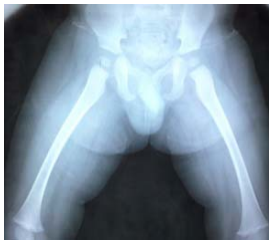

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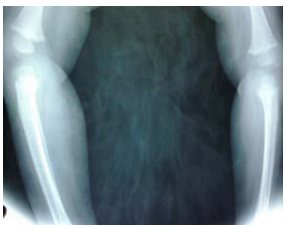

Fig 13

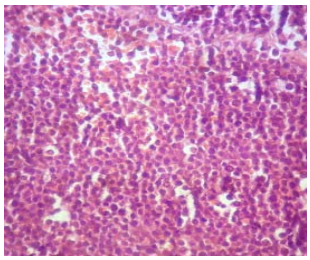

Fig 14

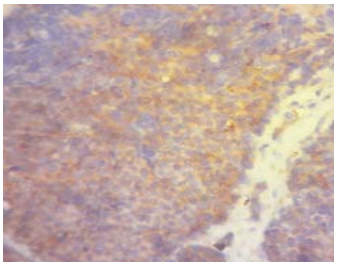

Fig 15