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Embryonal Rhabdomyosarcoma of Orbit- A Case Report

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ABSTRACT

Rhabdomyosarcoma is a highly malignant neoplasm of pleuripotent embryonic mesoderm, which commonly differentiates to form cells similar to rhabdomyoblasts of the foetus. Because of the presence of elongated cells that contain abundance of eosinophilic glycogen rich cytoplasm, it is generally referred to as embryonal form of rhabdomyosarcoma. Rhabdomyosarcoma is one of the most common primary malignant orbital neoplasms of child hood. It usually produces a precipitously progressing unilateral proptosis of sudden onset. We report a case of embryonal rhabdomyosarcoma of orbit presenting as large mass on medial side of right upper lid since 1 month in a 15 year old female patient.

INTRODUCTION

Rhabdomyosarcoma can occur in any anatomic location of the body where there is skeletal muscle, as well as other sites without skeletal muscle, such as the soft tissues of the orbit. It is presumed that most of these lesions, particularly the embryonal variants, arise from undifferentiated or pluripotential mesenchyme. The primary sites of rhabdomyosarcoma include[(1)] head and neck (40%), genitourinary tract (20%), extremities (20%), trunk (10%) and others (10%). In the eye, it can affect orbit, eyelids, conjunctiva and uveal tract[(3-5)]. Rhabdomyosarcoma is a rare tumor, with an annual incidence of 4.3 cases per million children[(2)]. Even though, it is the most common soft tissue sarcoma in childhood, with a mean age of 6 to 8 years at diagnosis[(1)].We present a case of a child with a rare tumor in which we had a high suspicion of malignancy, and early

diagnosis and treatment rendered the child free of systemic disease.

CASE REPORT

A 15 year old female patient was brought to the M.G.M. Medical College, Ophthalmology outpatient department with swelling over right upper and lower lid since 1 month. The mass initially developed on upper lid which gradually increased in size but after 10-15 days swelling involved the lower lid also, along with ecchymosis of both lids.which was not associated with apparent diminution of vision. There was no history of any injury, fever or chronic cough or any family history of malignancy.

CLINICAL EXAMINATION

Patient was conscious, cooperative, well oriented to time, place &person, with average build for age. The patient had no apparent diminution of vision. Right eyeball is displaced laterally because of the mass.There was gross restriction of ocular movements of the right eye in all positions of gaze.

On external examination a large multilobulated mass measuring about 5 *6 cm was seen over medial side of right eye associated with both upper & lower lid swelling and ecchymosis (figure 1). Mass was bluish red in colour, irregular in shape, skin over the mass was necrosed and bleeding area was present over the mass with no abnormal pulsations. Swelling was firm in consistency, warm as compared to surrounding area, tender, fixed to the underlying structure and overlying skin. It was non reducible & non compressible, pulsations were not felt. Preauricular and submandibular lymph nodes were fixed and hard on palpation. Conjunctiva was chemosed. Rest anterior segment and posterior segment was normal. Left eye visual acuity,(6/6),anterior segment and fundus examination was normal.

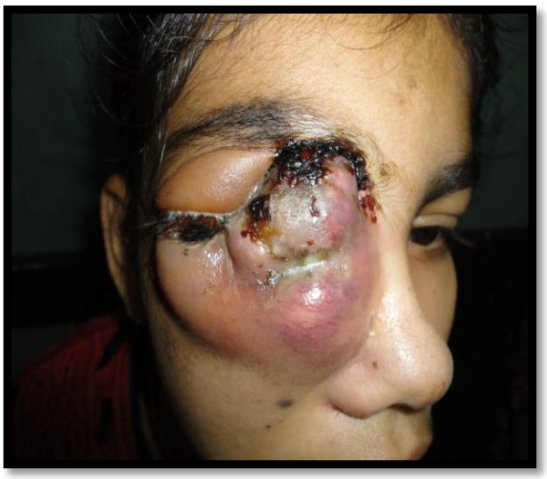

Figure 1

INVESTIGATIONS

- Routine haematological examination were within normal limits.
- X ray chest and PNS,USG abdomen revealed no abnormality.
- MRI Orbit revealed (figure 2, 3) a large multilobulated solid appearing soft tissue mass lesion seen in the right medial orbital fossa displacing the globe laterally and it is also seen extending into the underlying right half of ethmoid sinus as well as nasal cavity. Inferior it is seen extending into the right maxillary sinus. In the orbit, lesion appears to be extra conal. There is small extension into the right frontal recess also. Soft tissue component is seen extending into the right premaxillary subcutaneous soft tissue. Retained secretions are seen in

right maxillary sinus. Minimal mucosal thickening is seen in sphenoid sinus and right posterior ethmoid sinus. No abnormal blood vessels, calcific foci or fat intensity is seen. No necrotic or hemoorhagic areas are seen.

- Entire lesion measures around 5.5*5*4.8 cms. No intra cranial extension is seen. Left orbit and sino nasal region appear normal. An irregularly mixed dense well defined shadow in the retrobulbar space of the right orbit. The optic nerve could not be separated out and the lesion seemed to be adherent to the eye-ball (figure 2 & 3).

Anterior orbitotomy of right orbit was done, under G.A. and a bilobed globular mass with stalk was excised.The mass was examined his-topathologically and diagnosed to be embryonal rhabdomyosarcoma (figure 4). The patient was then subjected to radiotherapy.

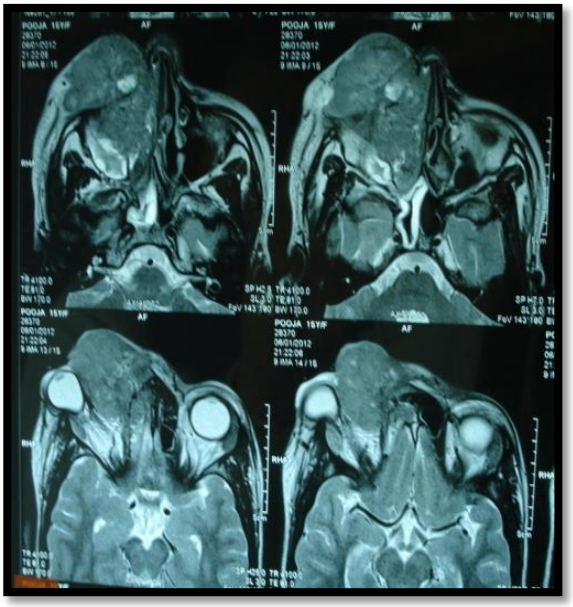

Figure 2

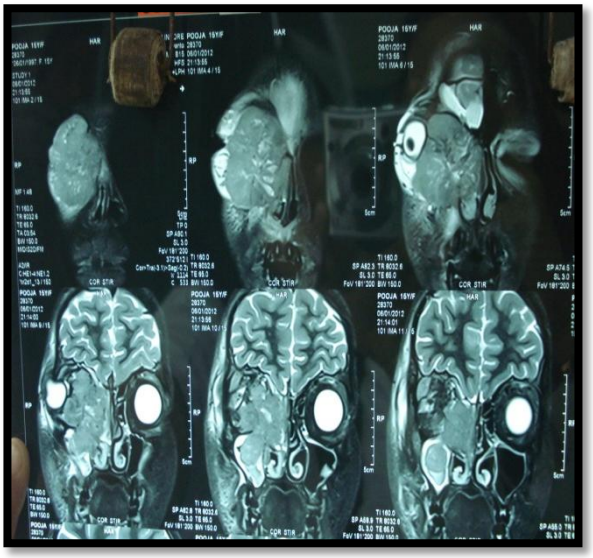

Figure 3

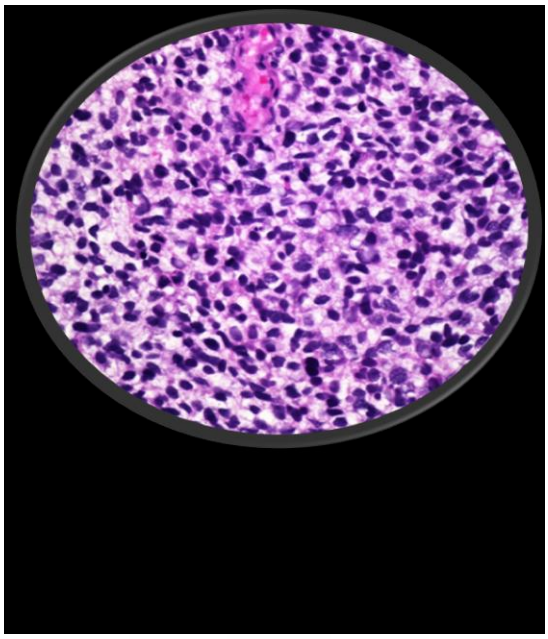

Figure 4

DISCUSSION

From the clinical point of view, rhabdomyosarcoma is of an adult and infantile form.[4] The adult rhabdomyosarcoma arises from voluntary muscle and is extremely rare since the fully mature muscle cell is not prone to malignant change. The infantile form is much more common and indeed is the most frequent malignant tumour in the orbit. It is seen at an average age of 6 or 7

years although in our case it has occurred earlier i.e. at 4 years.

From the pathological point of view several types have been described - embryonal sarcoma arising from the embryonic mesenchymal tissue either prospective muscle or undifferentiated tissue capable of heteroplastic differentiation into muscle, non striated embryonal rhabdomyosarcoma and striated embryonal rhabdomyosarcoma. [5] Reports have stressed a preponderence in males and we also found a male child to be affected.

The mode of presentation of this tumour varies considerably and in our case it presented with swelling in the right medial canthus.

The embryonal rhabdomyosarcoma consists of a loosely knit mass of primitive mesenchymal cells among which pleomorphism is a prominent feature; most of the cells are round, oval or stellate but sometimes spindle shaped. The cytoplasm is usually scanty while some have long ribbons or streamers of eosinophilic cytoplasm, often containing cross-striations. The nucleii are round or oval and are rich in chromatin; mitosis is common. The cells are frequently arranged in a loose syncytium but sometimes in the form of compact masses or in alternating bands of loosely arranged and closely packed cells.

REFERENCES

1. Porterfield J.T. and Zimmerman. L.E.: Rhabdomyosarcoma of the orbit of clinicopathologic study of 55 cases. Virchow Arch. (Patho. Anat.) 335: 329. 1962.
2. Frayer, W.C. and Enterline. H.T.: Embryonal rhabdomyosarcoma of the orbit in children and young adults. Arch . Ophthal. 62 : 203, 1959
3. Horn, R.C.: Enterline. H.T.: Rhabdomyosarcoma - A clinico - pathological study and classification of 39 cases. Cancer 11:181, 1958
4. Bard. Arch. Physiol. Norm. Path. 5:247. 1885
5. Ashton and Morgan: J. Clin. Pth. 18:699, 1965