# Rhabdomyosarcoma a rare case soft tissue tumor of extra ocular muscle of eye

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***Abstract-***
**Objective- To study a rare case soft tissue tumor of extra ocular muscle known as rhabdomyosarcoma.**
**Material and method- This is a cross sectional, noncomparative study of rhabdomyosarcoma of extra ocular muscle of eye in GSVM medical college Kanpur presented with left upper lid swelling.**
**Result- A female patient presented with left upper lid swelling with 2-month history for that CT scan and FNAC is done and patient is referring to oculoplastic unit for further management.**
**Conclusion- Rhabdomyosarcoma is a malignant mesenchymal tumor with skeleton muscle differentiation. It has mainly 3 subtypes on pathological basis alveolar (20%), embryonal (60%) and pleomorphic (20%)[1]**

***Key ward-*** **Upper lid swelling, CT scan, FNAC.**

## INTRODUCTION

Soft tissue sarcoma refers to non-epithelial tissue excluding the skeleton joints, central nervous system, hemopoietic and lymphoid tissue [1]

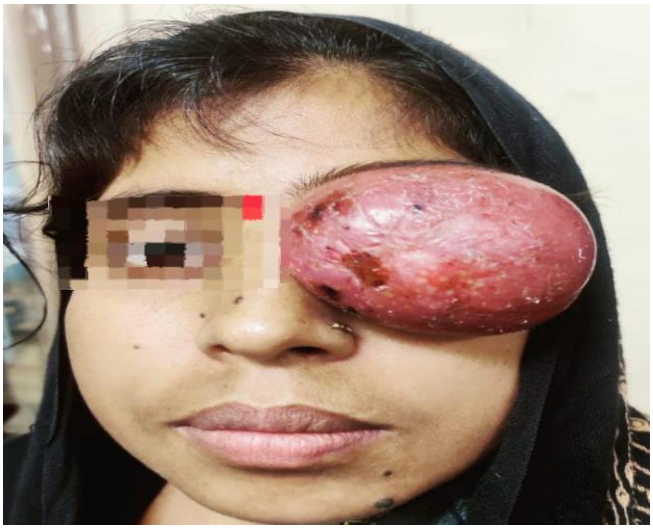
Rhabdomyosarcoma is a malignant mesenchymal tumor with skeleton muscle differentiation. It has mainly 3 subtypes on pathological basis alveolar (20%), embryonal (60%) and pleomorphic (20%)[1]. Alveolar and embryonal most common in childhood and adolescent. Pleomorphic forms arise in adults [1]. Pediatric forms arise from location that normally do not contain skeletal muscles. Embryonal and pleomorphic subtype are g1enetically heterogeneous. Alveolar rhabdomyosarcoma frequently contains fusions of FOXO1 gene to either PAX3 or PAX7 gene rearrangement marked by the presence of (2;13) and (1;13).

## INCIDENCE

Among all solid tumor malignancies, soft tissue sarcoma form <1% in adults [2]. Among these Rhabdomyosarcoma are about 3%. Head and neck (35-40 %) being the most affected site followed by genitourinary tract and extremities [3]. Orbital Rhabdomyosarcoma forms 9% of all cases of head and neck. Average age of diagnosis of orbital Rhabdomyosarcoma is 7-8 years with incidence about 4-7 cases per million. So aggressive orbital Rhabdomyosarcoma in adulthood could be a rare case. There is a slight male to female predilection with a male: female ratio of 5:3 [4].

## CASE REPORT

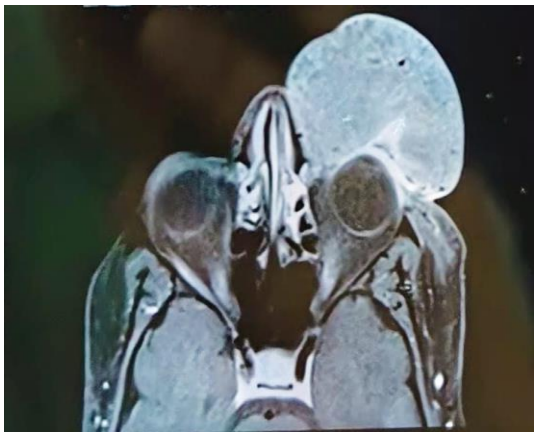
A 36-year-old female came to our side outpatient department at Department of ophthalmology GSVM medical college Kanpur with chief complaint of small hemangioma over left upper lid for which excisional biopsy was performed and it showed capillary hemangioma. But within a span of 3 months after excision that small hemangioma grew into a huge mass over left upper lid (figure-1).

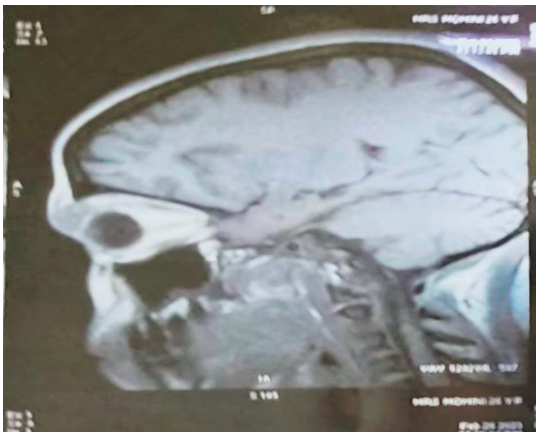


**INVESTIGATION**

Dramatic worsening of symptoms led to performing a new biopsy and computed tomography. The biopsy showed a malignant small round cell tumor consistent with orbital Rhabdomyosarcoma with embryonal type. The diagnosis was made dependent on immunohistochemistry staining using leucocyte common antigen /Desmin/ CD 99 (positive for Desmin).

CT scan showed a hyperdense mass lesion of size 6.6×5.3×5.6 cm arising from left upper eyelid, superior rectus, lateral rectus and extending to involve the left orbicularis oculi muscle, medially the lesion is abutting levator labii superioris. No bony involvement or erosion seen by the lesion. On post contrast images showing enhancement with vascular supply likely from superficial temporal artery -? Orbital Rhabdomyosarcoma? Vascular lesion. Calcified granuloma of size 3 mm without perifocal edema seen in left parietal lobe. (Figure-2 & 3).





**TREATMENT**

The patient was referred to oncology department. Aggressive Neoplasm are usually treated with surgery and chemotherapy with or without radiation therapy.[1]

As far as treatment is concerned orbital exenteration could be one of the options. But usually, it leads to poorer prognosis. So, after the diagnosis has been confirmed by excisional biopsy. The patient is given orbital irradiation later followed by chemotherapy in some patients.

From the stage of regional lymph nodes involvement up till distant metastasis radiation in the range of 4000 to 5000 cGy given for 4-5 weeks. The length and number of chemotherapeutic agents used for treatment is determined by staging. Traditionally vincristrine and actinomycin are used but among newer agents Ifosfamide and Etoposide are also beneficial [5].

**FOLLOW-UP**

After completion of treatment patient is followed initially for 3-4 months. Systematic evaluation to predict metastasis and extensive ophthalmic assessment is required:

- Best corrected visual acuity of both eyes.
- External ocular examination for dry eye, proptosis, masses, and motility disturbance of both eye
- Slit-lamp bio microscopy to detect cataract
- Ophthalmoscopy to rule out radiation retinopathy
- To detect any residual or recurrent tumor orbital CT and MRI is must.

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