

## Advanced Case of Rhabdomyosarcoma of Orbit Mimicking Orbital Cellulitis

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### Abstract

Rhabdomyosarcoma is a malignant neoplasm of primitive mesenchyme exhibiting skeletal muscle differentiation. Rhabdomyosarcoma, which can histopathologically be of embryonal, alveolar, botryoid and pleomorphic types; is a rare tumor in children, with an annual incidence of 4.3 cases per million. Pleomorphic rhabdomyosarcomas is a rare variant, occurring more commonly in adults. We report a rare case of ocular pleomorphic rhabdomyosarcoma in an 18-month-old child with atypical presentation mimicking orbital cellulitis. A provisional diagnosis of orbital cellulitis was made based on clinical and radiological examination and was planned for surgery. Histopathology confirmed the diagnosis of pleomorphic rhabdomyosarcoma. This case shows that pleomorphic rhabdomyosarcoma may also occur in children, it may mimic symptoms of orbital cellulitis, and any fast growing swelling should be carefully examined with a high degree of suspicion of this entity.

**Keywords:** Advanced rhabdomyosarcoma; Mimicking orbital cellulitis; Orbital rhabdomyosarcoma

### Introduction

Rhabdomyosarcoma is a malignant soft tissue neoplasm of skeletal muscle origin. Rhabdomyosarcoma is the most common soft tissue sarcoma in the pediatric population, accounting for approximately 5% of all childhood cancers and for about 20% of all malignant soft tissue tumors [1]. The primary sites of rhabdomyosarcoma include head and neck area (45%), trunk (40%), and extremities (15%) [2]. About 25%-35% of head and neck rhabdomyosarcomas arise in the orbit [3,4]. Based on histopathologic observation microscopically, the four major histopathologic types of rhabdomyosarcoma are embryonal (57%), alveolar (19%), botryoid (6%), and pleomorphic (1%) [5]. The embryonal type of rhabdomyosarcoma is the most common variant occurring in children, whereas the pleomorphic type more often in adults [6,7]. The occurrence of pleomorphic rhabdomyosarcoma is exceedingly rare in children. In this report, we present a case of pleomorphic rhabdomyosarcoma in the left eye of an 18-month-old male child with extension of tumour into paranasal sinuses.

### Case Report

A 18 month old male child brought by parents to ophthalmology outpatient department with complaints of mass in left eye progressively increasing in size for one and half months associated with low grade fever on and off for 15 days. Antenatal and birth history revealed nothing abnormal. On general examination, no palpable mass was felt in other parts of the body. Per auricular and submandibular lymph nodes were not palpable. On systemic examination, no lump was felt in abdomen.

Ocular examination of left eye showed proptosis, lid edema and severe exposure keratopathy which made examination of anterior and posterior segment impossible. On local examination, the swelling was tender, soft in consistency. The mass was extending from superior orbital margin to nasolabial fold vertically and nasal bridge to lateral

canthus horizontally. Complete examination of right eye was within normal limit. On the basis of history and clinical examination, provisional diagnosis of orbital cellulitis was made.

Computed tomography showed a mass displacing the globe and medial rectus laterally. Other childhood tumors were ruled out by hematological & radiological studies. Patient was planned for surgery and the biopsy of the protruding mass was sent for histopathology studies which showed spindle shaped cells with elongated nuclei, eosinophilic cytoplasm and cross striations were present. Thus a definitive diagnosis of advanced grade iii pleomorphic rhabdomyosarcoma was made. The patient was referred to department of oncology, where chemotherapy was started. Patient underwent 6 cycles of chemotherapy but did not turn up for radiotherapy. He succumbed to his illness 4 months later.

### Discussion

Rhabdomyosarcoma is a malignant mesenchymal tumor composed of cells with histologic features of striated muscle in various stages of embryogenesis [8-10]. It originates from pluripotent mesenchymal cells that have the capacity to differentiate into skeletal muscle [11]. It is the most common soft tissue sarcoma arising in the pediatric population, accounts for about 5% of all childhood cancers [12]. Rhabdomyosarcoma can occur in several sites in the body, including the ocular region. Primary sites of rhabdomyosarcoma include head and neck area (45%), trunk (40%), and extremities (15%). Orbital rhabdomyosarcoma accounts for about 25-35% of head and neck rhabdomyosarcomas and for about 10-20% of all rhabdomyosarcomas. The ocular region, particularly the orbital soft tissues, represents a major anatomic location for rhabdomyosarcoma [13-16].

Pleomorphic rhabdomyosarcoma is a rare variant, which occurs predominantly in adults aged 30 to 50 years and is rarely seen in children.

Classic clinical picture is sudden onset and rapid evolution of proptosis without history of previous trauma or sign of upper respiratory tract infections. There is often a marked lid edema,

proptosis, paraxial globe displacement, eyelid edema, conjunctival congestion, blepharoptosis and periocular discoloration. Ptosis and strabismus may also be present.

In general, most patients have an advanced disease even at the stage of initial presentation because rhabdomyosarcomas are known to show rapid growth and the patients generally tend to delay medical consultation.

Being very rapidly growing with all inflammatory signs, it may be misdiagnosed as orbital cellulitis. If rhabdomyosarcoma is suspected, CT and MRI can be used to define the location and extent of the tumor. CT is particularly helpful if tumor has caused bony destruction, although the orbital walls remain intact in most cases.



Figure 1: Clinical photograph.

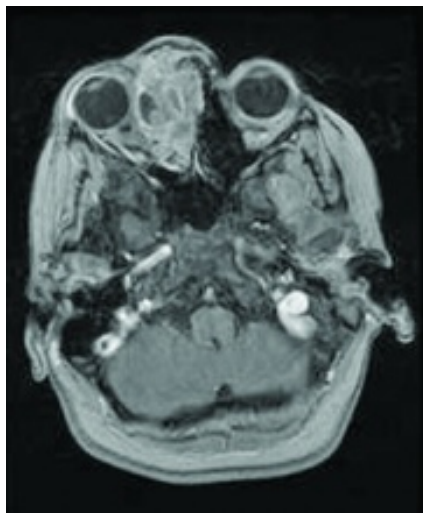


Figure 2: Computed tomography showing orbital rhabdomyosarcoma displacing the globe and medial rectus laterally.

Prognosis of rhabdomyosarcoma is relatively poor compared to that of other orbital soft tissue malignant lesions and depends on the clinical staging and the anatomical site of the tumor. Treatment of rhabdomyosarcoma consists of surgical resection, when possible, associated with chemotherapy and/or radiotherapy.

In our case, 18 month old male child presented with massive proptosis of left eye since one and half month duration and low grade

fever on and off 15 days. He had extensive orbital mass, lid edema, proptosis and tenderness mimicking orbital cellulitis. Radiological investigations showed extension of tumour in paranasal sinuses which is rarely seen because orbital tumours tend to remain localized for a long period and are usually diagnosed earlier because of proptosis.

Orbital rhabdomyosarcoma is one of the few life-threatening diseases seen initially by ophthalmologists and prompt diagnosis and treatment can save the life of the affected patient. Therefore, eye care physicians should be aware of this tumor, recognize its clinical features, and refer the patient for prompt treatment (Figures 1-3).

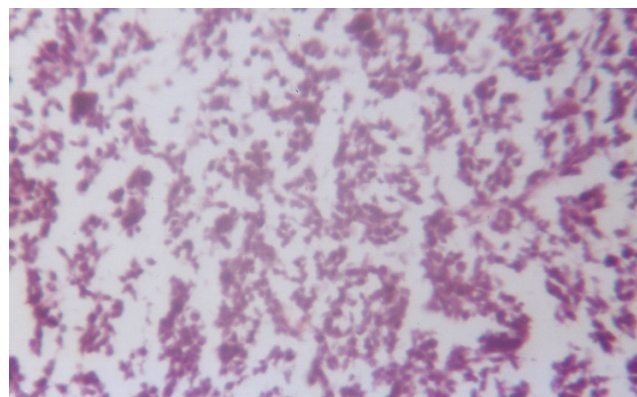


Figure 3: HP showing spindle shaped cells with elongated nuclei, eosinophilic cytoplasm and cross striations.

## Conclusion

This is a rare case of pleomorphic rhabdomyosarcoma in an 18 month old child who had signs & symptoms mimicking that of orbital cellulitis. High index of suspicion and histopathological confirmation along with prompt treatment in early stage can go a long way in increasing the survival rates of such patients.

Our case had several distinctive features including age of presentation (early onset), type of presentation (enlarging mass with tenderness mimicking orbital cellulitis), and type of tumor (pleomorphic).

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