

Rhabdomyosarcoma masquerading as orbital cellulitis

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Key words: Orbital Cellulitis, orbital tumors, rhabdomyosarcoma

Orbital rhabdomyosarcoma (RMS) is the most common orbital malignancy in children. RMS accounts for approximately 4% of the orbital masses in children.^[1] The mean age of diagnosis is 6–8 years. However, RMS might be observed at birth.^[2] Orbital RMS presents as rapid-onset proptosis, chemosis, restriction, and painful ocular movements, similar to orbital cellulitis. Diagnosis of RMS is necessary to prevent the disease from spreading and threatening patients' vision and life.

We present a case of orbital RMS that mimicked orbital cellulitis in a 2-year-old girl with rapid-onset proptosis and ocular movement restrictions.

A 2-year-old girl with rapid-onset protrusion of the right eye for 3 days was referred to our institution. The protrusion was associated with swelling of the right side of the cheek for 1 month. No history of trauma was observed, and the patient was not on any immunosuppressive drug. Initially, the patient was monitored by a pediatrician who prescribed oral antibiotics and anti-inflammatory medicines. Transient relief was obtained for 2 days. However, lid swelling and protrusion of the eye increased over time. When referred to our hospital, the child exhibited fever with periorbital swelling and proptosis of the right eye in the temporal direction with restricted ocular movements in all directions [Fig. 1]. Chemosis and hazy cornea with stain-positive epithelial defect due to exposure keratopathy were observed. The right eye pupil was semi-dilated with a sluggish response to light. Faint retinal glow was observed on indirect ophthalmoscopy. Systemic workup, including complete blood count, urinalysis, and serum electrolytes, was unremarkable.

Due to local signs of inflammation, we suspected orbital cellulitis. The patient's left eye was normal. Additionally, the patient exhibited a running nose with an upper respiratory tract infection. Therefore, the diagnosis of the right eye orbital cellulitis was confirmed. Intravenous antibiotics provided

no relief. Therefore, a multislice computed tomography scan of the orbit and brain was recommended. The scan revealed well-defined, heterogeneously enhancing soft tissue density lesion filling the retro-orbital space with the destruction of orbital walls spreading to the right maxillary sinus [Fig. 2]. The tumor size in the maxillary sinus was approximately $8.7 \times 6.2 \times 6.7$ cm³. The right temporal bone was involved without any significant surrounding edema. Complete erosive destruction of the maxillary sinus and floor walls was observed, which extended along the right orbital floor of intra-orbital and extraconal compartments [Fig. 3]. The tumor was abutting and mildly deforming the sclera posteriorly, involving the right cavernous sinus. Bony erosion involving sella turcica and the pituitary base was also discovered. The right optic nerve was normal with a few nonenhancing necrotic areas. No evidence of calcification was noted. Involvement of hard palate and nasal septum was also observed [Fig. 4].

The features suggested neoplastic etiology over infective etiology, mostly RMS. However, to confirm the diagnosis, incisional biopsy was taken. Histopathologic examination revealed the presence of elongated spindle cells with a highly eosinophilic cytoplasm and hyperchromatic nuclei suggestive of alveolar type of RMS. Due to extensive involvement of the patient's orbit and brain, the patient was referred to the radiotherapy and chemotherapy department for further management.

Discussion

RMS is the common primary orbital malignant tumor in children. The initial stage of RMS development is insidious. The most frequent primary site of RMS is the orbit. The incidence of RMS is the highest in 1–4 years and the lowest in 15–19 years of age. The classic presentation of RMS involves sudden onset and rapid progression of proptosis. Additionally, orbital cellulitis is presented with sudden-onset proptosis, lid edema, chemosis, orbital tenderness, ocular movement restriction, fever, headache, and runny nose. Our patient exhibited the signs and symptoms suggestive of orbital cellulitis. Moreover, the most common cause of proptosis in children is orbital cellulitis.^[3] Injectable antibiotics and anti-inflammatory agents did not relieve the swelling and proptosis. Therefore, the patient underwent a computed tomography scan of the orbit and brain, which illustrated a heterogeneous mass involving the right orbit's retro-orbital region, extending to the maxillary sinus. Amir *et al.*^[4] revealed that orbital cellulitis can mimic RMS. However, a high degree of suspicion is essential in RMS diagnosis as the disease can be vision- and life-threatening.

The diagnosis of RMS is usually performed through imaging and histopathology analyses. Ultrasonography has limited efficacy because limited penetration into the deeper tissues

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Figure 1: Photograph showing proptosis of right eye with chemosis and epithelial defect



Figure 3: Computed tomography post-contrast coronal image at the level of maxillary sinus, mid orbit, and cavernous sinus. Complete erosive destruction of all walls of maxillary sinus and floor, medial and lateral walls of right orbit (arrow)

can be achieved using this technique. Incisional biopsy of the lesion showed presence of elongated spindle cells with a highly eosinophilic cytoplasm and hyperchromatic nuclei suggestive of alveolar type of RMS.

The closest differential diagnosis in the aforementioned age group includes retinoblastoma optic nerve glioma, leukemia, and neuroblastoma. Retinoblastoma presents initially with the white reflex in the pupillary area, with dilated pupil and raised intraocular pressure. Ultrasonography of the eye reveals calcification of the tumor. Optic nerve glioma presents with diminished vision, proptosis, dilated pupil, strabismus, disk pallor, or edema. Our patient had rapidly progressive proptosis, lid edema, and chemosis of the conjunctiva with restricted ocular movements. Therefore, the initial diagnosis of orbital cellulitis was made. However, computed tomography and histopathologic examination helped in solving the mystery.

Acute myeloid leukemia in the form of granulocytic sarcoma (chloroma) may present as unilateral proptosis. The clinical manifestations include chemosis, lid edema, proptosis, extraocular muscle palsies, diplopia, and retinal hemorrhages.^[5] The diagnosis of the condition may be made on bone marrow

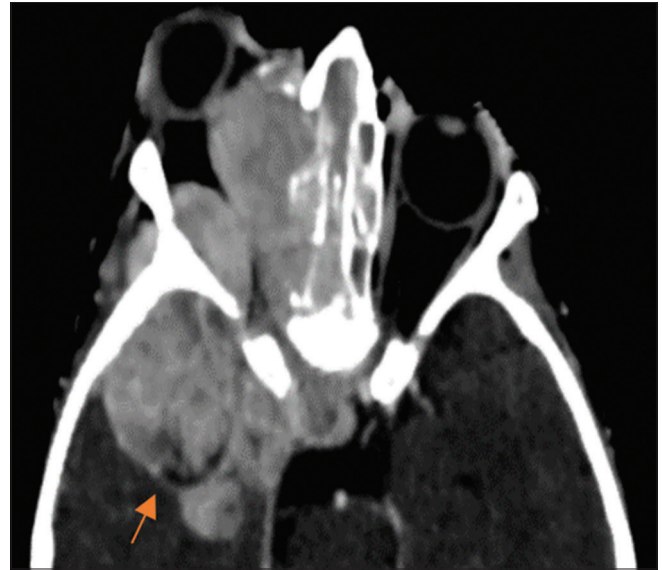


Figure 2: Computed tomography post-contrast axial image at the level of mid brain, right orbital apex, and uvula showing well-defined, heterogeneously enhancing soft tissue density lesion filling the retrobulbar region of right orbit and involving right temporal fossa (arrow)

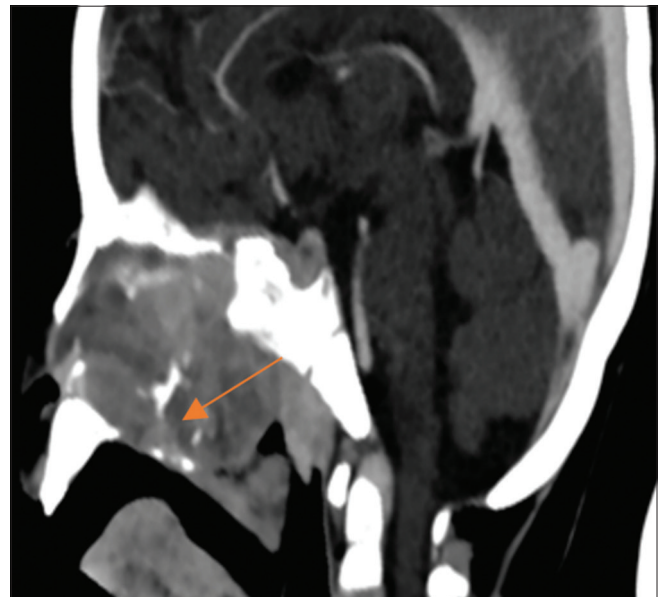


Figure 4: Computed tomography post-contrast sagittal image at the level of right orbital apex showing erosion of hard palate and bony nasal septum (arrow)

biopsy, which shows leukemic cells. Systemic antileukemic chemotherapy helps to resolve the condition.

Another differential diagnosis could be orbital neuroblastoma. Neuroblastomas are neuroendocrine tumors that can originate anywhere in the sympathetic nervous system and are most commonly found in one of the adrenal glands. The symptoms and signs of orbital neuroblastoma are not specific. Clinical features, like periorbital ecchymosis and proptosis, in combination with radiological analysis and histological features are important for the diagnosis of orbital neuroblastoma.

The treatment regimen for RMS is multimodal that comprises multidrug chemotherapy, radiotherapy, and excision of growth in early stages.^[6] However, completely excising the mass is often tricky due to the tumor's proximity to the orbital structures and the high risk of complications.^[7]

RMS may masquerade as orbital cellulitis. The present case study demonstrates that an acute onset and rapidly progressing proptosis must be aggressively investigated and immediately referred for optimum management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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