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Case Report

# Orbital immature teratoma: A rare entity with diagnostic challenges

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## Abstract

Childhood orbital teratomas are congenital lesions that presents most often at birth with progressive, severe unilateral proptosis. Due to the rarity of such tumors, the diagnosis is often missed with delay in the patient's management. We are presenting a unique case of an immature right orbital teratoma with extensive growth in a full-term newly born baby boy. In this case report, we provide description of the clinical findings, initial misdiagnosis and the eventual management with review of similar reported cases.

**Keywords**: Orbit, Proptosis, Congenital, Immature teratoma


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## Introduction

Teratomas are congenital germ cell tumors derived from the three germinal layers. They are classified into mature or immature teratomas according to the degree of cell differentiation. Mature or well-differentiated teratoma tends to act benignly, whereas, immature teratoma is characterized by the presence of neuroectodermal elements and has the tendency to be malignant.[1] The sacro-coccygeal region is the commonest site for such lesions.[1,2] Head and neck location is considered the second most common site contributing for 5–14% of cases in infancy.[2] Females have predominance compared to males with a ratio of 2:1.[2] Teratoma of the orbit is believed to be a rare condition.[2,3] Hence, we are presenting a case of immature orbital teratoma where the diagnosis was not suspected clinically at initial presentation and the case was referred with the provisional clinical diagnosis of lymphangioma.

## Case report

A full-term baby boy of 3.2 kg was born with right severe proptosis (Fig. 1A). Details of the pregnancy, birth and family history were unremarkable. On examination, there was a huge orbital lesion with trans illumination causing completely prolapsed right globe (Fig. 1B). The anterior chamber was shallow with conjunctival chemosis. The nasal part of the cornea showed moderate haze and temporal opacification. The mass was hard in consistency and fixed to the globe. The left eye was normal. Computerized tomography (CT) scan showed a huge right orbital mass with a compressed elliptical globe, which was pushed antero-laterally. The mass was hypodense with fluid density and a localized area at the temporal aspect, which appeared multi-lobulated. Large dense linear and clustered calcifications were also noted. The Magnetic Resonance Imaging (MRI) showed partially hyperintense lesion posteriorly and along both transverse sinuses. No

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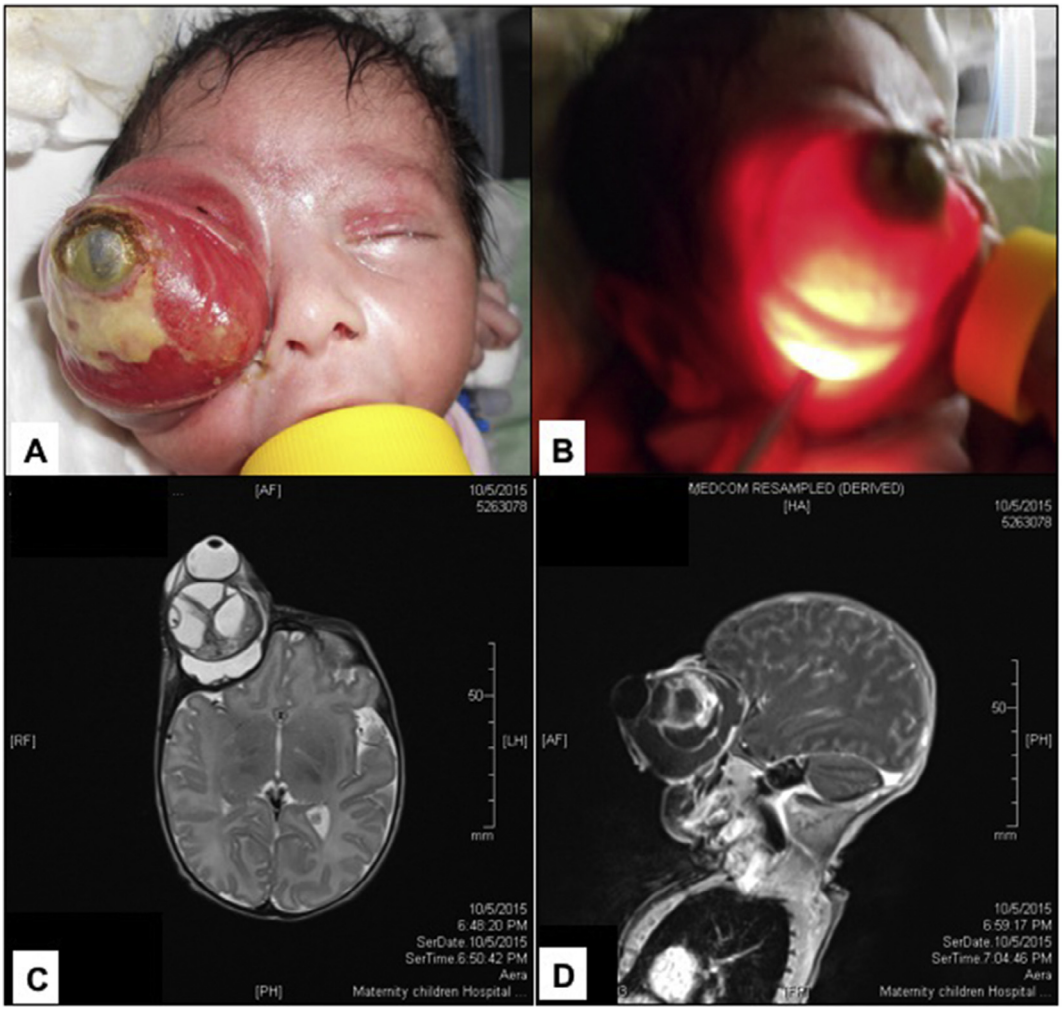


**Fig. 1.** A: The clinical appearance of the massive right side proptosis with corneal exposure keratopathy. B: The trans illumination of the right huge orbital mass showing cystic areas. C: Axial magnetic resonance T-2 weighted image showing the mult-loculated mass with massive proptosis of the right globe. D: Sagittal T-1 weighted post-contrast image showing right orbital mass with focal hyper-intense area posteriorly.

definite focal cerebral lesions were detected with normal appearing ventricular system (Fig. 1C and D). The baby was initially seen at a local hospital in Al-Qassim region and was referred to King Abdulaziz University Hospital (KAUH) in Riyadh for further management. The ultrasound demonstrated multiple hemorrhagic cystic spaces filling the orbit, pushing the globe anteriorly and replacing all orbital ocular muscles suggestive of lymphangioma. The patient underwent left eye examination under anesthesia and debulking of the right orbital lesion with salvaging of the globe (Fig. 2A). The specimen consisted of a well demarcated multilobulated mass measuring mm × mm with apparent cystic areas (Fig. 2B). The patient was seen few months postoperatively with no evidence of recurrence (Fig. 2C). The histopathological examination showed: a large encapsulated tumor mass consisting of different tissues from all 3 germline components, which included respiratory epithelium, secretory epithelium, bone, cartilage, skeletal muscle, and neural tissue (Fig. 3A–C). The middle sections of the mass showed immature neural tissue with mitotic figures in focal areas (Fig. 3D–F). Based on the histopathologic appearance, the lesion was diagnosed as an immature teratoma with grade 2.

## Discussion

Teratomas are congenital germ cell tumors that contain components of the three germinal layers and are classified into mature or immature teratomas according to their degree of cell differentiation. Several theories about the pathogenesis of these extra-gonadal tumors have been recognized. One theory suggested an abnormal migration of these germ cells resulting in further proliferation in extra gonadal location. Another theory supported the thought that these cells arise from normal somatic cells with complete genetic coding.[2] These extra gonadal tumors are classified histopathologically into mature teratoma, which is composed of fully differentiated adult tissue of several types, and immature teratoma, which is composed of fetal and partially differentiated tissue.[4] Childhood orbital teratomas are usually benign and tend to contain both mature and immature elements. Multiloculation, cyst formation, calcification, ossification, fat deposition and admixture of tissue are indicators of benign teratoma.[3] Clinically, orbital teratomas are usually detected early in life or more precisely at birth due to their obvious presentation.[2] However, the diagnosis is often missed in spite

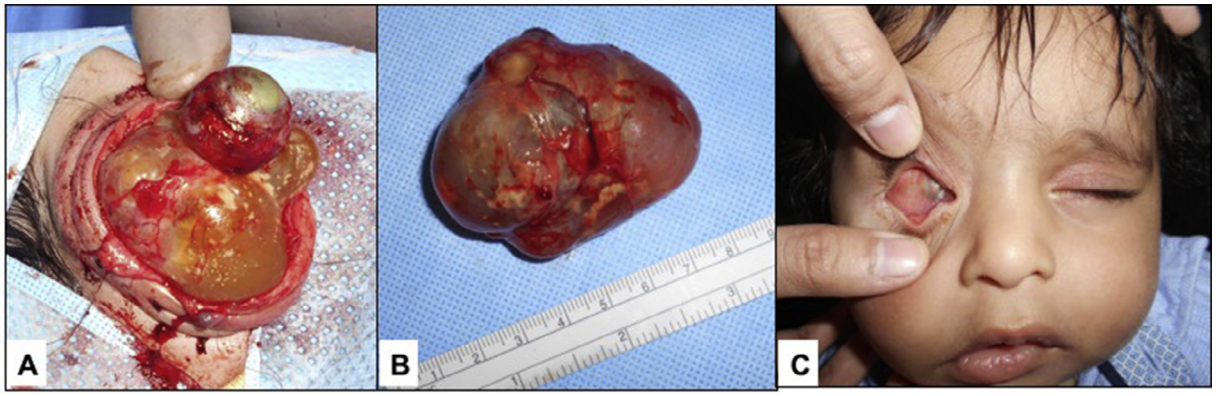


**Fig. 2.** A: Intra-operative appearance of the isolated capsulated right orbital mass for excision. B: The gross appearance of the mass excised. C: The appearance of the right salvaged globe several months after excision.

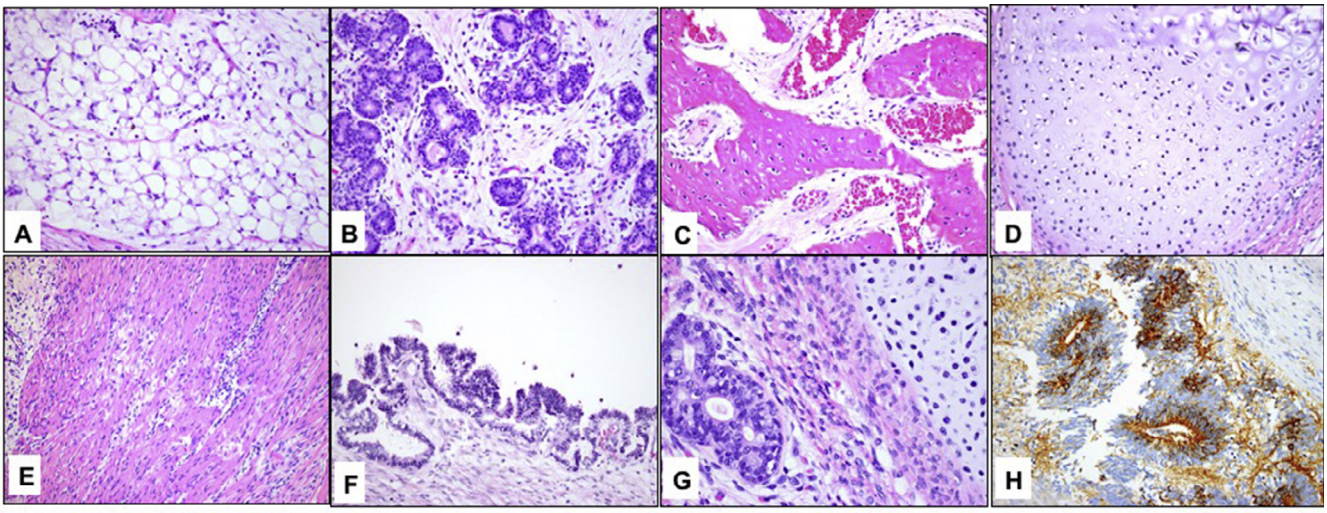


**Fig. 3.** Several elements of the teratoma in this case demonstrating adipose tissue, glandular structures, bone, cartilage, smooth muscle and respiratory epithelium in A through F (Original magnification× 200 Hematoxylin and Eosin). G: The primitive neuroepithelium of the immature teratoma with adjacent cartilage (Original magnification× 400 Hematoxylin and Eosin). H: The neuroepithelial component expressing synaptophysin with rosette-like configuration (Original magnification× 200).

of the alarming proptosis in such cases resulting in the delay in diagnosis and management leading to significant morbidity and loss of a useful vision on the affected side which was the case in our patient. Similar cases to ours with huge orbital teratomas have been reported. Ogun in 2012 reported a case of orbital immature teratoma presenting in the second day of birth and was almost identical to our patient in terms of the clinical presentation and radiological appearance.[5] The presence of a rapidly enlarging orbital mature teratoma masquerading as orbital cellulitis has been also reported to cause confusion and delay in the diagnosis.[2] Orbital teratomas might either originate primarily in the orbit or could be secondary as intracranial extension from another nearby location. Firat and his group reported a 3-day old female who presented with feeding difficulties accompanied with breathing disorder. Her examination showed an intra-oral lesion that extended to the left orbit through the left maxillary sinus and nasal space. The histopathological diagnosis in their case was grade 3 immature teratoma based on the presence of immature neuroepithelial cells, chondroid and osteoid elements.[4] Careful histopathological examination of all tissue submitted is essential for diagnosis with special attention to the surgical margins in immature teratomas and the presence of non-germ cell malignancy (such as neuroblastoma), which may affect the decision for adjunctive chemotherapy or radiation.[5] The tissue in our case consisted mostly of mature elements with focal areas only of immature neuro-epithelial elements not reaching the margins of excision.

Immediate management is preferable in order to avoid rapid growth resulting in necrosis, surrounding tissue destruction, hemorrhage or even rupture.[2,4,6] In cases of mature teratoma, complete surgical resection is advisable.[1–4,6] Adjunctive chemotherapy or radiotherapy have been tried but were believed to be ineffective especially in high grade immature teratomas.[1,2,4] Oculoplastic intervention is needed in advanced cases to maintain adequate cosmetic outcome.[6] Several factors might influence the prognosis of the extragonadal tumors including size and extension of the mass, age, gender, histopathological components and degree of tissue differentiation.[1,2,5]

In conclusion, orbital teratomas should be considered in the diagnosis of a rapidly progressing unilateral proptosis in newborns. Careful histopathological examination of the whole specimen is essential to classify the type and the grade of the teratoma and to identify the rare occurrence of non-germ malignancies. Finally, early surgical intervention aiming

at complete surgical excision is recommended with possibility of vision preservation.

## Conflict of interest

The authors have no financial disclosures to declare or conflict of interest in relation to this case.

## Compliance with ethical standards

This case study was performed in accordance with the ethical standards of the institutional and national research Human Ethical Committee (HEC) and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. IRB/Ethics Committee has ruled that approval was not required for the study. However, a General Informed Consent was obtained from the legal guardian of the child.

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