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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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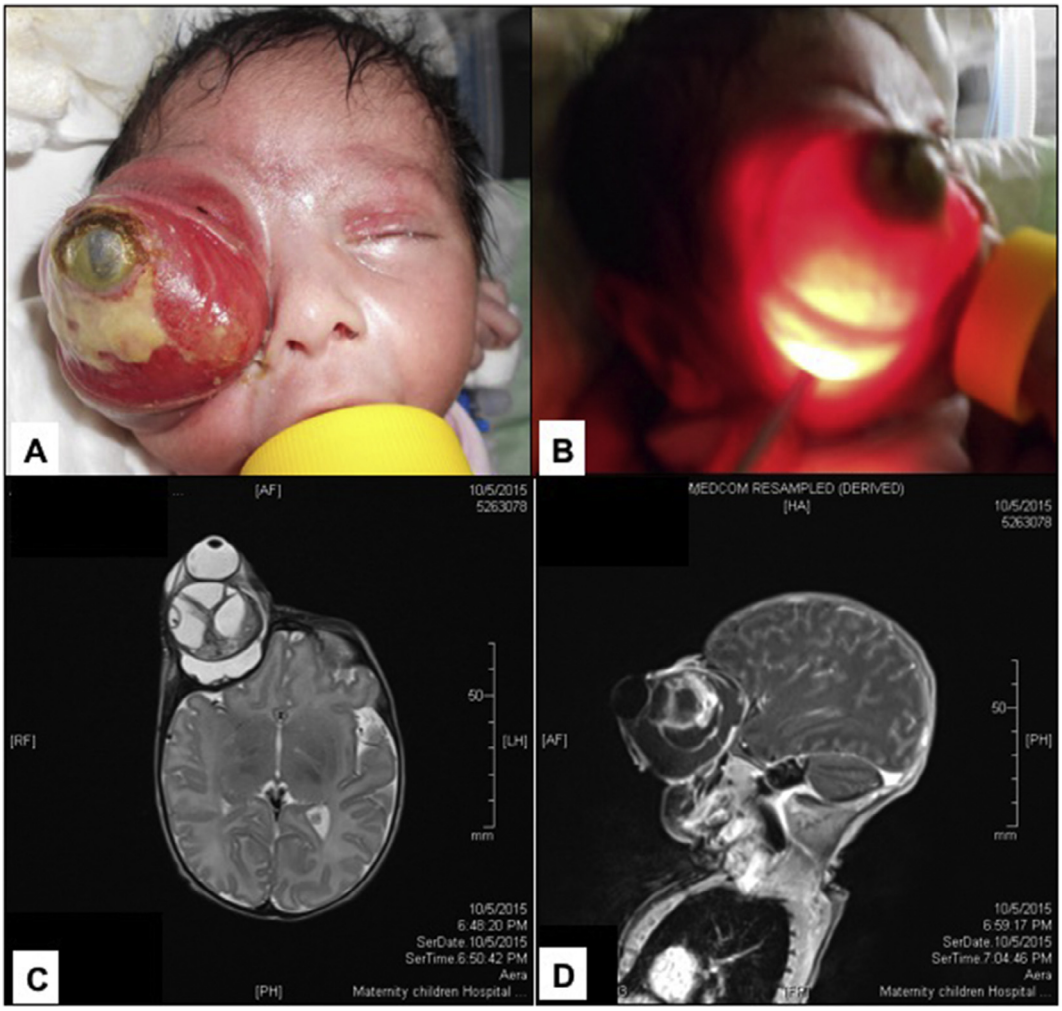

Fig. 1. A: The clinical appearance of the massive right side proptosis with corneal exposure keratopathy. B: The trans illumination of the right huge orbital mass showing cystic areas. C: Axial magnetic resonance T-2 weighted image showing the mult-loculated mass with massive proptosis of the right globe. 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