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Case Report

A Case Report on Intraorbital Extraconal Dermoid Cyst

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ABSTRACT

Orbital dermoid cyst represents 3 to 4% of all primary orbital tumors. The intraorbital location is relatively rare and it commonly occurs at childhood. We reported the case of a 4-year-old male child with an intraorbital dermoid cyst, who presented with progressive left proptosis, which had noticed 6 months earlier. Visual acuity, ocular motility and fundoscopic examination were normal. Magnetic resonance Imaging (MRI) revealed an extraconal orbital mass situated on superior & mildly posterior aspect of orbit and which was iso-to-hypointense in T1, hyperintense in T2, restriction of diffusion in DWI, hyperintense signal on ADC (Shine through) and no enhancement after giving contrast. The tumor was completely excised via a left fronto-orbital access. Histopathologically, the tumor was confirmed as dermoid cyst.

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Introduction

Dermoid cysts are squamous epithelium-lined sacs that result from abnormal migration of ectodermal cells. These cysts are composed of keratinized stratified squamous epithelium, have a fibrous capsule, and also have skin appendices like sweat glands, sebaceous glands and hair follicles [1]. Both dermoid and epidermoid are lined by keratinized stratified squamous epithelium and have a fibrous wall but A dermoid has dermal appendages, whereas an epidermoid has none [2]. Orbital dermoid cyst represents 3 to 4% of all primary orbital tumors. The intraorbital location is relatively rare [3]. Dermoid cysts are common orbital masses in childhood. Most cysts are present at birth or in the first 3 months of life. Patients usually present with mass lesion or proptosis [4]. Inflammatory reaction in surrounding tissues as the result of leakage of cyst content may rarely be the reason for admission. They should be surgically removed at ages 1 - 2 as they may enlarge and lead to deformation in orbital bony structures and erosion [5].

Good quality imaging (MRI) plays a major role in the etiological diagnosis, providing a precise analysis of the location of the lesion, its components, and its effects on adjacent and nearby structures, as well as in planning the surgical approach [6].

Case Report

A 4-year-old male child presented with progressive left proptosis, which had noticed 6 months earlier. Visual acuity, ocular motility and fundoscopic examination were normal. Magnetic Resonance Imaging (MRI) revealed an extraconal intraorbital dermoid (2.1 x 1.75 x 1.75 cm) situated on superior & mildly posterior aspect of orbit and which was iso-to-hypointense in T1, hyperintense in T2 (Figure 1), restriction of diffusion in DWI, hyperintense signal on ADC (Shine through) and no enhancement after giving contrast (Figure 2). The tumor was completely excised via a left fronto-orbital access (Figure 3). Histopathologically, the tumor was confirmed as dermoid cyst. Postoperative CT (Computerized Tomography) scan revealed no residual dermoid cyst (Figure 4).

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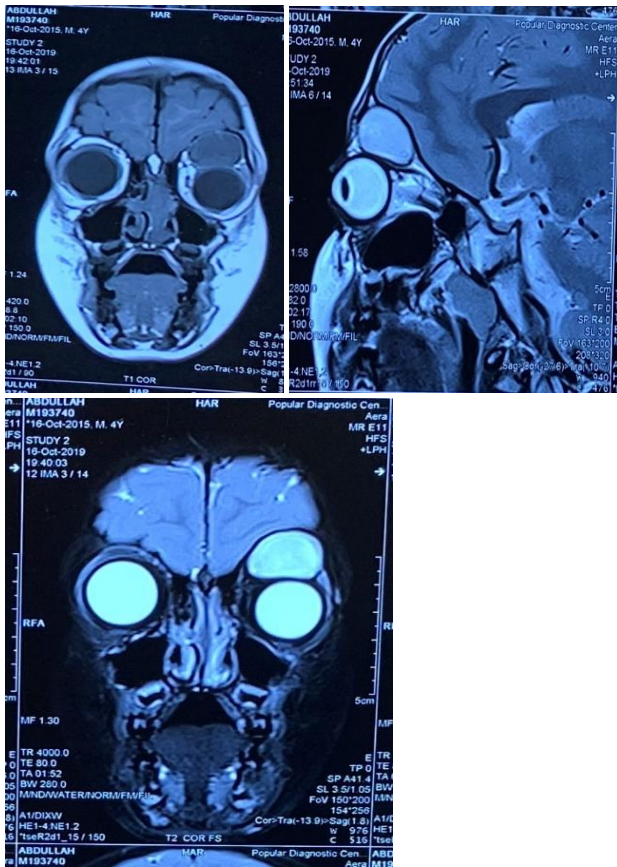


Figure 1: T1 weighted coronal & T2 weighted sagittal & coronal MRI showed left Intraorbital extraconal dermoid.

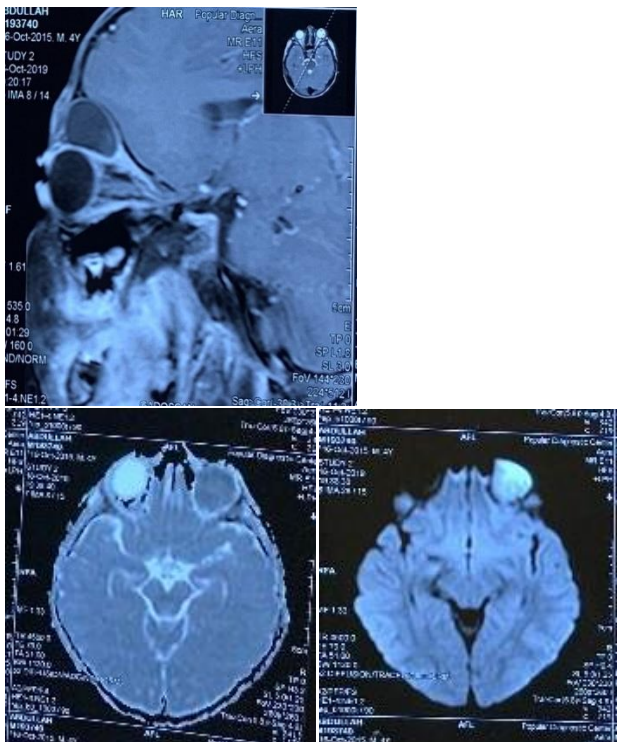


Figure 2: MRI contrast sagittal & DWI & ADC showed left intraorbital dermoid.

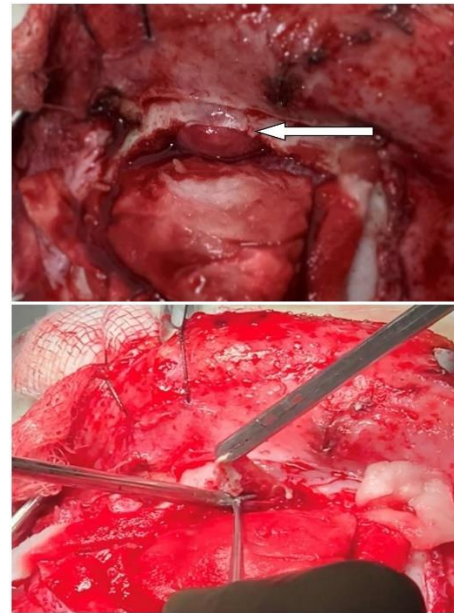


Figure 3: Per-operative view of dermoid tumor.



Figure 4: Postoperative CT scan showed no residual dermoid cyst & present status of the patient.

Discussion

Dermoid cysts are abnormally located masses arising from abnormal involution of superficial ectoderm during closure of sutures between bones [1]. This cyst is the commonly used clinical term for benign cystic teratoma, derived from sequestration of surface ectoderm into underlying mesenchyme along embryonic lines of closure [2].

Dermoid cysts occur predominantly in children, with few reported cases in the literature in adults [7]. They are presented in more superficial or periorbital forms with precocious clinical presentation and diagnosis, as well as in deeper or intraorbital forms, which usually progress for a long period until clinical manifestations arise and the diagnosis is established [8]. When found in the intraorbital region, they mainly occupy the upper

lateral quadrant of the orbit, and its clinical presentation depends on the exact location and size of the tumor [9]. They are usually slow-growing lesions, but they can progress more rapidly if they affect people during adulthood. The deepest forms of the dermoid cyst commonly manifested with a progressive proptosis [8]. In some cases, the cyst may compress the optic nerve, impairing the visual acuity of the affected side, or manifest itself also associated with inflammatory complications or hemorrhagic complications [10].

Image exams such as CT (Computed Tomography) and Magnetic Resonance (MRI) help in the diagnosis of dermoid cysts. Computed tomography (CT) usually performed preoperatively in order to determine the presence of intracranial extension of cysts and orbital bone defect. Magnetic resonance imaging (MRI) is beneficial for detection of cyst content and differential diagnosis. CT images usually present the cyst as hypodense lesions and not highlighted by contrast, and may present calcifications; in MRI, it commonly presents hyposignal in T1 and hypersignal in T2, restriction of diffusion in DWI & hypersignal in ADC, characteristics that were observed in the reported case; in both exams, it may show evidence of the presence of oily contents [11]. Diagnostic confirmation occurs through histopathological examination of the lesion, that characterizes a laminated pavement epithelial wall and a heterogeneous content with dermal appendages that also observed in our case [12].

The principle of treatment in dermoid cyst reported in the literature is the complete removal of the mass [3, 6]. Traditional treatment is excision of the mass with an incision over the mass. After this incision, long-standing erythema and sometimes a bad looking sunk scar may remain due to thickness of the skin in this area. Scarring occurs more in children as the skin is tight in children. Incisions through, below or above the brow are used to remove these cysts. Although the scar incision is hidden in the brow in this incision, alopecia and deformity may develop [13]. The best option for deep intraorbital dermoid cyst, the cyst extending into frontal sinus or cranium is bicoronal. When the lesion is intraorbital, the procedure involves complex techniques, due to the distinct anatomy and fragility of the orbital structures and depends, among other aspects, on the location and extent of the lesion, as well as the experience of the surgeon who performs the procedure [12]. In the case reported, the chosen access was left fronto-orbital access followed by osteotomies on the orbital roof. Similar access was used in the approach reported by Taha *et al.* [3].

Cyst rupture is a complication that should be avoided mostly. Cyst content should be removed from wound site by washing with abundant saline solution [8].

Conclusion

Intra-orbital dermoid cysts are rare lesions, which results in a few case reports regarding the approaches of these lesions. Thus, it is essential to add multicenter knowledge and to establish more holistic approaches for

these neoplasms and satisfying the surgical demands to guarantee the cure.

Funding

None.

Conflicts of Interest

None.

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