Short communication

Orbital giant conjunctival epithelial primary cyst

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ARTICLE INFO

Article history:
Received 5 October 2011
Accepted 8 April 2012
Available online 5 June 2013

Keywords:
Cyst
Conjunctival epithelial
Primary
Giant
Orbit

ABSTRACT

Case report: A 15-year-old male, with no previous traumatic or surgical ocular injury, presented with a right eye proptosis and inferior displacement which began 4 months earlier. Ultrasound and CT showed a cystic lesion of 3.5 cm × 2 cm in the superior orbit. Surgical resection was performed by transconjunctival orbitotomy. The histopathology examination showed a cyst with nonkeratinized epithelium, and without goblet cells. After 10 years of follow-up, there have been no new lesions.

Discussion: Conjunctival primary orbital cysts are very uncommon. They are usually of small-moderate size; giant cysts are exceptional. Diagnosis by imaging is essential to establish the differential diagnosis and surgical treatment.

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Quiste conjuntival epitelial gigante primario orbitario

RESUMEN

Caso clínico: Varón de 15 años de edad, sin antecedente traumático o quirúrgico, que presenta proptosis con desplazamiento inferior del globo ocular derecho de cuatro meses de evolución. La ecografía y la TAC muestran una lesión quística de 3,5 × 2 cm. Se reseca mediante orbitotomía transconjuntival. La histopatología muestra un quiste con un epitelio no queratinizado sin células caliciformes. Después de 10 años de evolución no ha recidivado.

Discusión: Los quistes epiteliales conjuntivales primarios son muy infrecuentes. Suelen ser de pequeño-moderado tamaño; los gigantes son excepcionales. El diagnóstico por imagen es fundamental para el diagnóstico diferencial y el tratamiento quirúrgico.

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Introduction

Cystic lesions account for 8% of adult orbital tumors. Dermoid cysts are most frequent while conjunctival cysts are comparatively exceptional in the orbit. These can be primary or more frequently secondary (67%) to the implementation of conjunctival tissue in the orbit, either due to trauma or iatrogeny, derived from surgery due to strabismus, enucleation or retinal surgery.

Only 27 cases of primary orbital conjunctival cysts have been previously described.

Case report

Male, 15, without ocular traumatic or surgical history, exhibiting proptosis with inferior displacement of the right ocular globe and slight inflammatory reaction beginning 4 months back.

Exploration revealed a superior orbit tumor protruding at the level of the sac fundus having a cystic translucent appearance with serous contents, without vascularization and a size of 3.5 cm × 2.5 cm (Fig. 1), as well as internal conjunctival symblepharon. Echography revealed a round and capsulated cystic mass located between the upper eyelid elevator and upper rectus muscles, reaching the equatorial level from the upper orbital edge (Fig. 2A). An orbital CAT scan revealed a cystic lesion with a thin and well-defined line and hypo-intense content (Fig. 2B), which discarded a dermoid cyst, lymphangioma or vascular tumor and indicated possible retention cyst. The possibility of a hydatid cyst was discarded upon reception of the normal results of serology, latex, and abdominal and thoracic echographies.

Trans-conjunctival antero-superior orbitotomy was performed removing the cyst in toto, which exhibited a thin wall and was translucent and comprising serous liquid content (Fig. 3).

The anatomopathological study revealed a non-keratinized epithelial wall with mucin-excreting cup-shaped cells without the presence of dermic elements, which allowed for a diagnosis of primary and idiopathic simple conjunctival inclusion cyst (Fig. 4).

After 10 years follow-up the patient did not exhibit relapse or developed a new lesion.

Discussion

The pathogenesis of orbital conjunctival cysts is unknown. A postulated cause is excessive invagination of the caruncular epithelium or the fornix during embryonic development similar to the pathogenesis of dermoid and epidermoid cysts. Other authors have considered the possibility of the sequencing of ectodermal pluripotential superficial cells with the ability to differentiate in one or more epithelial types.

At the histopathological level, dermoid cysts comprise keratinized stratified squamous epithelium with cutaneous appendices, while in conjunctival cysts the epithelium is squamous but not keratinized, generally without cutaneous appendices although in some cases said appendices can be present in the walls, or mucin-secreting cup-shaped cells.

The age of presentation of said cysts varies from birth up to 70 years of age and in patients over 40 it appears in only 25% of cases. Previous evolution time can range between several months up to 60 years and the typical clinic consists in a painless cystic mass of small-moderate size which can associate proptosis and ocular globe displacement although generally extrinsic muscle motility is respected as well as the bone structure.

The most common location is in the supero-nasal region, followed by the supero-temporal region where bone involvement cases have been described.

Fig. 1 - Cyst protruding through the upper right palpebral sac, with translucent wall and serous content.

Fig. 2 - (A) Echography and (B) orbitary computerized axial tomography demonstrating the existence of a cyst in the right upper orbit with hypotense content, thin wall and large volume.
As primary conjunctival cysts are typified as non-keratinized epithelium, histopathological diagnostic mistakes with mucoceles can occur\(^1\) which includes bone involvement with communication to paranasal sinuses.

With rare exceptions, orbital conjunctival cysts are masses of thin walls and low pressure which generally do not induce significant mechanical alterations.\(^2\) However, there are cases in which a recurring conjunctival cyst has eroded the bone structure and produced visual symptoms as well as spontaneous hemorrhage which increases orbital pressure and associates inflammation and pain.\(^{1,4}\) This demonstrates its potential capacity to produce anatomic and functional alterations.

The differential diagnostic of orbital cystic lesions is established between lesions that characteristically have a hyper-dense capsule image in its central content, in contrast with other orbital tumors. The diagnostic is established between epidermoid cysts, mucoceles, hydatic, hematic, cholesteatoma and conjunctival (or inclusion cysts). Data such as age, location, history of trauma or surgery or sinusitis are useful for correct diagnostic, with imaging tests being essential for circumscribing and treating the lesion.

Small asymptomatic tumors can be managed with a conservative approach by means of follow-up, whereas large and symptomatic tumors require surgery and generally exhibit good post-surgery evolution without recurrences.\(^3\)

**Conflict of interests**

No conflict of interest has been declared by the authors.

**REFERENCES**