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Short communication

Dermoid cyst in childhood, diagnosed as ptosis

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A B S T R A C T

Clinical case: A four year-old boy, diagnosed of palpebral ptosis since he was 2 by his paediatrician. At the initial ophthalmological review he had asymmetrical upper eyelids. In the follow-up a mild ocular hypotrophy appeared in his left eye and an increasing astigmatism, which made us suspect an orbital disease. The MRI confirmed a mass, compatible with a dermoid cyst.

Conclusion: Due to the slow growth of these tumours, it is only with clinical follow-up and the aid of imaging techniques that we may achieve the diagnosis and offer a definitive treatment with surgical extirpation.

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Quiste dermoide en la infancia bajo el diagnóstico de ptosis

R E S U M E N

Caso clínico: Niño de 4 años diagnosticado por su pediatra de ptosis congénita en ojo izquierdo a los 2 años. En su valoración oftalmológica inicial, únicamente se aprecia discreta asimetría entre los párpados superiores. Es en consultas posteriores cuando aparece discreta hipotropía del ojo izquierdo, y un astigmatismo creciente que hacen sospechar sobre patología orbitaria. La RMN confirma la presencia de una masa compatible con un quiste dermoide. **Conclusión:** Debido al crecimiento lento de estos tumores, solo un seguimiento de la clínica y la ayuda de pruebas de imagen llevará al diagnóstico y al tratamiento definitivo mediante extirpación quirúrgica.

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Palabras clave:

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Introduction

Orbital tumors are infrequent in children, with dermoid cyst being the most common. It constitutes 3-9% of orbital

masses¹ and can appear in any part of the body, the orbit being a frequent area of expression.

This short paper presents the case of a 4 year-old child who at age 2 had been diagnosed by his pediatrician with

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palpebral ptosis with slight facial asymmetry. He was referred for ophthalmological assessment. In regular visits and due to the progressive development of the mass an orbital lesion was suspected and imaging studies were requested.

Clinical case

Patient, age 4, referred to the Ophthalmology practice in June 2008 by the Pediatrics area, with a diagnosis of left eye congenital ptosis which, according to the parents, became evident at age two. They also related that the child tended to rub that eye frequently. The mother's pregnancy developed normally and the child was born with a Caesarian intervention within term. The only relevant history is a coagulation problem of the mother during the pregnancy, which was treated with subcutaneous heparin.

The exploration revealed a discrete asymmetry between upper eyelids, with the inside third of the upper left eyelid discretely lower. No masses were palpated in that area. The visual acuity is of 0.8 in both eyes with the Snellen E test. Normal ocular motility, negative Cover test. Ocular fundus without alterations.

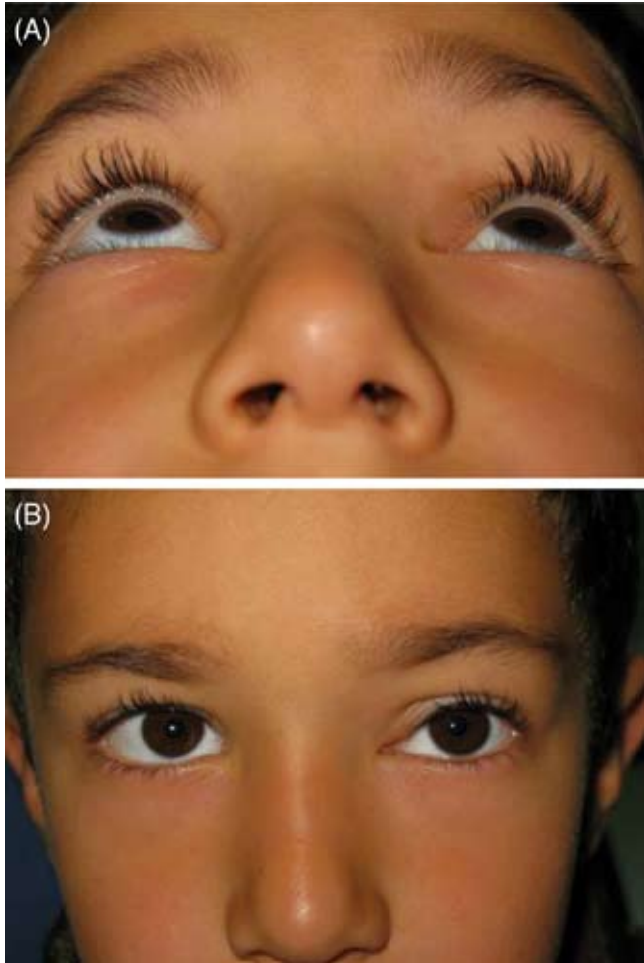


Figure 1 – A: Supraduction view. B: Front view. Both views evidence asymmetry between both upper eyelids. Left eye in discrete hypotropia.

The child was brought for a further assessment five months later and referred itching in the left eye. The new exploration revealed discrete bulging in the internal third of the left eye upper eyelid. No clear motility limitation was evident in said eye but it seems to be in slight hypotropia (fig. 1). Rest of exploration normal. A differential diagnostic is considered between motility alteration or orbital mass. It was decided to request an assessment by pediatric Ophthalmology and imaging technique is requested.

In the children's Ophthalmology practice one week later, the child exhibited a visual acuity without correction of 1 in right eye, and 0.7 in left eye which, with 1.25 at 40°, reaches the unit. The Cover test was negative in the primary position with discrete limitation in adduction and elevation, stereopsis normal and ocular fundus without alterations. Palpation gave

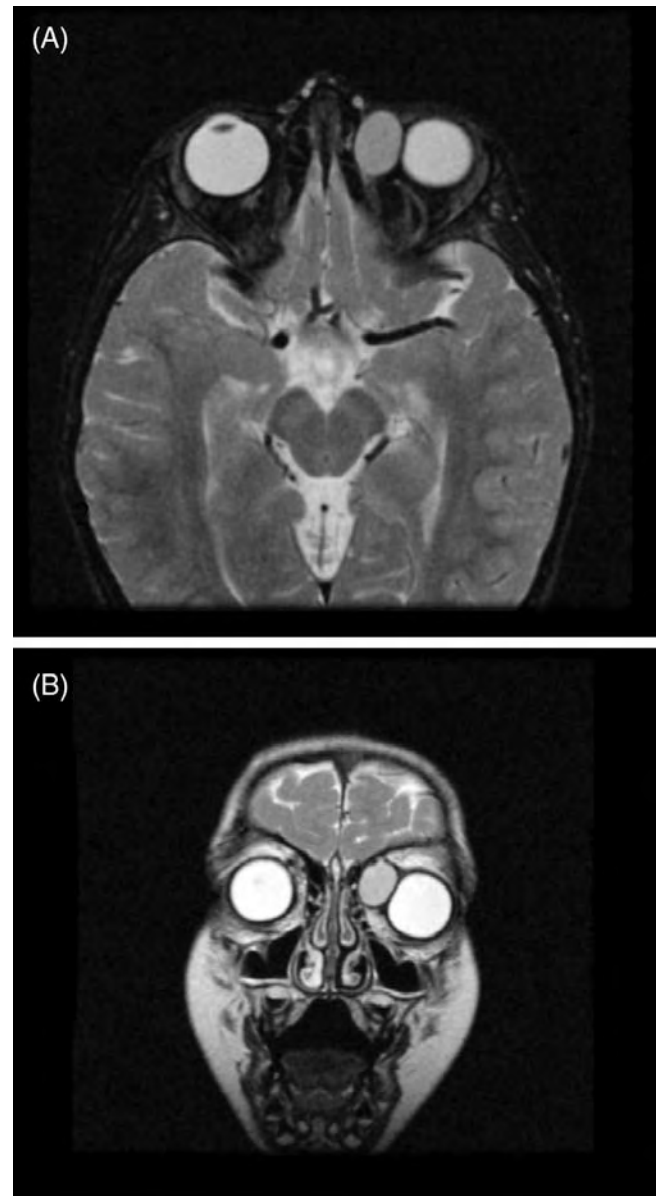


Figure 2 – A: NMR axial section in T2. B: NMR coronal section in T2. Both views show tumoration in upper internal region of left orbit that compressed and slightly displaced the ocular globe. Molding of internal bone wall.



Figure 3 – Surgery photograph. The tumor was extirpated in one piece through the internal third of the upper palpebral fold.



Figure 4 – View one week after extirpation of the cyst. Symmetry between both upper eyelids has been recovered and the left ocular globe has returned to its normal position.

a clue to suspect a mass in the internal third of the upper eyelid. While awaiting the requested nuclear magnetic resonance (NMR) the astigmatism increased.

The NMR evidenced tumoration in the upper internal angle of the left orbit which compressed the ocular globe, compatible with dermoid cyst (fig. 2). Accordingly, surgical treatment was indicated. An anterior orbitotomy through the internal third of the palpebral fold was performed. Intraoperatively a well-defined lesion was identified in the superior-internal orbital quadrant which was removed in its entirety (fig. 3). One week post-op the patient recovered the symmetry between both upper eyelids and the eye position (fig. 4).

Discussion

Dermoid cysts are one of the most frequent tumors in children. Their origin is congenital and are made up by epidermic

inclusions due to the defective closure of the embryonic facial slits.² Dermoid cysts are a type of choristoma, i.e., a tumoration formed by an aggregation of tissue which is not normally at the site. In this case, it exhibited a keratinizing epithelium with dermal appendices.³

The most frequent presentation of dermoid cysts is the upper orbit temporal quadrant, adjacent to the frontal zygomatic suture, with the medial location being second in order of frequency. They are slow-growing, painless and smooth oval-shaped masses. According to the location and size, the age of emergence as well as clinical signs will vary between proptosis, diplopia, restriction of eye movements or tumoration.¹ Posterior cysts do not emerge until adulthood.³

The treatment consists in complete extirpation. It is important to maintain the cyst intact during surgery because a loss of internal cyst material or remains produce an acute inflammatory response in the orbit or eyelid.⁴

On the basis of the orbital location, the initial diagnostic is clinical. Imaging techniques such as CAT or NMR are necessary to confirm the presence of an orbital mass and to determine location and extension.⁵

In the case presented here, the initial symptoms and signs were very subtle due to its progressive growth and the discrete deviation of the ocular globe associated to astigmatism caused by ocular globe compression, leading to the suspicion of an orbital mass. The NMR showed a mass which deformed the globe and produced a sort of remodeling of the medial orbital wall. Both findings confirmed the slow and progressive growth of the cyst from birth.

As a conclusion, the presence of dermoid cysts must be clinically suspected in children exhibiting small palpebral asymmetries which do not clearly indicate congenital ptosis or motility alterations. Imaging tests must be made to confirm the suspicion and the existence of a space-occupying lesion and to plan its surgical treatment.

Conflict of interest

The authors declare they have no conflict of interests.

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