Short communication

Amblyopia secondary to iris cyst


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A B S T R A C T

Clinical case: A 5 year-old child diagnosed with moderate anisometropic amblyopia secondary to primary cyst of iris pigment epithelium. He was evaluated with ultrasound biomicroscopy (BMU) and optical coherence tomography (OCT) of anterior segment.

Discussion: The OCT, although with some limitations, is a useful tool to study the anterior segment. It is probably more recommendable than BMU in the childhood.

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R E S U M E N

Caso clínico: Varón de 5 años de edad con ambliopía anisotrópica meridional secundaria a quiste de epitelio pigmentario de iris. Es evaluado mediante biomicroscopia ultrasonora (BMU) y tomografía de coherencia óptica de polo anterior (OCT Visante).

Discusión: La OCT de polo anterior, aunque con limitaciones, es una herramienta útil en la evaluación de lesiones de polo anterior. Puede ser preferible, en la infancia, a la BMU.

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Introduction

Cysts of iris pigment epithelium are the most frequent anterior segment benign tumour in childhood. As they are generally asymptomatic, they tend to be infradiagnosed. In their evaluation, the slit lamp exam or biomicroscopy (BMC) and the ultrasound biomicroscopy (UBM) play a significant role.

We present the case of a five-year-old child diagnosed with amblyopia secondary to multiple unilateral cysts of iris pigment epithelium, who was evaluated using UBM as well as optical coherence tomography (OCT) of anterior segment.

Clinical case

The patient is a five-year-old child, with no significant personal or family history, referred to paediatric ophthalmology consultation due to left eye (LE) amblyopia, who had been under treatment with refractive correction and part-time occlusions for six months.

Examination

The child showed a visual acuity (VA) of 0.8 in his right eye (RE) and of 0.5 in his LE, with +1 and +3 −5 to 130°, respectively. He had no stereopsis, evaluated by TNO test. RE dominance; orthophoria. Refraction under cycloplegia was repeated several times given the LE astigmatic axis and amount variability. No lesions are observed in the fundus of eye.

A corneal topography was performed using Pentacam (Oculus®), and the following keratometric data were obtained: RE K1 41.9−6.5° and K2 43.6−96.5°; LE K1 42−172.9° and K2 44−82.9° (Fig. 1). Given the difference between the LE corneal and refractive astigmatism, the anterior segment was thoroughly examined. After being dilated, a pigmented lesion was observed protruding from the pupil edge at V hours (Fig. 2) and leaving a mark on the anterior crystalloid, where a sectorial cortical cataract responsible for lenticular astigmatism (Fig. 3) was starting, which corresponded to a densitometry measurement of 9.8 in such area (Fig. 4).

Due to a suspected cyst of iris pigment epithelium, a UBM and OCT of the anterior segment were performed.

The LE UBM (OTI System®, 35 MHz probe) showed multiple cystic lesions of the iris posterior wall, in the medial portion, with thin, smooth, well-defined, hyperreflective walls with anechoic content, some of which were multilobate, more prominent at the area of IV–VI hours of iris, with no masses in the ciliary body (Fig. 5).

The LE OCT (OCT-Visante Zeiss®) of anterior segment showed the previously described cysts, which moved the iris anterior wall creating a localized iris plateau, without occluding the angle (Fig. 6).

Fig. 1 – Left eye refractive map showing a regular astigmatism in favour of 2 D rule.
Fig. 2 – Photograph taken with a slit lamp where a pigmented lesion was observed protruding from the inferior temporal pupil edge.

Fig. 3 – Cyst mark on the crystalline lens anterior wall and sectorial anterior cortical cataract.

Fig. 4 – (A) Scheimplug image of the left eye crystalline lens and specific densitometry measurement at crystalline lens centre level (6.1). (B) Scheimplug image of the left eye crystalline lens and specific densitometry measurement at inferior temporal sectorial cataract level (9.8).
Fig. 5 – Ultrasound biomicroscopy test of the pigmented lesion. The hyperechoic, smooth, thin walls and hypoechoic content of the lesion are observed. This image specifically shows a multilobate cyst, with no evidence of masses at ciliary body level.

Exploration via BMC, UBM and OCT of anterior segment of the RE was normal.

Treatment was started with RE total and direct occlusion according to a 15/1 schedule and graduation was adjusted. After two months, VA was 1 in both eyes. After a three-year follow-up, VA, the cataract and the cysts have remained stable.

Discussion

Cysts of iris pigment epithelium of the middle part of the iris account for 9% of the iris cysts in childhood and are more frequent in boys. They may be multiple and bilateral. They tend to go unnoticed, becoming more evident when the pupil gets dilated. In general, they remain stable and do not require any treatment. In this case, the clinical manifestation was an astigmatic anisometropic amblyopia, a very rare form of presentation.3,4

They must be differentiated from other causes of anterior segment cystic lesions in children, such as the ciliary body melanoma, the ciliary body medulloepithelioma and the inclusion cysts. The lesion’s cystic nature and its multifocality, evidenced by UBM and OCT, made it possible to rule out the iris melanoma, since the described cases of cavitated melanoma show uneven and thick-walled cysts, associated with a nodular dense lesion.3 In addition, the UBM ruled out the presence of masses at ciliary body level, as well as the ciliary body medulloepithelioma from the differential diagnosis.5,6 Inclusion cysts may appear after amniocentesis or anterior segment penetrating trauma, which have not been reported in this patient.

The use of UBM in children is rather complicated given the fact that it is a contact exploration, performed through an immersion system. In this case, and exceptionally, it was possible to carry out the procedure during the consultation, but in general, it has to be performed in an operating room and under sedation at this age.

The OCT has the advantage of being a non-contact technique that requires a lower level of cooperation. It enables assessment of lesions in cameral angle and iris in 360°, thus obtaining high-resolution images, even better that those obtained by the UBM.7 However, it does not enable the assessment of the ciliary body since the iris pigment epithelium has a screen effect on the laser beam.

In conclusion, in the light of an astigmatic anisometropy, it is necessary to explore the optical system as a whole, without focusing on corneal astigmatism only.

The clinical case presented shows how, on certain occasions, the OCT is more useful than the UBM for the diagnosis of iris cystic lesions in children, as well as for their follow-up.

Fig. 6 – Optical coherence tomography image of anterior segment, showing the lesion’s cystic nature and the iris plateau configuration.
given the higher ease of use and the non-invasive nature of the OCT.

Conflicts of interest

The authors declare that they do not have any conflicts of interest.

REFERENCES