



Bilateral peripupillary pigment epithelium iris cysts About a case

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Introduction

Iris cysts are a rare pathology, even more so in children. They develop asymptotically and can, in some cases, have an impact on the anterior segment (glaucoma, endothelial damage, lens luxation). The treatment is not codified, and remains to be defined on a case-by-case basis, depending on the situation, size and morphology of each cyst.

We report the case of a 5-year-old child, with no particular pathological history, who consulted for a bad visual behavior (squinting, approaching objects).

The ophthalmological examination revealed a visual acuity of 2/10 in the right eye and 1/10 in the left eye. Slit lamp examination identified pigment epithelial cysts around the pupillary rim of the eyes. The intraocular pressure was correct and the lens was clear. The fundus was unremarkable. We completed the examination with a refraction under cycloplegia which showed a myopia of -6 dioptries on the right and -6.50 on the left. The visual acuity increased to 6/10 in both eyes after the refraction.

A UBM (Ultra Biomicroscopy) was performed, confirming the cystic nature of the above-mentioned lesions, round anechogenic lesions surrounded by a hyperechogenic wall at the pupillary margin, without associated ciliary cysts. An OCT of the anterior segment was also performed, which showed elevations of the pupillary rim keeping the central visual axis clear. The general examination of the child did not reveal any particularities.

Given the non-obstruction of the visual axis and the corrected visual acuity of 6/10 at eye level, we decided to delay the surgical removal of the cysts and to establish a close surveillance.

The evolution was marked by the persistence of a stable visual acuity without increase in the size of the cysts.

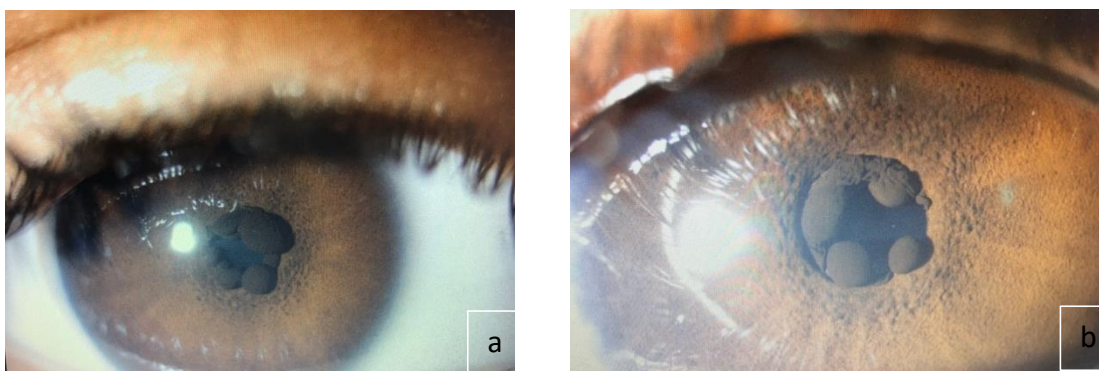


Figure 1: Photograph of iris cysts (arrowhead) right eye (a) and left eye (b)

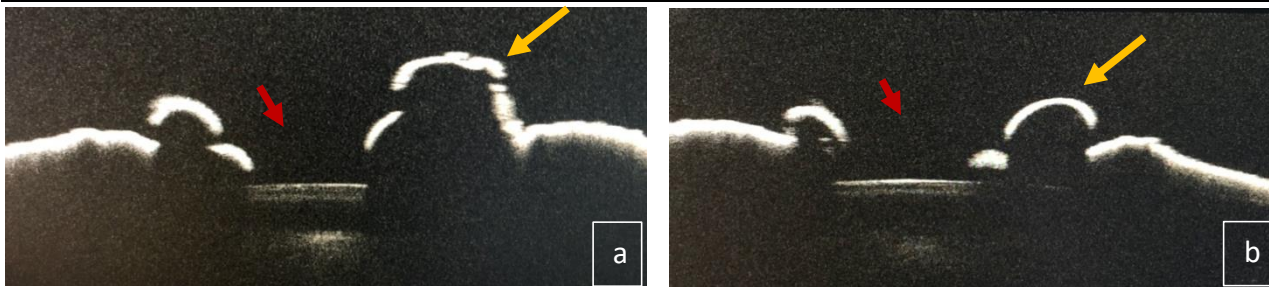


Figure 2: Anterior segment OCT right eye (a) and left eye (b) showing iris elevation (yellow arrow) with a clear visual axis (red arrow)

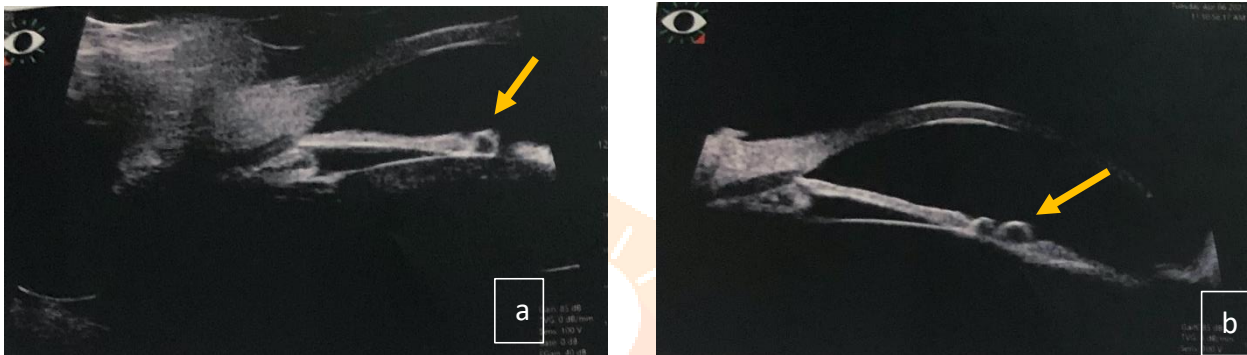


Figure 3: UBM right eye (a) and left eye (b) showing cystic lesions (yellow arrow)

Discussion

Iris tumors can be separated into two groups: cystic and non-cystic, and originate either from the pigment epithelium or the iris stroma.

According to Shields' classification in 1981(1), iris cysts are divided into congenital or secondary (post-traumatic, parasitic, tumor). Primitive cysts of the pigment epithelium, also known as iris flocculi, are divided according to their location: central (peripupillary), peripheral and medial.

Central primitive cysts are rare, about 3%. They present as rounded ovoid lesions, peripupillary, pigmented (dark brown to black) because they originate in the iris pigment epithelium, and move with the movements of the iris sphincter.(2) Generally, the lesion is single in unilateral affection, and multiple if bilateral affection.

The development of the cyst often remains asymptomatic for a long time. The diagnosis is usually made fortuitously during a routine ophthalmologic examination(4).

They can also lead to certain complications such as glaucoma (angle closure or plateau iris arrangement), or stromal opacity (rubbing against the endothelium). These complications are mostly encountered in peripheral cysts and in the medial zone due to their location and size.

For a long time, iris cysts were under-diagnosed and confused with iris tumors such as melanoma or iris adenoma, or medulloepithelioma. UBM (Ultra Biomicroscopy), has allowed to better study and analyze these lesions, and especially provide an effective means of monitoring (increase in size, thickening of the walls ..).

Primary pigment epithelial cysts do not require systematic treatment. The therapeutic decision takes into account several factors: location, size, presence of complications, invasion of the visual axis. Removal of the cyst can be done surgically or less invasively, by laser photocoagulation of the cyst walls.(2) In the case reported, given the absence of visual axis invasion and the improvement of visual acuity after optical correction, as well as the benign aspect of the lesion, no treatment was undertaken.

In a 2011 study by Disabella et al, the presence of iris flocculi may be associated with dissecting aortic aneurysms because of a common mutation in the alpha-actin 2 (ACTA2) and/or myosin heavy chain (MYH11) gene, which affects both the iris and the aorta. Indeed, the most common extravascular involvement found in TAA (Thoracic Aortic Aneurysm and/or Dissection) was the presence of iris cysts.(5) In our case, a general examination of the child did not find any abnormalities.

Conclusion

Primary pigmentary epithelial cysts are clinical entities whose diagnosis and management remain simple. The UBM keeps an important place in the diagnosis, evaluation and monitoring of these lesions. Their possible association with aortic aneurysms or dissections justifies and should encourage each practitioner to perform a cardiac evaluation in order to detect a potential aortic aneurysm.

Bibliography

1. Shields JA, Kline MW, Augsburger JJ. Primary iris cysts: a review of the literature and report of 62 cases. Br J Ophthalmol. 1 mars 1984;68(3):152-66.
2. Freton A, Caujolle JP. Cas bilatéral de kystes péripupillaires de l'épithélium pigmentaire irien. 2011;(2):3.
3. Mghaieth F, Chaker N, Limeim R, El matri L. Kystes de l'iris chez l'enfant. a propos de 2 cas Bull. Soc. belge Ophtalmol.,295, 23-27, 2005.
4. Köse HC, Gündüz K, Hoşal MB. Iris Cysts: Clinical Features, Imaging Findings, and Treatment Results. Turk J Ophthalmol. 1 janv 2020;50(1):31-6.
5. Disabella E, Grasso M, Gambarin FI, Narula N, Dore R, Favalli V, et al. Risk of dissection in thoracic aneurysms associated with mutations of smooth muscle alpha-actin 2 (ACTA2). Heart. 15 févr 2011;97(4):321-6.