



Bilateral congenital stromal iris cyst with secondary glaucoma

Toshit Varshney^{*}, Shikha Gupta, Karthikeyan Mahalingam, Viney Gupta, Seema Kashyap

Dr Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, 110029, India

ARTICLE INFO

Keywords:

Ultrasound biomicroscopy
Congenital stromal iris cysts
Glaucoma

ABSTRACT

Purpose: To report a case of bilateral congenital stromal iris cyst in a neonate.

Observations: A one-month-old male child was brought with complaints of watering in both eyes and enlargement of the left eye since birth. Examination under anesthesia revealed an elevated IOP in both the eyes. The left eye had buphthalmos with hazy cornea and central corneal scarring. Ultrasound biomicroscopy revealed the presence of a stromal cyst in both the eyes and a cataractous lens in the left eye. While the right eye responded to topical glaucoma medications, the left eye continued to have a high IOP and hence, it was managed surgically.

Conclusions and Importance: This rare case of bilateral stromal iris cyst in a neonate presented with a unique diagnostic and management challenge.

1. Introduction

Congenital stromal iris cysts are relatively uncommon, with a higher incidence in the pediatric age group. They are usually unilateral, with majority being asymptomatic and stationary in course.^{1,2} Here, we report a case of an infant with bilateral stromal cysts who presented with secondary complications.

2. Case report

A one-month-old male infant was brought to the outpatient department with complaints of watering in both eyes and enlargement of the left eye since birth. The child was of a full-term normal vaginal delivery with no significant family history of ocular disorders. Examination of his parents and 2-year-old sibling was unremarkable. On examination under anesthesia, his IOP was 24 mmHg and 28 mmHg (Perkins applanation tonometer) in his right eye (RE) and left eye (LE), respectively. The RE cornea was clear with an iris mass lesion in the lower half of anterior chamber covering the pupillary plane. The LE cornea was hazy with a central scar and had a horizontal corneal diameter of 13mm [Fig. 1 A and B]. Ultrasound Biomicroscopy (UBM) with a 50MHz probe of both the eyes revealed a single, thin-walled intrastromal iris cyst with internal lucency. The surrounding iris was thin and atrophic. The cyst appeared to be adherent anteriorly to the corneal endothelium. The RE lens appeared to be normal. The cyst in the LE was larger in dimensions, and the lens was cataractous [Fig. 1 C and D].

A posterior segment B/A-mode ultrasonography (USG) of both the eyes with a 10Mhz probe showed an anechoic posterior segment with optic nerve head cupping in both eyes. Axial length was 24.4mm and 25.6mm in the right and left eye, respectively. A diagnosis of bilateral primary congenital stromal iris cyst with secondary glaucoma was made, and the patient was started on topical betaxolol 0.25% twice daily and topical dorzolamide 2% thrice daily in both eyes. On subsequent follow-up, sleeping IOP of 10 mmHg and 26 mmHg was recorded in the right and left eye, respectively. Due to high IOP in the LE despite maximum medications, surgical excision of cyst was planned. Although, the cyst was obscuring the visual axis in the RE as well, the parents denied consent for bilateral eye surgery in view of poor prognosis and need for multiple surgeries. Under general anesthesia, complete visco-dissection of anterior cyst wall was done from corneal endothelium followed by cyst excision and sectoral iridectomy. In a subsequent surgery, a lens aspiration was performed combined with trabeculectomy. The eye was left aphakic. Histopathology confirmed the diagnosis of intrastromal iris cyst lined by non-keratinized squamous epithelial cells [Fig. 2]. Postoperatively, aphakic glasses were prescribed, and amblyopia therapy was initiated. On follow-up EUA at three months, no recurrence was noted in the LE, and IOP in both the eyes was controlled.

3. Discussion

Congenital iris cysts can be of pigment epithelial or stromal types. Stromal cysts form 11% of all cysts, with a higher incidence in the first

; IOP, Intraocular pressure; UBM, Ultrasound Biomicroscopy.

* Corresponding author.

E-mail address: t1varshney@gmail.com (T. Varshney).

<https://doi.org/10.1016/j.ajoc.2022.101261>

Received 23 July 2021; Received in revised form 10 January 2022; Accepted 11 January 2022

Available online 20 January 2022

2451-9936/© 2022 The Authors.

Published by Elsevier Inc.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

two decades. Though pigmented epithelial cysts tend to be bilateral, stromal cysts are usually unilateral.¹⁻⁴ To the best of our knowledge, this is the first reported case of a congenital bilateral stromal cyst. The pathogenesis most widely accepted is the entrapment of surface ectodermal cells into iris stroma during the fourth week of embryogenesis when the lens vesicle is formed.⁴ Due to this, the epithelial lining of stromal cyst varies from stratified squamous to unilayered cuboidal epithelium.²

Primary iris cysts are usually asymptomatic owing to their stationary or slow-growing nature and are usually an incidental diagnosis. They are more aggressive in younger children and present with secondary complications like glaucoma, uveitis, strabismus, or amblyopia.^{2,5} Our patient had raised IOP in both eyes. The left eye had buphthalmos, corneal scarring, and total cataract as well. Although these cysts are usually visible on routine anterior segment examination, high frequency (50–100Mhz) ultrasound biomicroscopy (UBM) remains the gold standard for diagnosis, especially when the corneal clarity is compromised. The UBM offers a resolution of 20–50 μm with tissue penetration of 3–5mm. The size, location, extent of iris cysts can be assessed in vivo, especially in cases with corneal opacity. UBM reveals the entire structure of the iris cyst, including the posterior extent as well as its relationship with surrounding tissues and lens status. All these help differentiate benign iris cysts from solid tumors and other anterior segment lesions simulating iris lesions.^{2,6,7}

Stationary or slowly growing uncomplicated cysts are usually observed. Treatment options for symptomatic cysts include fine-needle aspiration of cyst followed by cauterization or cryotherapy of cyst base, intracystic injection of mitomycin or absolute alcohol followed by irrigation, laser photocoagulation of the cyst wall or surgical excision.²

Recurrence after treatment is a common complication of iris stromal cysts. Simple aspiration of the cyst is associated with a higher chance of cyst recurrence along with potential complications like iritis and glaucoma due to the sudden release of cystic contents. Irrigation with absolute alcohol following aspiration reduces failure rate; however, its side effects include transient corneal edema and anterior chamber inflammation requiring corticosteroid treatment.^{5,8} Incomplete or partial resection of stromal cyst also results in recurrences.⁹ While a complete surgical excision is highly successful with minimal recurrences, it is also associated with damage to other adjoining ocular structures.^{5,10} Further, a subsequent filtration surgery may be required in eyes with uncontrolled IOP.¹⁰ In our patient, a radical cyst excision was performed for LE in view of secondary complications and was followed by an IOP lowering surgery. However, the visual prognosis in our case remained poor because of amblyopia and pre-existing secondary glaucoma.

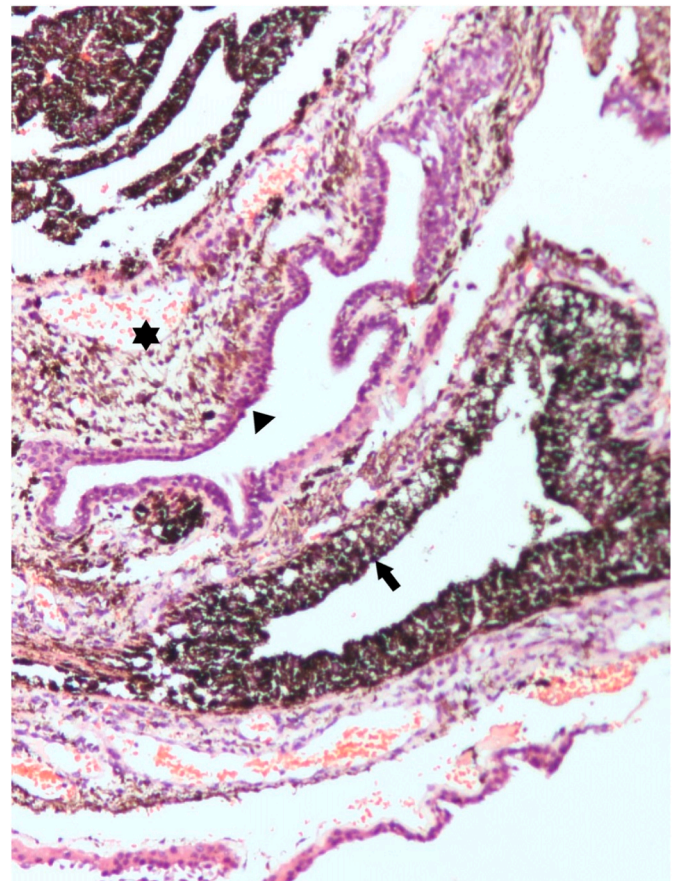


Fig. 2. Histopathological section showing iris stroma (asterisk), posterior pigmented epithelium of iris (arrow) and stromal cyst with a cystic lumen lined by non-keratinized stratified squamous epithelial cells (arrowhead).

4. Conclusion

While most stromal cysts are asymptomatic, an early diagnosis and recognition of complications like glaucoma is necessary. Treatment and visual rehabilitation are of paramount importance, especially at a young age, to prevent amblyopia.

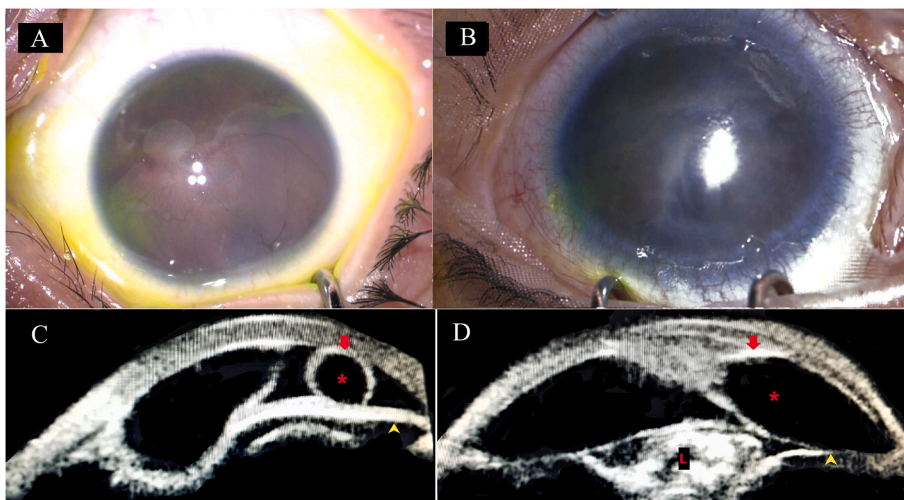


Fig. 1. (A) Clinical photograph of the intrastromal iris cyst in the lower half of right eye covering the pupillary plane. (B) Clinical photograph of the left eye showing corneal edema and scarring that has obscured the details of the anterior segment. (C and D) Ultrasound biomicroscopy (UBM) of right and left eye respectively, showing thin-walled iris stromal cyst (asterisk) with endothelial corneal touch (arrows) anteriorly and a surrounding atrophic iris (arrowheads). A thickened and scarred cornea with a cataractous lens (L) in the left eye is seen.

Patient consent

Consent to publish this case report has been obtained from the patient's guardians in writing as the patient himself is an infant.

Funding

No funding or grant support.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

None of the authors have any conflicts of interest related to this submission.

The following authors have no financial disclosures: TV, SG, KM, VG, SK.

Acknowledgements

None.

References

1. Shields CL, Kancherla S, Patel J, et al. Clinical survey of 3680 Iris tumors based on patient Age at presentation. *Ophthalmology*. 2012;119(2):407–414. <https://doi.org/10.1016/j.ophtha.2011.07.059>.
2. Georgalas I, Petrou P, Papaconstantinou D, Brouzas D, Koutsandrea C, Kanakis M. Iris cysts: a comprehensive review on diagnosis and treatment. *Surv Ophthalmol*. 2018;63(3):347–364. <https://doi.org/10.1016/j.survophthal.2017.08.009>.
3. Shields JA. Primary cysts of the iris. *Trans Am Ophthalmol Soc*. 1981;79:771–809.
4. Shields J, Shields C, Lois N, Mercado G. Iris cysts in children: classification, incidence, and management. *Br J Ophthalmol*. 1999;83(3):334–338.
5. Lois N, Shields CL, Shields JA, Mercado G, Potter PD. Primary iris stromal cysts: a report of 17 cases. *Ophthalmology*. 1998;105(7):1317–1322. [https://doi.org/10.1016/S0161-6420\(98\)97041-5](https://doi.org/10.1016/S0161-6420(98)97041-5).
6. El-Seht. Ultrasound biomicroscopy as a guideline for management of secondary iris cysts: a case series study. <http://www.djo.eg.net/article.asp?issn=1110-9173;year=2018;volume=19;issue=1;spage=24;epage=29;aulast=El-Seht>. Accessed April 27, 2020.
7. Rao A, Gupta V, Bhadange Y, Sharma R, Shields JA. Iris cysts: a review. *Semin Ophthalmol*. 2011;26(1):11–22. <https://doi.org/10.3109/08820538.2010.541319>.
8. Shields CL, Arepalli S, Lally EB, Lally SE, Shields JA. Iris stromal cyst management with absolute alcohol-induced sclerosis in 16 patients. *JAMA Ophthalmology*. 2014;132(6):703–708. <https://doi.org/10.1001/jamaophthalmol.2014.160>.
9. Casey M, Cohen KL, Wallace DK. Recurrence of iris stromal cyst following aspiration and resection. *J AAPOS*. 2002;6(4):255–256. <https://doi.org/10.1067/mpa.2002.124647>.
10. Haller JA, Stark WJ, Azab A, Thomsen RW, Gottsch JD. Surgical management of anterior chamber epithelial cysts. *Am J Ophthalmol*. 2003;135(3):309–313. [https://doi.org/10.1016/S0002-9394\(02\)01960-8](https://doi.org/10.1016/S0002-9394(02)01960-8).