

Significance of Accurate Diagnosis and Optimal Management Approach for Bilateral Anterior Lenticonus

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ABSTRACT

A 16-year boy sought a second opinion for bilateral, gradual, and painless visual impairment, initially diagnosed as keratoconus. Despite a previous clinical diagnosis, a careful examination and corneal topography excluded keratoconus. Distant direct ophthalmoscopy revealed a central symmetrical oil-droplet sign and anterior segment examination showed a conical protrusion of the anterior lens capsule. Anterior segment-optical coherence tomography (AS-OCT) confirmed the diagnosis of anterior lenticonus. With a best-corrected visual acuity of 6/36 in the right eye and 6/24 in the left eye, the patient was diagnosed with Alport syndrome (AS), supported by bilateral sensorineural hearing loss and proteinuria/hematuria. The recommended approach involved bilateral clear lens exchange (CLE) with irrigation/aspiration (I/A), emphasising meticulous care during continuous curvilinear capsulorhexis (CCC) using utrata forceps. This case underscores the importance of a holistic approach, combining thorough history, examination, and investigations for accurate diagnosis, as well as discusses the surgical management of the relatively rare finding of anterior lenticonus associated with AS.

Key Words: Alport Syndrome, Anterior lenticonus, Anterior segment-OCT.

How to cite this article: Mazhry ZU, Asif L, Mazhry MA, Saeed M. Significance of Accurate Diagnosis and Optimal Management Approach for Bilateral Anterior Lenticonus. *JCPSP Case Rep* 2024; **2**:68-70.

INTRODUCTION

Bilateral anterior lenticonus is an uncommon condition typically linked to Alport syndrome (AS). It is characterised by a genetic defect in the synthesis of type IV collagen.¹ This collagen defect eventually results in increased fragility of the lens capsule. AS can be inherited through various modes, with X-linked inheritance being the most common. The key ocular abnormalities associated with this syndrome include anterior lenticonus, perimacular changes, and peripheral fleck retinopathy.² This report focuses on the case of a patient with bilateral anterior lenticonus, highlighting the approach that facilitated the timely diagnosis of AS.

CASE REPORT

A 16-year male patient presented to Eye OPD with reported steady and painless deterioration in vision of both eyes over the past 7 to 8 years. The boy came for a second opinion about corneal cross-linking (CXL) that he had been advised from elsewhere. His best-corrected visual acuity (BCVA) was 6/36 in the right eye and 6/24 in the left eye.

Distant direct ophthalmoscopy showed a positive oil droplet sign that was unusually symmetrical and central. During the slit lamp examination of the anterior segment, everything appeared normal except for the presence of a conical protrusion of the anterior lens capsule right in the visual axis (Figure 1). The fundus examination revealed a cup-disc-ratio of 0.3/0.3 bilaterally, with no abnormal findings observed in the macula. Intraocular pressure (IOP) was within normal range bilaterally. Upon further questioning, the patient also complained about difficulty in hearing. In family history, it was disclosed that the parents were closely related. While the parents did not display any symptoms, their older sibling had unfortunately passed away due to renal failure of unknown origin.

Anterior segment optical coherence tomography (AS-OCT) (OCT Nidek 3000 Advance) confirmed the diagnosis of bilateral anterior lenticonus (Figure 2). Coloured fundus photograph showed normal fundi (Figure 3). Corneal Topography using Galilei G6 Dual Scheimpflug analyser showed normal corneal parameters. Pure tone audiometry revealed moderately severe bilateral sensorineural hearing loss. Urine analysis showed proteinuria 3+ and hematuria.

Taking into account the supporting evidence, a diagnosis of bilateral anterior lenticonus associated with AS was established. To address the visual impairment, clear lens exchange (CLE) using irrigation/aspiration (AI) with intraocular lens (IOL) implantation was performed.

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Received: October 30, 2023; Revised: January 24, 2024;

Accepted: January 28, 2024

DOI: <https://doi.org/10.29271/jcpspcr.2024.68>

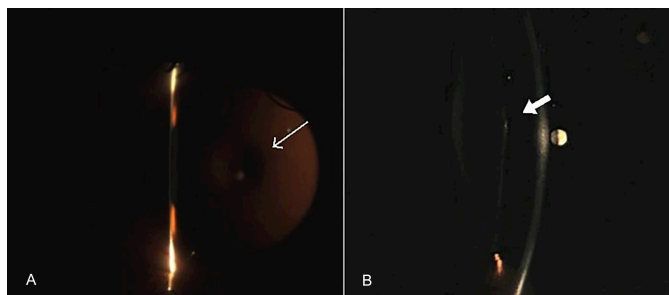


Figure 1: Illustration of the right eye via Slit Lamp Photography - (A) Displaying characteristicsymmetricaloil dropletreflection underretro-illumination (indicated by arrow). (B) Cross-section through the anterior lens capsule highlighting conical protrusion within the pupillary zone (highlighted by bold arrow).

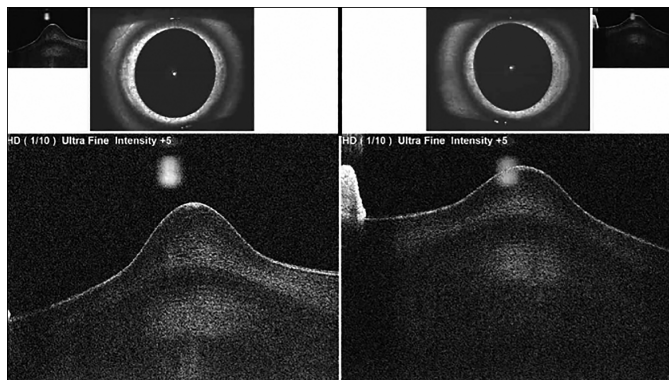


Figure 2: Visualisation of anterior lens capsule conical protrusion utilising Nidek 3000 advanced anterior segment module in anterior segment optical coherence tomography.

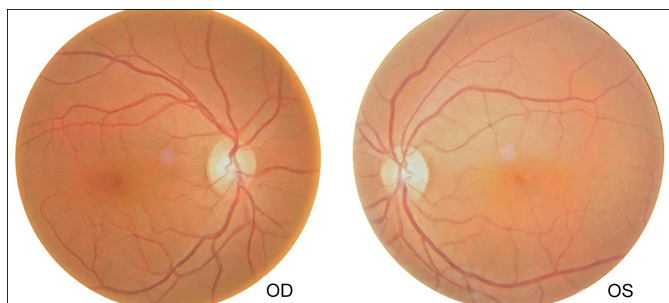


Figure 3: Coloured fundus photograph showing normal fundi.

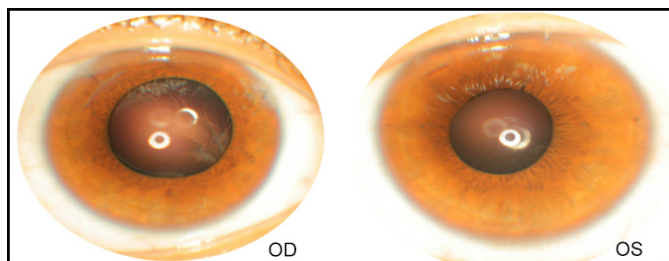


Figure 4: At 4 weeks post-operatively, early posterior capsule opacification (PCO) in the right eye not involving the visual axis. The best-corrected visual acuity (BCVA) was 6/9 in both eyes.

The surgical procedure was scheduled sequentially with a one-week gap between the two surgeries, and it was conducted under local anaesthesia. Prior to the surgery, optical biometry using Zeiss IOL Master was performed. Single side port at 10 o'clock and a main incision of 2.75 mm was fashioned. Hydroxypropyl methylcellulose (HPMC) was used to maintain the anterior

chamber. Delicate manipulation of the anterior capsule was carried out to ensure the creation of a central continuous curvilinear capsulorhexis (CCC) using utrata forceps. A suture was placed in the end to close clear corneal incision (CCI) to prevent wound leakage resulting from fish mouthing of incision lips due to decreased scleral rigidity and tissue elasticity.

One day after the initial surgery, the visual acuity (VA) was recorded as 6/36 bilaterally. The postoperative medications prescribed consisted of topical prednisolone acetate 1% drops, to be administered every hour for the first week and then gradually reduced over the following 4 weeks. Additionally, moxifloxacin 0.5% eye drops were recommended to be used four times a day during the initial week. At 4 weeks postoperatively, the BCVA was 6/9 in both eyes with correction of -1.50 Cyl D@70° in the right eye and $+0.50$ Cyl D@35° in the left eye (Figure 4). For near vision, an addition of $+2.50$ was prescribed bilaterally. The patient was also referred for auditory rehabilitation and to a nephrologist for further management.

DISCUSSION

Bilateral anterior lenticonus is a rare ocular condition often associated with AS, a genetic multisystem disorder primarily affecting the basement membranes of kidneys, eyes, and ears.³ Other common ocular features include posterior lenticonus, perimacular changes, and dot and fleck retinopathy.⁴ Timely evaluation of the patients with bilateral anterior lenticonus using a holistic approach can lead to the establishment of early diagnosis of AS.

During clinical examination of these patients, it should be kept in mind that the bulge in the capsule can create a central, almost perfectly symmetrical oil droplet appearance on retroillumination, which when initially examined on distant direct ophthalmoscopy can give rise to a false impression of keratoconus. However, a detailed examination on slit-lamp can omit this confusion. Moreover, the significance of use of AS-OCT, as we did in our case, cannot be overemphasised to confirm and document the presence of this unique lenticular abnormality as also highlighted by Sedaghat *et al.*⁵

Due to defective collagen, the capsule becomes thin and elastic, making capsulorhexis technically challenging. Boss *et al.* reported that the capsule has an unusual cogwheel tearing pattern, which can lead to rips and runoffs.⁶ We followed a careful approach, initiating the rhexis with cystotome at a mid-peripheral position and completing it with utrata forceps. IA was performed cautiously, considering the fragile nature of the posterior capsule. There were no associated posterior capsular abnormalities in this case. After hydrodissection and hydrodelineation, we implanted a foldable acrylic aspheric IOL, taking care to avoid contact with the posterior capsule.

Early recognition of ocular features can lead to early diagnosis and subsequent management as end-stage renal disease can be delayed by angiotensin converting enzyme (ACE) inhibitors and angiotensin receptor blockers (ARBs) in a time-dependent manner.⁷

In conclusion, anterior lenticonus of AS can be successfully managed with CLE and IOL implantation with careful handling of delicate capsule. This case underscores the importance of a holistic approach, combining thorough history, physical and detailed ocular examination, and investigations for accurate diagnosis, as well as discusses the surgical management of the relatively rare finding of anterior lenticonus associated with AS.

PATIENT'S CONSENT:

The informed consent was obtained from the patient to publish the data concerning this case.

COMPETING INTEREST:

There authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

ZM: Concept, design, and manuscript review.

LA, MS: Data acquisition, drafting, manuscript review, and editing.

MAM: Drafting, manuscript review, and editing.

All authors approved the final version of the manuscript to be published.

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