Inverse atoll sign as preoperative indicator of posterior capsular defects in posterior lenticonus

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CASE 1

A 7-year-old boy presented with a white reflex in the right eye along with diminution of vision, which was noticed by his parents for the previous 3 months. Ocular examination revealed a corrected distance visual acuity (CDVA) of 20/400 with no manifest refractive error in the right eye and 20/30 with +1.50 +0.75 × 90 in the left eye. On slitlamp examination, the right eye had posterior polar cataract along with the posterior coning of the lens. The dense white central lenticular opacity corresponded with the apex of the cone. This central opaque zone was surrounded by a clearer zone or halo, which was further surrounded by a concentric rim of opacification (Figure 1, a). The nomenclature inverse atoll sign was proposed because the appearance described here is inverse of the appearance of an atoll reef. The left eye showed early posterior subcapsular cataract (PSC) with a normal contour of the posterior lens capsule (Figure 1, b).

Bilateral anterior segment imaging was done using SS-OCT (CASA 2, Tomey Corp.). The OCT images showed a conical contour of the posterior surface of the lens in the right eye, with lens thickness of 7.2 mm measured using lens analysis function on the SS-OCT (Figure 1, c). The posterior lens capsule, normally seen as a thin hyperreflective membrane at the posterior lens surface, showed discontinuity along with herniation of the cortical matter from the zone of capsular deficit. The edges of the posterior capsule at the margins of the defect showed a subtle outward angulation, which clinically could translate into the smooth, well-delineated appearance of the outer rim of the inverse atoll reef mentioned earlier. SS-OCT of the left eye showed a normal lens contour with intact posterior capsule and early cataractous changes corresponding to the PSC seen clinically (Figure 1, d).

A diagnosis of right eye posterior lenticonus with cataract was made, and thorough systemic evaluation was performed, which was unremarkable. Cataract surgery of the right eye was performed keeping the existence of a PCD in mind. A 5 mm continuous curvilinear capsulorhexis (CCC) was...
by an opaque ring (red arrows), indicating the edges of the PCD. b: Left eye on retroillumination showing an early posterior subcapsular cataract. c: SS-OCT of the right eye showing the intact posterior lens capsule visualized as a thin hyperreflective membrane (green arrows) and herniation of the lens matter through the PCD in a conical configuration (area between yellow arrows). The conical zone is hyperreflective, signifying opacification of the lens matter in this zone. The margins of the deficient posterior capsule are turned outward (red arrows). d: SS-OCT of left eye showing a normal lens contour and intact posterior capsule with central hyperreflectivity suggestive of early cataractous changes in the lens (PCD = posterior capsular defect; SS-OCT = swept-source optical coherence tomography).

CASE 2
A 9-year-old girl presented with complaints of decreased vision in both eyes since birth. CDVA in the right eye was 20/40, and hand motion in her left eye. The left eye had a cataractous lens with posterior coning. Similar to case 1, in this patient also the central opaque zone was surrounded by a relatively clear area that was further encircled by an opaque rim along the posterior lens surface, forming an inverse atoll reef (Figure 2, a).

Anterior segment OCT of the left eye showed the posterior lens capsule as a hyperreflective membrane that was deficient centrally, signifying a PCD in that area and a globular herniation of the lens matter into the vitreous cavity (Figure 2, b). A relatively hyporeflective area was seen on either side of the globular protrusion, the outer limits being defined by the edges of the posterior capsule, which in this case also demonstrated an outward angulation. These hyporeflective zones would correspond to the intermediate clear zone or halo seen clinically.

Keeping in mind the possibility of a large PCD as suggested by clinical picture and supported by SS-OCT, a slow controlled lens matter aspiration using a Simcoe cannula was planned to ensure minimal turbulence in the anterior chamber. A circular anterior CCC of 5 mm was created, hydrodissection was avoided, gentle viscodissection was performed, and lens matter was aspirated. A large PCD, the configuration of which corresponded to the concentric opaque rim noted preoperatively, was detected intraoperatively, with dimensions of approximately 5.8 mm as measured on CALLISTO eye rhexis guide. The edges of the cataractous deformity of posterior lens surface and a hyperreflective posterior capsule that is deficient centrally (area between yellow arrows) with herniation of lens matter into the vitreous cavity (area between white arrows). The margins of the deficient posterior capsule are turned outward (red arrows). The green arrow shows the hyporeflective zone adjacent to borders of PCD representing that part of the cortex which, although not covered by the posterior capsule, was not yet opacified. This corresponds clinically to the intermediate clear zone or halo. c: Intraoperative picture of the left eye after removal of the lens matter shows the presence of a large PCD, the shape of which corresponds to the opaque rim shown in the preoperative picture (red arrows). Blue arrows delineate the margins of the well-centered anterior continuous curvilinear capsulorhexis. Some vitreous membranes are seen in anterior vitreous. The PCD can be seen extending beyond the borders of anterior rhexis at 5 and 11 o’clock. d: SS-OCT of the right eye showing a normal contour of the lens with an intact posterior capsule and central hyperreflectivity, which is commensurate with the cataractous changes seen clinically (PCD = posterior capsular defect; SS-OCT = swept-source optical coherence tomography).
PCD extended beyond the borders of anterior CCC at 5 and 11 o’clock (Figure 2, c). Limited anterior vitrectomy was performed using a vitreous cutter, and a 3-piece posterior chamber IOL was implanted into the sulcus with optic capture.

Postoperative CDVA at 4 weeks was 20/60 with a refractive error of 1.0+1.75 × 80. The patient was prescribed spectacles and started on amblyopia therapy. The right eye of this patient had a PSC that accounted for the suboptimal vision. Figure 2, d shows the OCT of the right eye with a central hyperreflectivity arising from the posterior capsule and extending centrally (corresponding to the cataractous changes seen clinically), a normal contour of the posterior lens surface, and an intact posterior capsule.

Although extension of the PCD did not occur in either of the cases, the posterior capsule surrounding the PCD demonstrated excessive fluttering movements intraoperatively, suggestive of enhanced fragility. Considering the large size of the preexisting PCD combined with posterior capsular fragility, IOL implantation in the bag was not attempted in either case.

**DISCUSSION**

Many theories have been given to explain the pathogenesis of posterior lenticus, including posterior traction caused by the hyaloid artery, an inherently weak posterior capsule, overgrowth of the posterior lens epithelium, and cortex producing lens phakoma. Although most cases of posterior lenticus occurs sporadically, association has been found with Lowe syndrome, which is an X-linked disorder characterized by intellectual and developmental disabilities, renal dysfunction, corneal keloids, and glaucoma. Diagnosis of posterior lenticus is mainly clinical. Some clinical signs have been described previously including the oil droplet sign on retroillumination and fish tail sign (lenticular cortex hanging in vitreous cavity after posterior capsular dehiscence).

The 2 cases in this study had preexisting PCD, with anatomic picture exactly reverse of a typical atoll reef. To the authors’ knowledge, the central opaque zone corresponds to the herniating cataractous lens matter in the vitreous cavity, the outermost opaque rim represents the borders of PCD, and some lens matter that is not herniating into the vitreous cavity and is as yet noncataractous (probably supported by the anterior vitreous face) forms the halo between the central opaque zone and the peripheral opaque rim. This clinical impression was supported by the SS-OCT findings preoperatively and confirmed intraoperatively. Other recommended modalities for imaging the posterior lens capsule are 20 MHz echography, ultrasound biomicroscopy, and Scheimpflug imaging. Echography and ultrasound biomicroscopy are contact-based imaging devices for which young children might not cooperate. Imaging using Scheimpflug-based devices, although noncontact and fast to acquire, fails to provide a precise view of the posterior lens surface. Because of its rapid image acquisition (50,000 A-scans/s), it is an excellent imaging tool for young children.

In conclusion, the awareness of the operating surgeons about the possibility of a preexisting capsular defect in cases of posterior lenticonus and recognition of clinical signs that reflect the status of the posterior capsule aided by appropriate imaging can help in safe execution of cataract surgery in these patients.

**WHAT WAS KNOWN**

- Various clinical signs, such as fish tail sign and atoll sign, to describe the posterior capsular integrity help surgeons anticipate intraoperative complications while managing cases of posterior lenticus with cataract.
- Modalities such as ultrasound biomicroscopy (UBM) and Scheimpflug imaging are helpful in supporting the diagnosis because none of the clinical signs are definitive indicators of posterior capsular integrity.

**WHAT THIS ARTICLE ADDS**

- A new inverse atoll sign for preoperative detection of a posterior capsular defect (PCD) in cases of posterior lenticus can serve as a reliable predictor of PCD even in the absence of other diagnostic modalities. Longer-wavelength swept-source optical coherence tomography to assess the integrity of posterior capsule was more advantageous than other conventional imaging modalities, such as UBM and Scheimpflug imaging.

**REFERENCES**

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