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New Sign for Recognizing Posterior Lenticonus Complicated with Cataract and its Surgical Management

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Abstract:

Aim: To describe a new diagnostic sign and improved technique for cataract extraction in cases with posterior lenticonus.

Setting: Kasr Alainy hospital, Cairo University, Cairo, Egypt

Methods: A consecutive series comprised of 16 eyes of 15 patients with definite or suspected posterior lenticonus were diagnosed and surgically treated by a modified technique to prevent premature opening of posterior capsule

Results: The posterior cone can be seen in early cases, but is obscured by the cataract in advanced cases. We have recognized irregular posterior opacity as a sign of posterior lenticonus. Our technique for cataract extraction prevented premature opening of posterior capsule.

Conclusion: Recognition of posterior lenticonus is key for uncomplicated cataract removal in these cases. We describe a new sign and a new technique to manage posterior lenticonus safely.

Keywords: Posterior lenticonus; Cataract; Developmental lens anomaly; Intraocular lenses

Introduction

Posterior lenticonus is a developmental anomaly of the lens in which the posterior portion of the lens bulges posteriorly in a cone shape. It may be well circumscribed or not. Most of the cases are unilateral and sporadic. Bilateral cases may have a hereditary factor. Autosomal-dominant, autosomal-recessive and X-linked pedigrees with bilateral posterior lenticonus have been reported [1,2]. Posterior lenticonus is usually associated with cataract. It is often progressive and limited to the posterior subcapsular and cortical region of the lens [3-5].

Many cases, especially those with dense cataract can be easily missed. We have recognized a sign that can help diagnose posterior lenticonus before entering the anterior chamber. Premature opening of the posterior capsule with vitreous prolapse is a major concern during removal of cataractous lenses with posterior lenticonus. We suggest a modified surgical technique to avoid premature opening of posterior capsule. We have searched PubMed and no similar technique has been described. An article was published at ophthalmic hyperguide.com which used nearly the same technique with a limited number of patients.

The aim of this work is to describe diagnosis and a technique for cataract extraction in cases with posterior lenticonus.

Patients and Methods

This consecutive series comprised 16 eyes of 15 patients with definite or suspected posterior lenticonus who had surgery between January 2009 and March 2011. This study was approved by Cairo University Research Ethics Committee.

All patients had a complete ophthalmological examination. Visual acuity was assessed according to the age of the patient. Direct ophthalmoscopy, retinoscopy and slit lamp examination and B-scan ultrasonography were done in all cases. Intraocular pressure (IOP) was measured preoperatively by either Goldman applanation tonometer or Perkins handheld tonometer. Vision assessed by Snellen chart for children ≥ 5 years while in younger children were assessed by fixation pattern.

Surgical technique

All surgeries were performed by the same surgeon (A.M.K). A superior 3.0 mm clear-cornea incision and 2-20 gauge MVR paracentises were made. An anterior capsulorrhexis was performed. Partial hydrodissection was accomplished by using multiple, small strokes in a circular peripheral fashion. The aim was to separate the peripheral cortex but not reaching the edge of the posterior lenticonus. Using a bimanual technique, the peripheral portion of the cataract is removed first. The central portion of the cataract served as a shield to protect the defect/thin portion of the posterior capsule (Figure 1).

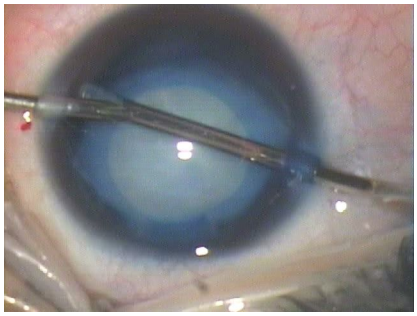

Figure1: Aspiration of peripheral cortex leaving central cortical shield.

The posterior cortex was removed cautiously and slowly to decrease the incidence of posterior capsule rupture. A 3-piece posterior chamber IOL was implanted in the bag in 14 eyes and in the sulcus in 2 eyes. A pars plana posterior capsulotomy and anterior vitrectomy were perfomed through a sclerotomy 2.5 mm posterior to the limbus in 10 eyes after placement of the IOL in the bag. The sclera and conjunctiva were closed separately using 10-0 nylon sutures. In the other 6 eyes, a posterior capsular defect was noted and anterior vitrectomy was performed through the corneal incision prior to lens implantation (Figure 2). In 4 of these cases the IOL was placed in the bag and in 2, the posterior capsular defect was too large for safe in-the-bag placement and the IOL was placed in the sulcus. The 3 mm corneal wound was closed with one 10-0 nylon suture. Subconjunctival triamcinolone was injected in the inferior fornix. Post-operative treatment was dexamethasone/tobramycin drops for 4 weeks. Patients were followed up for at least 6 months. In each follow up visit vision, IOP, position of IOL and visual axis were assessed.

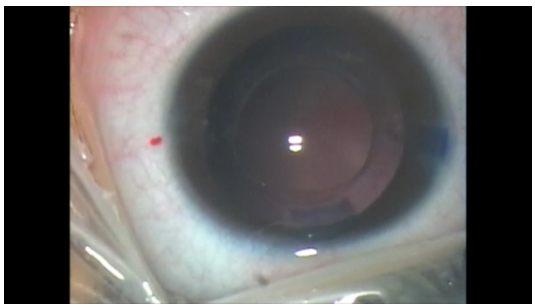

Figure 2: Posterior capsular defect after aspiration of central cortex.

Results

Sixteen eyes of 15 patients with posterior lenticonus were included in this study. Mean patient age was 5 ± 1.4 years (range, 3-7 years). Thirteen eyes (81.25%) were for males and 3 (18.75%) were for females. The left eye was more commonly affected (10 eyes). The mean axial length was 23.72. The mean keratometry was 43.75. The parents complained of poor vision in just one case with bilateral involvement; the others complained of deviation of one eye. Direct ophthalmoscope examination showed an "oil droplet" reflex in 6 patients; however, in the other cases the cataract was too extensive for this to be seen. Retinoscopically, the oil droplet appearance produced a scissors movement of the light reflex. The cone was easily seen in early cases by slit lamp examination as a posterior bulging of the posterior capsule (Figure 3).

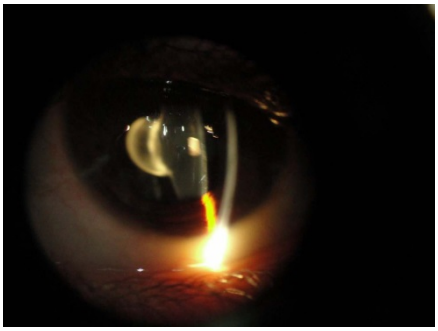

Figure 3: Posterior bulging of posterior capsule on slit lamp.

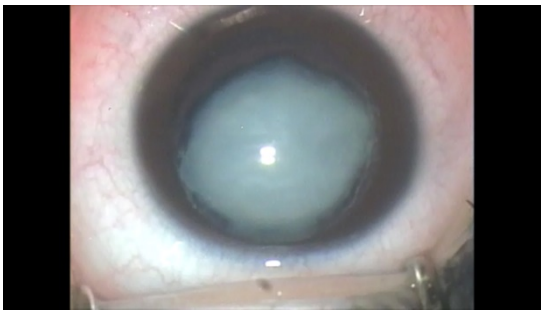

Figure 4: Irregular posterior cortical opacity of posterior lenticonus.

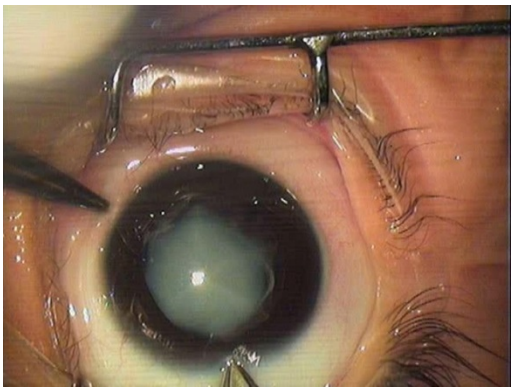

Figure 5: Another example showing the sign of irregular capsular opacity.

In cases with dense posterior subcapsular cataract and posterior cortical cataract, no definite cone could be seen. Identification of these cases as having posterior lenticonus was essential to avoid premature vitreous presentation during conventional hydrodissection or cortex removal. We observed that in these advanced case, the edge of the posterior cortical opacity was irregular and not well defined as we see in non-posterior lenticonus cases (Figures 4 and 5). In all these dense

cataract cases this sign was the clue to predict posterior lenticonus. It was positive sign in all cases. B-scan ultrasonography was done for all cases and bulging of the posterior capsule was noted in 10 eyes (Figure 6).

Figure 6: B-scan ultrasonography showing posterior lenticonus.

Postoperative follow up showed that IOLs were centered with good opening of the posterior capsule and clear visual axis.

Discussion

Posterior lenticonus is a rare condition first described by Meyer in 1888 [6]. It was reported to occur in 1 in 100 000 people [5]. In the current study, all cases were unilateral except one case with bilateral lenticonus. No family history in all our cases.

Early findings of posterior lenticonus appear ophthalmoscopically as an "oil droplet" in the central red reflex that produces pathognomonic scissors movement by retinoscopy. The posterior subcapsular and posterior cortical region develop cataract changes. The diagnosis of posterior lenticonus can be missed easily in patients with large lens opacities. Many surgeons do not discover it until they find a large defect of the posterior capsule during irrigation aspiration of cortex. Early cases of posterior lenticonus can be seen easily on slit lamp as posterior bulging of the lens. It becomes more difficult in the presence of dense cataract. We were able to predict the presence of posterior lenticonus in our cases by observing the cataract limited to the subcapsular and cortical region with an irregular edge. This irregular edge sign was present in all cases of posterior lenticonus with dense cataract. In these cases we could prepare for the possibility of an absent or thin posterior capsule. B-scan ultrasonography may help in the diagnosis by showing posterior bulging of the posterior capsule, but it is not consistent sign in all cases.

Various surgical techniques have been described for the treatment of these cases. Simons and Flynn [7] suggest a pars plana lensectomy and insertion of an IOL into the ciliary sulcus. Other authors [8,9] suggest standard phacoemulsification and IOL implantation in the bag in adults or in cases of a small lenticonus. Montanes et al. [10] implanted the haptics in the bag and the optic was situated behind the posterior capsulotomy with optic entrapment. Sukhija [8] and Montanes et al. [10] did not do hydrodissection to avoid rupture of thin posterior capsule. In our series we did multiple small areas of hydrodissection, not reaching the edge of the suspected weak or absent capsule, to facilitate removing the sticky cortex of children. The peripheral cortex was removed easily and rapidly leaving the central part to protect the weak posterior capsule. Then the central and posterior cortex was aspirated cautiously, as if peeling of the posterior capsule. In most cases the capsule was left intact so the PCIOL was implanted in the bag, followed by pars plana posterior capsulotomy and anterior vitrectomy. In 6 cases a posterior capsular defect was found at the end of the surgery; however IOL placement in the bag was possible in 4 eyes. PCIOL in the sulcus was performed when large defects occurred in the other 2 eyes. Failure to recognize posterior lenticonus will lead the surgeon to do routine hydrodissection with premature opening of posterior capsule. This may lead to vitreous prolapse, incomplete removal or dropped lens material.

Careful examination of congenital cataract is necessary to distinguish the cases with posterior lenticonus. It is easy to detect early cases of posterior lenticonus when it evident on slit lamp but it is difficult to detect the cone in advanced cases when the cataract is dense. The sign of irregular, ill-defined cataract edge was present in all advanced cases. Recognition of these cases is important for proper management and optimal visual prognosis.

What was known?

- Posterior lenticonus complicated with cataract can be easily missed. It can be unexpectedly discovered intraoperatively by finding large opening of posterior capsule.
- Management of these cases is controversial, with no definite preferred surgical technique.

What this paper adds:

- A new sign was found to recognize cases of posterior lenticonus with dense cataract.
- An improved surgical technique was described to decrease the incidence of vitreous prolapse and dropped lens material to improve the outcome.

Acknowledgments

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