Review Article

Cataract surgery in pseudoexfoliation syndrome

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Abstract
Pseudoexfoliation syndrome (PXE) is multifactorial disease genetically related and influenced by age and environmental factors, it is characterized by increase production of an elastic material and its accumulation in the intra and extraocular tissues with increasing risk of cataract. PXE represents an obstacle for surgeons during cataract surgery because of increase the risk of intraoperative and postoperative complications due to weakness of the zonules and poor pupillary dilation. So, this review outlines the approach to patient with PXE and planned for cataract surgery to avoid occurrence of complications.

Introduction
Cataract is the leading cause of blindness and vision impairment in elderly all over the world, despite, the age is the common risk factor for progression of cataract, pseudoexfoliation represents an additional hazard for nuclear sclerosis which requires cataract surgery [(1)].

Preoperative assessment:
Firstly, PXE is diagnosed using slit lamp examination where typical grayish white deposits are seen at the pupillary margin and anterior lens capsule, however, these signs may be difficult to be seen in cases of advanced cataract [(1)].
After diagnosis is established and surgery is planned, signs of decrease zonular integrity should be investigated to predict the risk of intra-operative complications as phacodonesis, lens subluxation, zonule dialysis, and a reduction of anterior chamber depth[(2)].

Iridodonesis is assessed prior to dilation which may be subtle or focal, however, phacodonesis, lens subluxation, and zonule dialysis are investigated with maximum pupil dilation[(3)]. In addition, quantitative and qualitative zonulopathy severity grading is necessary for selecting the appropriate surgical approach and devices [(4)]
Corneal evaluation by specular microscopy should be done to evaluate endothelial cell counts and morphology, cell hexagonality and coefficient of variation[(5)]. Moreover, intraocular pressure should be assessed as the prevalence of glaucoma is reported from 26% - 49% at the time of cataract surgery in PXE patients [(6)].

Intraoperative management:
PXE represents a challenge for the surgeon during phacoemulsification because of poor pupillary dilation and zonular weakness which are responsible for intraoperative complications[(7)]. A poorly dilated pupil makes capsulorhexis difficult, increases iris prolapse incidence and increases surgical risk due to poor visualization of the lens, therefore, preoperative dilating eye drops is used if not dilated, intraoperative using of a highly viscous cohesive ophthalmic viscosurgical device (OVD)[(8)]. Lysis of posterior synechiae; if present; should be done using a blunt spatula, also, iris hooks or pupillary rings can be used to ensure roomy and stable dilation during surgery if pupil not sufficiently dilated [(9)].

Zonular weakness causes difficulties on puncturing the anterior capsule due to decreased anterior capsule tension, so using a 25-gauge sharp needle bent at the tip helps performing the initial capsulotomy[(10)]. Then. propagation of the capsule tear is not easy because of the absence of usual zonule countertraction, this is overcome by using a hook or capsule retractors at the cut edge of the capsulorhexis. A 5–6 mm centered circular continuous capsulorhexis (figure 1A) is needed to ease the steps of phacoemulsification procedure, intra ocular lens (IOL) support, and implantation of devices needed in preservation of capsular stability during and post-operative [(11)].

Following capsulorhexis is completed, visco-dissection is preferred before hydrodissection (figure 1B) as it allows better corticocapsular cleavage. Some surgeons advice hydride-lineation to separate the endonucleus from epinucleus to facilitate removal of endonucleus and a safe epinuclear shell is left especially in soft lenses [12].

A capsular tension ring (CTR) is used to distend and upholds the capsular bag during all phacoemulsification steps, it also supports areas of zonular dehiscence through redistributing the zonular tension around the capsule and allows IOL centration [13]. CTR implant reduces intraoperative complications in PXE eyes in cases of mild zonulopathy. Whereas in case of severe zonular weakness, capsule retractors should be used during all steps of hydrodissection and phacoemulsification followed by the implantation of a modified CTR sutured to the sclera to assure IOL - bag complex stability [14].

Phacoemulsification technique is similar to what is done for complicated cataract cases. The chop technique uses horizontal and vertical chopping which allows less zonular stress, centering the surgical maneuvers especially in a small pupil (figure 1C). If vitreous prolapse occurs a limited anterior vitrectomy is performed and using suitable OVD helps to prevent more vitreous prolapse, avoid aspiration in the peripheral, and reduce collapse of the anterior chamber [15].

Removal of lens cortex is mostly difficult and traumatic for the capsule and zonules. The presence of a CTR helps cortex removal by stenting the capsular equator and facilitates its separation from the capsule, however cortex may be trapped behind the device, making the removal more challenging. Repeated hydration of cortex may help to soften it and facilitate its aspiration. In general, careful slow tangential stripping, as well as gentle centripetal traction, help cortical removal accurately [16].

The choice of the IOL Implanted (figure 1D) in PXE patients depends on the state and future risk of capsular instability. A one piece or three piece acrylic IOL may be implanted according to the surgeon predilection. One piece IOL is of choice in PXE patients. Toric IOLs and multifocal IOLs are not recommended due to unsatisfactory visual outcomes. Also, placing IOL in the sulcus is not preferred due to probability of decentration and posterior dislocation. If a complete capsular diaphragm is missed, any anterior chamber angle-supported IOL should not be implanted because of increased risk of glaucoma [17].

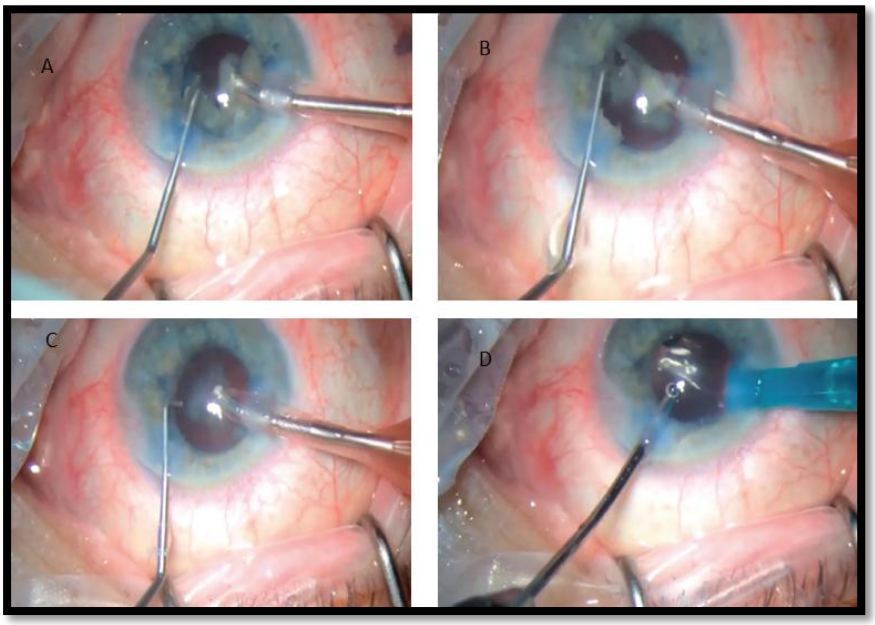

Figure (1A): an appropriately sized capsulorhexis. Figure (1B): Thorough hydrodissection is necessary to prevent further damage to the zonules. Figure (1C): When the last piece of nucleus is emulsified, the chopper is placed in front of the phaco tip to protect the posterior capsule. Figure (1D): An IOL is implanted under the irrigation quoted from https://crstodayeurope.com/wp [18].

Postoperative management:

The pathological changes occurs in PXE eyes lead to many early and late complications after cataract extraction and IOL implantation, so follow up either short term or long term after surgery is recommended [(1)].

Early Intraocular Pressure Spikes: it is more common in PXE eyes after phacoemulsification, and the risk increase with preexisting glaucoma [(19)].

Early Postoperative Inflammation: Postoperative inflammation of anterior chamber is common in PXE due to the alteration of the blood-aqueous barrier and ischemic changes in iris of these patients[(20)]

Severe postoperative fibrinous uveitis occurs frequently in PXE patients due to complicated surgery, iris manipulation and possible vitreous lossw hich resulted in synechiae, capsule contraction and cystoid macular edema [(21)].

Anterior Capsule Contraction (Phimosis): it is frequent in PXE eyes especially with aging, blood–aqueous compromise, chronic inflammation, retained cortex, material and type of IOL and compromised zonules. It is cause for IOL decentration and tilt. If postoperative phimosis develops, release should be done to decrease the risk of IOL displacement either using a neodymium: YAG (Nd:YAG) laser to relax the anterior capsule contractions by cruciate laser incisions through the circumferential anterior capsule band or surgically by a microforceps and scissors[(22)].

Corneal involvement: Abnormal endothelial cell morphology in the form of decreased endothelial cell counts and increase polymorphism is common in PXE eyes. PXE keratopathy needs penetrating keratoplasty [(23)].

Additional care must be given to protect the corneal endothelium as optimizing power modulation, decrease anterior chamber turbulence, use of suitable OVDs, and working away from the cornea [(24)].

Posterior Capsule Opacification: it is higher in eyes with PXE than normal eyes, this may be attributed to retained cortex and a defective capsule–zonular complex [(25)].

Conclusion

Cataract surgery in PXE patients has a significant risk of capsule rupture, vitreous loss and may be IOL dislocation, however with some precautions these complications could be avoided. In addition, postoperative follow up is necessary for those patients, so, satisfactory visual outcomes could be achieved in PXE patients.

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