

Pseudoexfoliation Syndrome and Cataract Surgery by Phacoemulsification

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ABSTRACT

Pseudoexfoliation syndrome is a systemic age-related disease in which abnormal extracellular material is produced and accumulates in many ocular tissues. Its ocular manifestations involve all of the structures of the anterior segment as well as conjunctiva and orbital structures. The presence of pseudoexfoliation should alert the physicians to the increased risks associated during and after cataract surgery. Increasing awareness of this condition are important in the detection and preoperative determination of patients inclined to be at greater risk for complications during surgery. Data regarding the rate of complications during phacoemulsification suggest a lower complication rate than with extracapsular extraction but still greater than in eyes without pseudoexfoliation. Despite the existence of a higher number of intraoperative and postoperative complications, experience with the phacoemulsification technique together with the improvement of the apparatus and instruments used enable similar results to obtained in eyes with pseudoexfoliation syndrome as in eyes without this pathology.

Key words: phacoemulsification, cataract, surgery, pseudoexfoliation syndrome

Introduction

Pseudoexfoliation syndrome (PEX) is an age-related disease characterized by the production and progressive accumulation of fibrillar-granular extracellular material in many ocular tissues. Its ocular manifestation involves all of the structures of the anterior segment, including the pupillary margin, the anterior lens capsule, the trabecular meshwork and the zonulla, as well as the conjunctiva and the orbital structures¹. The most consistent and important diagnostic feature of PEX are deposits of white material on the anterior lens surface. The classic pattern consists of three distinct zones that become visible when the pupil is fully dilated: a relatively homogenous central disc corresponding roughly to the diameter of the pupil; a granular, often layered, peripheral zone and clear area separating the two. Whilst the classic picture of manifest PEX has often been described, the early stages of exfoliation have not been well defined. Pigment loss from the iris sphincter region and its deposition on anterior chamber structures aid in the diagnosis. Making the diagnosis requires a careful slit-lamp examination after pupillary dilatation, and frequently goes undiagnosed, leading to unexpected problems in management and during surgery. The aware-

ness of the importance of PEX has increased considerably in recent years. PEX causes not only severe, chronic open angle glaucoma, but also lens subluxation, angle-closure glaucoma, blood-aqueous barrier impairment, serious complication during cataract surgery and it appears that it is etiologically responsible for cataract formation. Exfoliation-like fibrils have been found in many organs by electron microscopy, suggesting that PEX syndrome is a systemic disorder, long recognized only in the eye because of its visibility on slit-lamp examination and the fact that it causes glaucoma². Some reports suggest a relationship with transient ischemic attacks, stroke, heart disease, and aneurysms of the abdominal aorta^{2,3}. Despite its wide prevalence and clinical importance, the pathogenesis of PEX and exact composition of the material remain unknown. An overproduction and abnormal metabolism of glycosaminoglycans have been suggested as having one of the key roles in PEX. Based on electron microscopic identification of accumulations of exfoliation fibers in orbital tissue, specimens of the skin and visceral organs, PEX is considered to be a generalized or systemic disorder of the extracellular matrix⁴⁻⁶.

Epidemiology shows that disorder occurs in all geographic regions of the world, with varying frequency. The prevalence seems to be lower in Italy than in Finland (18.5 vs. 30.8%)⁷. PEX prevalence increases with age and is seldom observed in patients under the age of 60; however, it is not an integral process of normal aging⁸. Genetic factors predisposing to PEX susceptibility have merely begun to be explored, and no clear hereditary pattern has yet been discerned⁹. A number of nongenetic factors have also been evaluated for their possible implication in the development of PEX, including ultraviolet light, autoimmunity, slow virus infection, and trauma. It is possible that a combination of genetic and nongenetic factors may be involved in pathogenesis and etiology of PEX and it may be a multifactorial disorder¹⁰.

Pseudoexfoliative Syndrome and Cataract Surgery

Renewed interest in this long-known entity results from better awareness of the spectrum of intraocular risks during and after cataract extraction^{7,11–13}. Recent studies have shown that PEX syndrome is a systemic disorder of the extracellular matrix that involves all structures of the anterior segment of the eye². Local production and deposition of PEX syndrome fibers may lead to characteristic clinical and ultrastructural changes of the corneal endothelium (14) (PEX syndrome corneal endothelopathy), trabecular meshwork¹⁵ (capsular glaucoma), iris¹⁶ (PEX syndrome iridopathy), ciliary body¹⁷ (PEX syndrome cyclopathy), zonules¹⁸ (PEX syndrome zonulopathy), lens² (PEX syndrome phacopathy) and structures of blood-aqueous barrier^{16,19} (blood-aqueous barrier breakdown). Although all of the above-mentioned alternations may have consequences and cause complications for anterior segment surgical procedures, the most frequent and dreaded complication of PEX syndrome during cataract surgery is severe zonular weakness that may cause extensive intraoperative zonular dialysis, vitreous loss, and dislocation of the lens or of a posterior chamber intraocular lens during and after surgery^{7,11–13}. Possible consequences of reduced zonular integrity in PEX syndrome that can be seen at slit-lamp examination include phacodonesis²⁰, spontaneous dislocation of the lens into the vitreous space²¹ and anterior subluxation of the lens sometimes precipitated by the use of topical miotics leading to ciliolenticular angle-closure glaucoma²².

The incidence of phacodonesis and/or subluxation of the lens both in eyes with PEX syndrome have been reported to be between 8.4% and 10.6%^{11,20}. Furthermore, pupils of patients with PEX show a diminished response to mydriatic agents, probably for two reasons: the iris is infiltrated by the exfoliative material and consequently becomes fibrotic, and pupillary movements are mechanically restricted because the exfoliative material adheres to the iris pigment epithelium and anterior lens capsule⁷. Pupillary diameter and zonular fragility have

been suggested as the most important risk factors for capsular rupture and vitreous loss^{23,24}. Apparently, the instability of the zonular apparatus is more important than poor pupil dilatation^{7,23}. Zonular fragility increases the risk of lens dislocation, zonular dialysis or vitreous loss up to ten times²⁵. Vitreous loss has been reported to be five times more common than in patients without PEX (9.0% vs. 1.8%)²⁶, which is related to an increased incidence of zonular dialyses, lens dislocation and capsular rupture. Despite the involvement of the lens capsule in PEX, one study found no significant differences in the mean capsular thickness between the PEX eyes and normal eyes. Although the posterior capsule is of normal thickness, capsular rupture is more common in PEX eyes with an occurrence of up to 27% as compared to 2% in the control eyes². Kuchle found an intraoperative complication rate of 13.4% in the eyes with an anterior chamber depth less than 2.5 mm and 2.8% in eyes with an anterior chamber depth of 2.5 mm or more. This finding suggests that preoperative reduced anterior chamber depth indicates zonular instability and should therefore alert the surgeon to the possibility of intraocular complications related to zonular dialysis¹¹. Due to blood-aqueous barrier breakdown, PEX syndrome predisposes to the formation of synechiae between the iris pigment epithelium and the anterior lens capsule. Posterior synechiae are also more prone to form between the iris and intraocular lens (IOL) postoperatively. Posterior synechialysis or lysis of more peripheral iridocapsular adhesions and pupillary enlargement may be necessary²⁷. Due to chronic sphincter fibrosis, it is advisable not to overexert the pupil since it will often remain dilated, predisposing it to pupillary capture²⁸.

Postoperative decentrations of IOLs were reported to be significantly higher in eyes with PEX due to zonular rupture or capsular bag decentration. Lens decentration is even more common when the lens is entirely in the capsular bag, primarily due to decentration of the entire bag²⁹. Posterior chamber lenses may be implanted in the ciliary sulcus despite the presence of a small capsular break or area of zonular dehiscence, providing that enough support still exists for the implant³⁰. Postoperatively, patients with PEX are at a greater risk of developing an immediate elevation of intraocular pressure (IOP) and thus all viscoelastic should be removed from the eye at the time of surgery. Patients with extensive visual field loss or severe glaucomatous optic atrophy should have tonometry performed 4–6 hours after surgery, and an acute rise in IOP should be treated³¹. In glaucomatous patients, combined cataract and glaucoma surgery decreases the incidence of an acute postoperative rise in IOP and may improve long-term control of IOP^{31,32}. Posterior capsular opacification is increased in PEX eyes, most probably due to an aggravated blood-aqueous barrier breakdown³³. Both the increased rate of vitreous loss and the more frequent need of Nd:YAG capsulotomy for posterior capsular opacification could be expected to increase the risk of retinal detachment. The PEX syndrome might, therefore, be re-

garded as a risk factor in the development of retinal detachment².

Pseudoexfoliation Syndrome and Phacoemulsification

Improved by both technology and new surgical procedures, phacoemulsification (PHACO) of the nucleus has become increasingly important in extracapsular cataract surgery. PHACO within the capsular bag has proved to be a valuable surgical technique, yielding predominantly good results. PHACO in the presence of PEX presents surgeons with particular challenges. The frequency of intraoperative and postoperative complications such as zonular dialysis, capsular rupture, vitreous loss, and IOL decentration may be reduced with careful attention and precise surgical technique (34). Literature on the rate of complications during PHACO suggest a lower complication rate than with extracapsular extraction^{7,11,12,32,34}, but still greater than in eyes without PEX (2,11). Furthermore, PEX syndrome determines a statistically higher risk of YAG posterior capsulotomy in the first 6 months after the surgery⁷. Possible preoperative and intraoperative measures to avoid or minimize complications of cataract extraction in PEX eyes require increased awareness of potential intraoperative problem that this disease may evoke. Except for cases of very hard nuclei, controlled PHACO is the preferred surgical technique, but its use should be directed by the experience of the surgeon. Particular attention during preoperative examinations should be directed to the possible instability of the zonular apparatus. In case of marked zonular weakness, frequently anticipated before surgery by obvious phacodonesis or shallow anterior chamber, implantation of a polymethylmethacrylate capsule-supporting ring may allow for safe operation³⁵. In case of pronounced phacodonesis with subluxation of the lens, an intracapsular cataract extraction

followed by posterior chamber lens implantation fixed by trans-scleral sutures may be necessary.

Capsulorhexis should be performed within the limits of the outer ring: in this way, visualisation of the rhexis is enhanced, thus reducing the risk of rhexis dehiscences. Whilst this situation may lead to a small rhexis, it does prevent the nucleus »swing« inside the bag. If the pupil is inadequately dilated it is suggested to make the anterior chamber deeper by viscoelastic, and then only if necessary, to perform the pupil stretch technique. The pupillary stiffness and the pressure exerted by the viscoelastic substance enhance stability of the lens and maintenance of a mydriasis that is considered satisfactory in most of the eyes. The choice of IOL is also important in eyes with PEX syndrome. Heparin surface modified posterior chamber IOLs were found to be associated with fewer postoperative fibrinoid reactions, less frequent pigment and cellular deposits on the lenses and lower incidence of posterior synechia formation than other forms of IOLs²⁷. Furthermore, flexible silicone IOLs should not be used to prevent capsular contraction syndrome and valuating of intraocular lens optic². After surgery, frequent and thorough follow-up examinations are imperative for the early detection and treatment of complications such as IOP elevation, formation of synechiae or formation of fibrin.

Conclusion

Data regarding the rate of complications during PHACO suggest a lower complication rate than with extracapsular extraction but still greater than in eyes without PEX syndrome. Despite the existence of a higher number of intraoperative and postoperative complications, experience with the PHACO technique together with the improvement of the apparatus and instruments used enable similar results to be obtained in eyes with PEX syndrome as in eyes without this pathology.

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PSEUDOEKSFOLIJATIVNI SINDROM I OPERACIJA KATARAKTE FAKOEMULZIFIKACIJOM

SAŽETAK

Pseudoeksfolijativni sindrom je sustavna bolest vezana za dob, a odlikuje je stvaranje i nakupljanje abnormalne stanične tvari u različitim očnim tkivima. Bolest se očituje na svim strukturama prednjeg segmenta oka, kao i na spojnici i tkivima orbite. Nazočnost ove bolesti ukazuje na povećani rizik razvoja komplikacija tijekom i nakon operacije katarakte. Prema dosadašnjim podacima učestalost komplikacija operacije katarakte u očiju s pseudoeksfolijativnim sindromom fakoemulzifikacijom niža je u odnosu na ekstrakapsularnu ekstrakciju leće, iako je još uvijek viša nego kod očiju bez pseudoeksfolijativnog sindroma. Većim iskustvom u izvođenju operacije katarakte fakoemulzifikacijom te usavršavanjem instrumenata i aparata, u očiju s pseudoeksfolijativnim sindromom mogu se očekivati slični postoperativni rezultati kao i u očiju u kojih pseudoeksfolijativni sindrom nije nazočan.