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Age-Related True Exfoliation of the Lens Capsule: Phacoemulsification Surgery Results

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Key Words
True exfoliation · Lens · Cataract · Phacoemulsification

Abstract
Historically associated with glassblowers, true exfoliation of the crystalline lens involves a splitting or delamination of the capsule. We reviewed the phacoemulsification records of a single surgeon for patients with true exfoliation of the lens capsule. The incidence in our series was 2.2% (6 in 278 cases). The average age was 85.0 years. All patients had successful phacoemulsification outcomes, which may have been due to accurate recognition of the condition and appropriate surgical planning. Our findings support the notion that true exfoliation may be more often associated with advanced age rather than infrared radiation.

Introduction

True exfoliation of the crystalline lens involves a splitting or delamination of the capsule. Historically, the entity has also been referred to as glassblower's cataract due to the known risk of prolonged exposure to heat and infrared radiation causing the condition [1]. Other ocular conditions associated with true exfoliation include age, trauma, uveitis and glaucoma [2, 3]. Awareness of the link between advanced age and true exfoliation of the lens capsule has grown. For example, using transmission electron microscopy, one study found age-related ultrastructural changes in a patient with true exfoliation of the lens capsule [4].

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In this case series, we report 6 patients who underwent phacoemulsification with true exfoliation of the lens capsule. We recognized the true exfoliation either preoperatively with a rolled up and/or split anterior capsule or intraoperatively by the double-ring sign [1]. All cases were performed under local anesthesia by the same surgeon (J.C.C.Y.) over a 6-month period, out of a total of 278 cases performed during the same period.

Case Presentations

The clinical characteristics of the 6 patients are summarized in table 1. The average age of our series was 85.0 ± 7.1 years (range: 72–92). None of the patients had a history of prolonged infrared radiation or heat exposure, trauma or ocular inflammation. In 3 patients (cases 1, 5, 6), the clinical diagnosis of true exfoliation was made preoperatively by slit-lamp examination (fig. 1). The anterior capsules were sent for histological confirmation in cases 1 and 6 (fig. 2). In the other 3 patients (cases 2, 3, 4), the condition was only recognized intraoperatively, and we did not arrange for histological examination of the capsulorhexis specimen (fig. 3). In 2 patients (cases 1 and 5), the fellow eye was pseudophakic with previous cataract operation done elsewhere, thus we could not ascertain if true exfoliation was present. Of the other 4 patients, true exfoliation of the lens capsule was present bilaterally in 1 patient (case 6) and in only one eye of the other 3 patients.

Case 1

An 89-year-old female with no prior ophthalmic history presented with cataracts. Preoperatively, a delaminated anterior capsule floating in the anterior chamber was seen on slit-lamp biomicroscopy (fig. 1a), and the edge of delamination could be seen on retroillumination (fig. 1b). She underwent phacoemulsification, which required Tryptan blue (Vision Blue, 0.1%; DORC International BV, Zuidland, The Netherlands) staining of the capsule. Histology of the anterior capsule revealed areas of focal splitting (fig. 2a).

Case 2

The second patient was also an 89-year-old female. Preoperatively, she had old corneal scars, but the patient denied any history of ocular trauma or inflammation, and no specific cause of the corneal scars could be identified. The delaminated anterior lens capsule was noticed intraoperatively. Phacoemulsification was performed uneventfully without the need for capsular stains. Postoperatively, she was noted to have progressive increase in the cup-disc ratio but a normal intraocular pressure. She was subsequently diagnosed with normal tension glaucoma.

Case 3

An 85-year-old male presented with good past ophthalmic history. A partially delaminated anterior lens capsule was found intraoperatively. He had uneventful phacoemulsification without the need of capsular staining.

Case 4

A 72-year-old male had a bi-temporal hemianopia due to a pituitary macroadenoma. His history was otherwise unremarkable. He underwent uneventful phacoemulsification without needing any capsular staining.

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Case 5

An 83-year-old female presented with cataracts. Several years before, she had an acute angle-closure attack and had undergone laser iridotomy. Her intraocular pressure was normal since the angle-closure episode and did not require glaucoma medication. A delaminated anterior lens capsule was diagnosed preoperatively. She underwent uneventful phacoemulsification. Adequate visualization of the lens capsule required capsular stain. Figure 3 shows the double-ring sign, which was noted intraoperatively.

Case 6

A 92-year-old male with no prior ophthalmic history presented with a dense cataract. Delaminated anterior lens capsules were found in both eyes. Uneventful phacoemulsification was performed. Histology of the anterior capsule confirmed the diagnosis of true exfoliation (fig. 2b).

Discussion

We report on a series of patients of advanced age with true exfoliation of the lens capsule. The incidence in our single-surgeon single-center series was 2.2% (6 in 278 cases). To determine the true incidence of this rare condition would require a large, prospective study over a longer timeframe. The condition was consistent with an age-related etiology, as no other causes, such as a history of occupational exposure to prolonged heat or infrared radiation, were identified. Age-related degeneration of the lens epithelial cells could comprise the integrity of the lens capsule, causing vesicles to form. When the vesicles coalesce to form larger vacuoles, delamination of the anterior portion of the capsule may occur, which results in true exfoliation [4–6]. Wong et al. [2] recently reported a series of Chinese patients with a similar age-related pattern of true exfoliation. The authors also reported an association of the condition with glaucoma, with 29.2% of cases having preexisting chronic glaucoma and 20.8% with a prior history of laser iridotomy. In our series, 1 patient (case 5) had a history of acute angle closure with subsequent laser iridotomy performed.

Generally, one would expect bilateral occurrence in an age-related condition. Of the 4 patients with a phakic fellow eye, one had bilateral involvement while the other 3 had unilateral involvement. However, the possibility of asymmetry between the eyes exists. To exclude the possibility of subclinical involvement in the fellow eye, the lens capsule could be sent for histological examination in future cataract surgery. The remaining 2 patients had pseudophakic fellow eyes at the time of presentation; thus, in these cases the possibility of true exfoliation in the fellow eye could not be determined.

The average age of our series was 85.0 years, which was among the oldest reported in the literature. The reported age of true exfoliation ranged from 80.5 to 85.4 years [2, 7–9]. Cashwell et al. [7] reported 11 cases of presumed age-related true exfoliation cases with the highest average age of 85.4 years. With regard to the geographical distribution of true exfoliation, authors have reported case series from different continents, including Asia, North America and Europe. The relatively small number of patients reported in the literature precludes meaningful analysis of the role that geography may play in the development of true exfoliation.

In our series, the diagnosis of true exfoliation was made by recognizing the double-ring sign and/or the presence of a split or rolled-up anterior capsule, which was strongly associated with true exfoliation [1, 5, 6, 9–11]. In 2 patients from our series, the anterior capsule had histological confirmation of the true exfoliation syndrome, with delamination and rolling

up of the anterior capsule. The double-ring sign was due to partial splitting of the anterior capsule, where two complete continuous curvilinear capsulorhexis (CCC) procedures may be required in a single operation to achieve complete capsulorhexis [8]. Ataka et al. [5] demonstrated on electron microscopy that the anterior lens capsule in cases with the double-ring sign had horizontal splits in the anterior capsule, with concomitant abnormal epithelial changes including vacuole-like spaces and widened intercellular spaces, which supported the relation with true exfoliation.

The presence of true exfoliation of the lens capsule typically increases the risk of capsule complications such as radial extension and radial tear [9, 11, 12]. In our case series, all cases of age-related true exfoliation syndrome had a good outcome from phacoemulsification and capsular implantation of the intraocular lens. Kulkarni et al. [13] reported good safety outcomes from phacoemulsification in 5 cases with true exfoliation due to a history of prolonged heat exposure. Wong et al. [2] also noted good outcomes in their series. In all of our patients, the CCC was successfully performed, although in 2 cases capsular staining with Tryptan blue dye was required. The dye helped ensure the CCC was of full thickness instead of partial thickness.

We believe recognizing the true exfoliation, either preoperatively (splitting of the anterior capsule on slit-lamp biomicroscopy) or intraoperatively (double-ring sign), was the key to our good surgical outcome without any capsular complications. To detect true exfoliation syndrome preoperatively, image modalities such as the Pentacam or anterior-segment optical coherence tomography may also be employed [6, 14, 15]. To our knowledge, our series composed age-related true exfoliation cases with one of the oldest average ages reported. All cases demonstrated a good surgical outcome due to prompt recognition of the condition.

Statement of Ethics

The patients signed an informed consent before the surgery.

Disclosure Statement

The authors have no financial or proprietary conflicts of interest concerning the materials discussed in this article.

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Table 1. Clinical characteristics of 6 patients with true lens exfoliation

Case	Age, years	Sex	OD/OS	Medical history	Cataract grading	Fellow eye lens status	Past ocular history	Preop. VA	Postop. VA	Time of diagnosis of true exfoliation	Intraoperative complications and remarks
1	89	F	OS	Hypertension, atrial fibrillation	NS+CC++	Pseudophakic	Nil	0.3	0.7	Preoperative Confirmed on histology	Nil Vision blue-aided capsulorhexis
2	89	F	OS	Hypertension	NS++CC++	No lens exfoliation	Faint corneal scars (denies history of trauma) Normal tension glaucoma diagnosed after cataract surgery	0.2	0.5	Intraoperative	Nil
3	85	M	OD	Hypertension, gout, ischemic heart disease, hyperlipidemia	NS++CC++	No lens exfoliation	Nil	0.3	0.6	Intraoperative	Nil
4	72	M	OS	Hypertension, hyperlipidemia	NS++	No lens exfoliation	Bi-temporal visual field defect due to pituitary macroadenoma	0.1	0.7	Intraoperative	Nil
5	83	F	OS	Breast carcinoma (in remission)	NS++CC++	Pseudophakic	History of acute angle closure with laser peripheral iridotomy performed. Normal intraocular pressure	0.2	0.7	Preoperative	Nil Vision blue-aided capsulorhexis
6	92	M	OS	Hypertension	NS+++-PSC++	True exfoliation	Nil	0.03	0.7	Preoperative Confirmed on histology	Nil

NS = Nuclear sclerosis; CC = cortical cataract; PSC = posterior subcapsular cataract. VA = Visual acuity, in Snellen decimal notation; Preop. = preoperative; Postop. = postoperative.

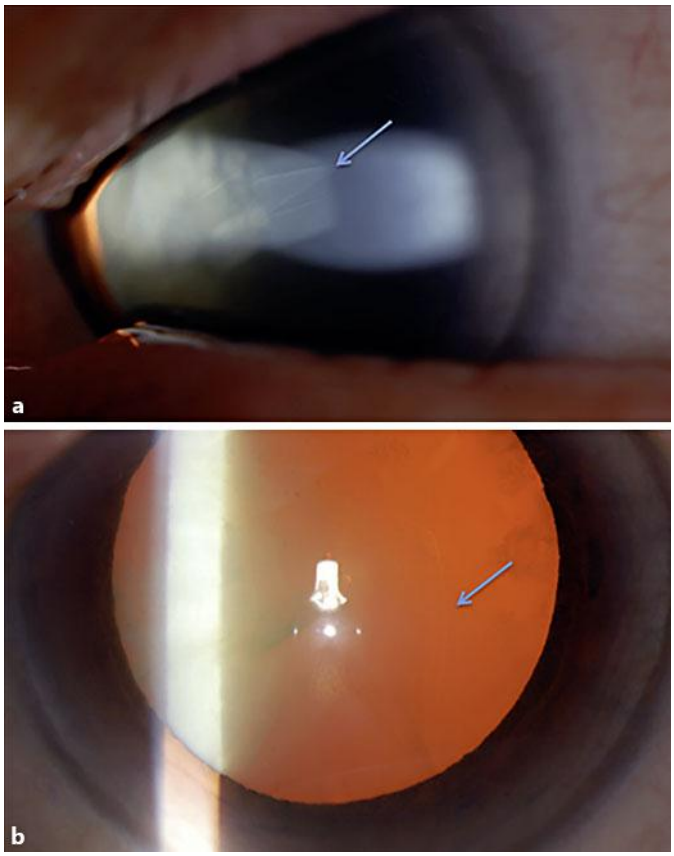

Fig. 1. **a** Slit-lamp biomicroscopic image showing a delaminated anterior lens capsule (arrow) floating in the anterior chamber. **b** The edge of delamination (arrow) as seen on retroillumination.

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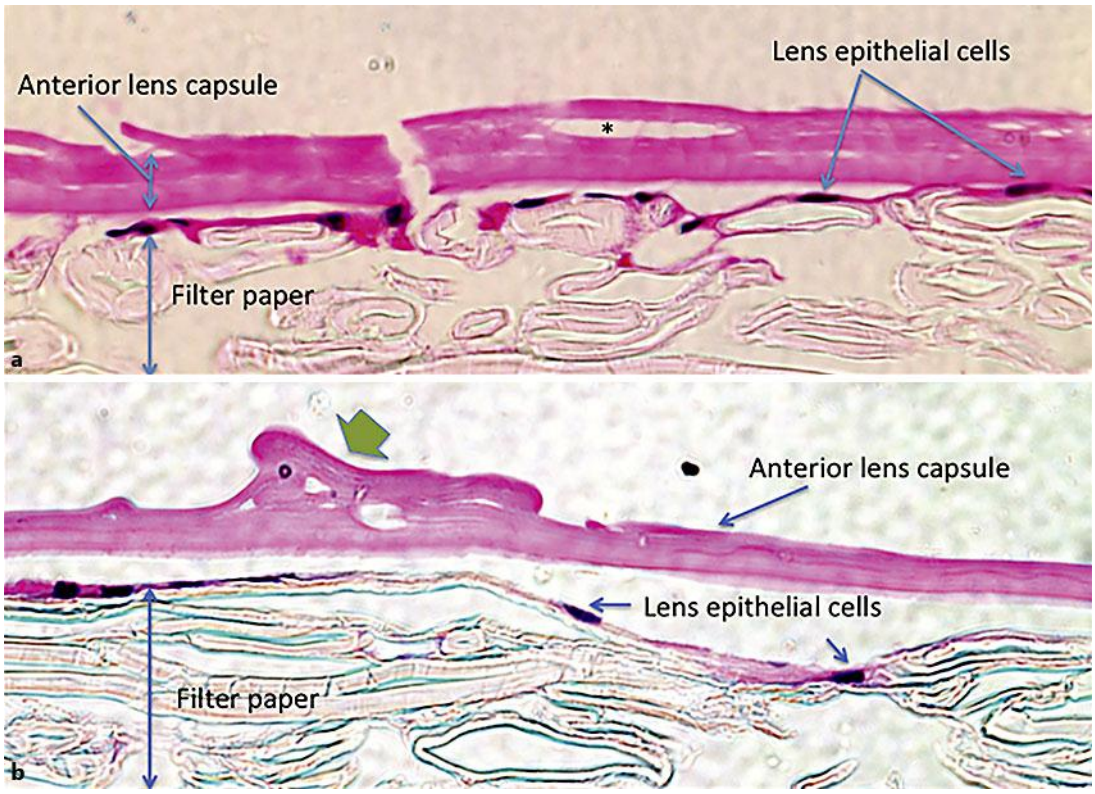

Fig. 2. Photomicrographs of true exfoliation of the anterior lens capsule from the capsulorhexis during phacoemulsification surgery. In both images, normal lens epithelial cells appear along the posterior surface of the anterior lens capsule. Filter paper was used to obtain a flat mount of the lens capsule. **a** Lamellar splitting along the anterior lens capsule is seen (HE. ×40). **b** A scroll of the anterior lens capsule is shown (green arrow; HE. ×20).

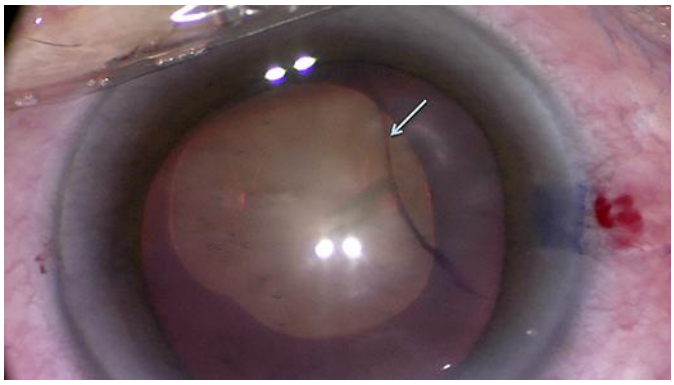

Fig. 3. Intraoperative image showing the double-ring sign in case 5. After completion of the capsulorhexis aided by Tryptan blue stain, the double-ring sign (arrow) was noted with another layer of anterior capsule remaining. A second capsulorhexis procedure was required.