

Multiple Layers Lens Capsule True Exfoliation in Patient with Primary Angle Closure Glaucoma

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Received: June 12, 2012 / Published online: August 21, 2012
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ABSTRACT

Introduction: True exfoliation of lens capsule is a relatively rare disorder that has a thin, fluttering membrane in the anterior chamber, mainly located in the pupil area, which is believed to have split from the anterior lens capsule.

Case Report: The authors present here a patient with true exfoliation and primary angle closure glaucoma in both eyes, the ultrastructure of one eye revealed that the delaminated membrane was composed of more than two layers in itself.

Conclusion: Lens capsule true exfoliation could happen in patient with primary angle closure glaucoma, with more than two layers in itself.

Keywords: Glaucoma; Lens capsule; True exfoliation; Ultrasound biomicroscopy; Ultrastructure

INTRODUCTION

True exfoliation of the lens capsule is a relatively rare disorder, which was first reported by Elschnig et al. [1] in 1922. The clinical manifestation has been described as a thin, fluttering membrane in the anterior chamber, mainly located in the pupil area, which is believed to have split from the anterior lens capsule. The pathogenesis of this disorder is not clear; traditionally it has been attributed to infrared radiation, exposure to ultraviolet light, inflammation, trauma, and idiopathic causes. Another risk factor is aging [1–16]. At present little is known about the pathology of the membrane splitting from the lens capsule, and the relationship between this disorder and glaucoma is ambiguous. Here the authors describe a case of bilateral true exfoliation of the lens capsule with primary angle closure glaucoma.

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CASE REPORT

General Information

The authors present a 74-year-old female farmer from a mountain village of Hubei province, China, with no history of excessive exposure to heat or surgery, no family history of this disorder or glaucoma. This study was carried out following informed consent from the patient, and the tissue was handled according to the Declaration of Helsinki.

Clinical Examination

The best corrected visual acuity was finger counting at 0.05 m in the right eye and 12/20 in the left eye, the vision loss of both eyes

could not be corrected with glasses. The intraocular pressure was 60 mmHg in the right eye and 17 mmHg in the left eye. Slit-lamp examination revealed that the right cornea was edematous and the anterior chambers of both eyes were shallow. The right pupil was ellipse and fixed, dilated to 7 mm, the diameter of the left pupil was 4 mm. Both lenses had moderate opacity and this was more serious in the right eye. The cup/disc ratio was 0.5 in the right eye and 0.3 in the left eye. Clear, diaphanous membranes were seen in both anterior chambers (Fig. 1). The capsular peeling was limited to the central anterior capsule. There was no iris transillumination, pseudoexfoliation, or phacodonesis, which may be accompanied with lens capsule true exfoliation. Gonioscopy of the left eye revealed

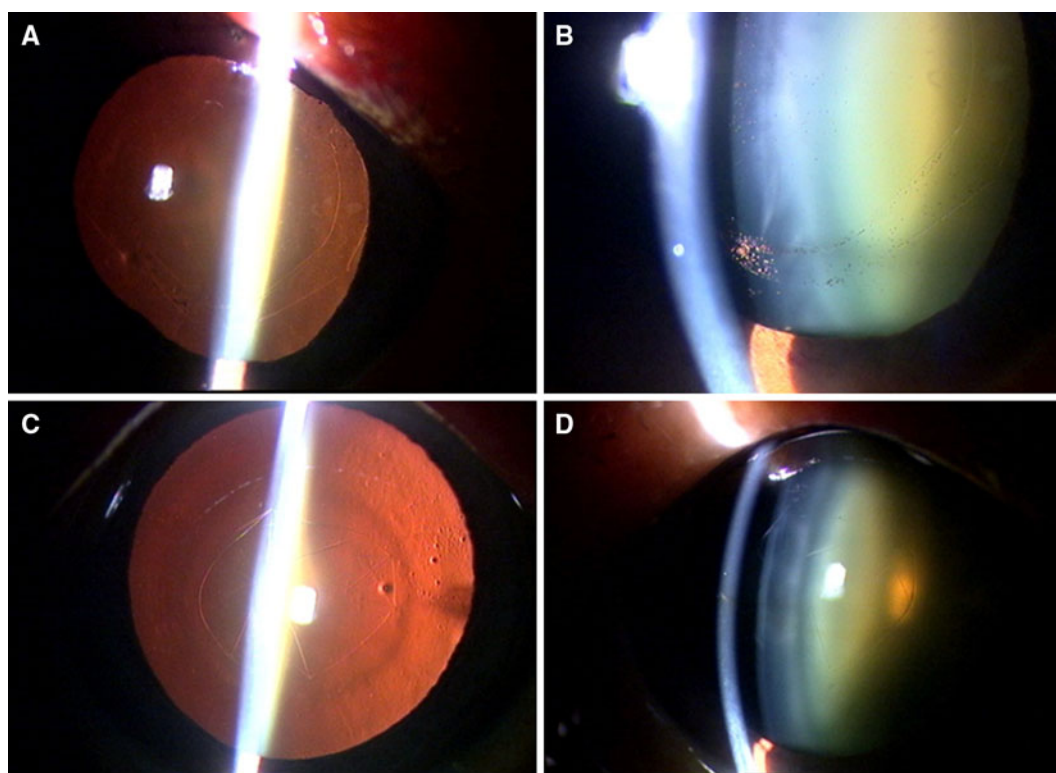


Fig. 1 Lens capsule true exfoliation under slit lamp microscopy. Clear, diaphanous capsular membranes were seen in both anterior chambers superior to the central anterior lens capsule (slit-lamp). The right eye, with a

dilated pupil as a result of an attack of primary angle closure glaucoma (a, b). The left eye, with pupil was dilated for phacoemulsification cataract extraction (c, d)

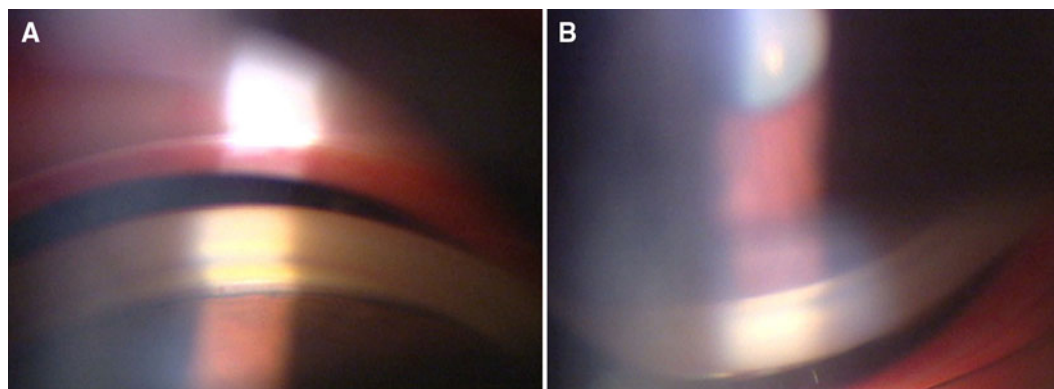


Fig. 2 Closed anterior chamber angle under gonioscopy. Gonioscopy of the left eye revealed a narrow but open angle at 6 o'clock (a) and a closed angle at 12 o'clock (b)

a narrow open angle at 6 o'clock and a closed angle at 12 o'clock (Fig. 2). Ultrasound biomicroscopy (UBM, SW-2100; Tianjin Suwei Electronic Technology Co., Ltd., China) demonstrated a membrane inserting into the anterior chamber from the surface of the anterior lens capsule (Fig. 3).

Diagnosis and Treatment

The major diagnosis was primary angle closure glaucoma in the right eye and eclipse period in the left eye. The authors prescribed mannitol, 1% pilocarpine and 2% mikelan eye drops, after which the intraocular pressure of the right eye dropped to 17 mmHg within 24 h, the corneal edema had decreased under the slit-lamp microscope. After a few days, the patient underwent a non eventful trabeculectomy in her right eye, and a cataract phacoemulsification with intraocular lens implantation in her left eye. The split membrane and the central anterior capsule of the left eye was extracted separately and fixed with 2.5% glutaraldehyde immediately after surgery, submitted for ultrastructural pathological examination (transmission electron

microscopy [TEM]: FEI Tecnai G2 12; FEI, Hillsboro, OR, USA).

RESULTS

Outcome of the Surgery

The best corrected visual acuity of the right eye returned to 10/20 and the left eye to 16/20 within a 3-months follow-up period. Because the cornea endothelial cell density of the right eye decreased to 657 cells/mm², the lens of the right eye received no further surgery. The intraocular pressure of both eyes was in the normal range, which was between 11 and 21 mmHg, and the right corneal edema subsided in the follow-up period of 12 months.

Light Microscopy

Light microscopy (Fig. 4) revealed that the membrane superior to the anterior lens capsule appeared as one layer in some parts and separated into two or three layers in others. The anterior lens capsule was always one layer, with normal lens epithelium.

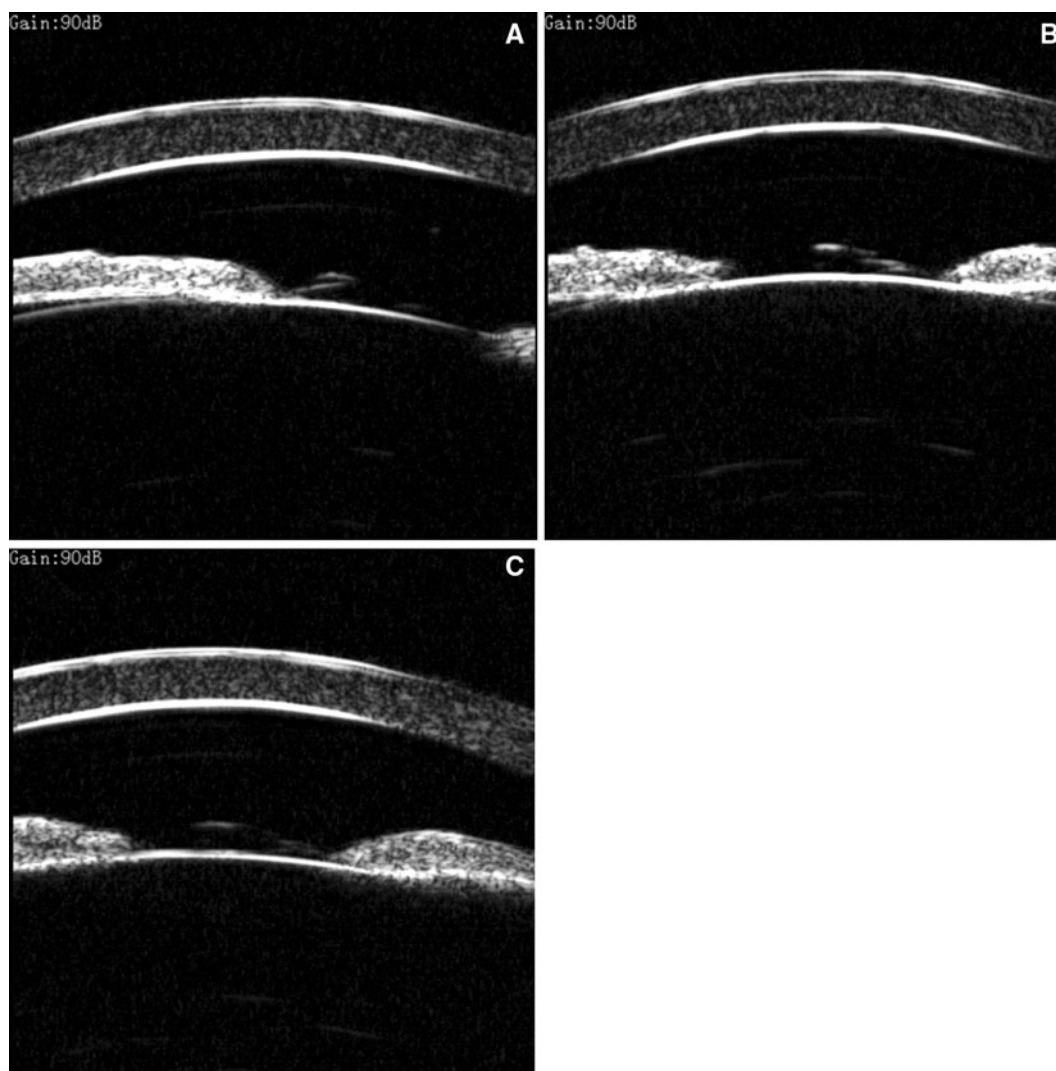


Fig. 3 Lens capsule true exfoliation under ultrasound biomicroscopy. Ultrasound biomicroscopy (UBM,SW-2100) demonstrating a membrane inserting into the anterior chamber from the anterior lens capsule in the left eye (a–c)

Transmission Electron Microscopy

TEM revealed an interesting phenomenon that has never been described before. Figure 5 shows that the so-called split membrane is composed of more than two layers in itself. With a total thickness of 12.5–18 μm , each layer was approximately 5–6 μm , composed of homogeneous granular material that was arranged in a linear or band pattern. A cleavage plane was present where the layers had separated. Figure 6 shows that the anterior lens

capsule is normal, with a thickness of 16 μm and normal epithelial cells.

DISCUSSION

The Relationship Between Glaucoma and True Exfoliation

Unlike pseudoexfoliation, a causative factor of open angle glaucoma, the relationship between true exfoliation and glaucoma is ambiguous.

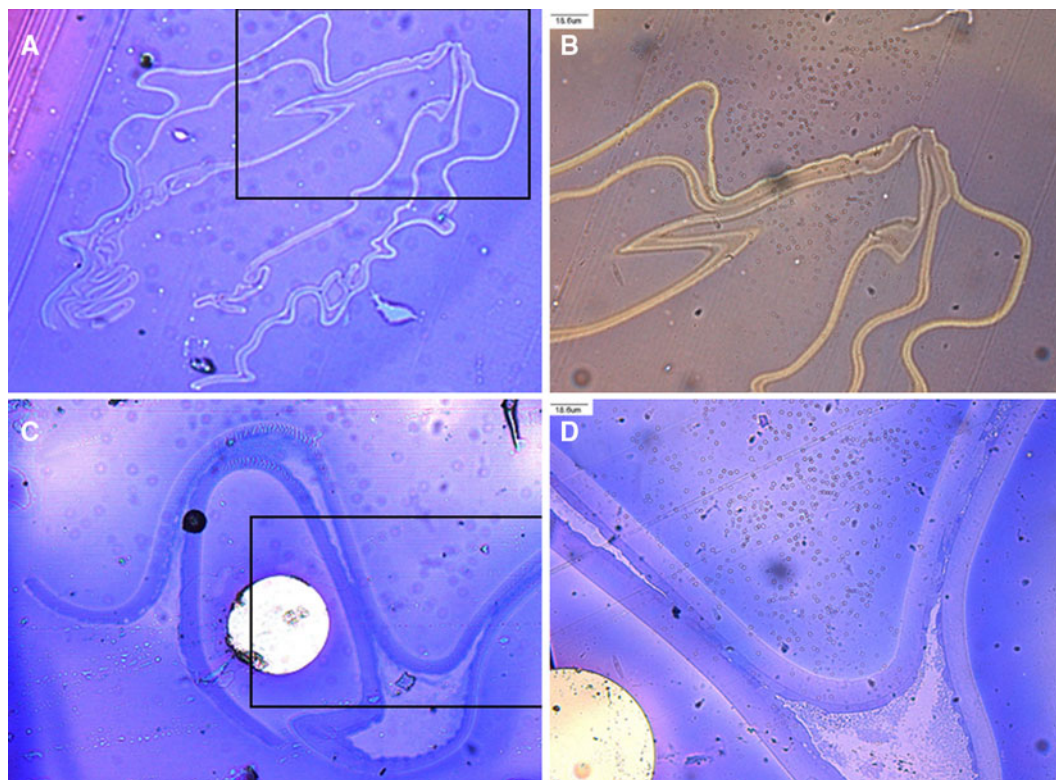


Fig. 4 Lens capsule true exfoliation and remaining capsule under light microscopy. Light microscopy of the membranes superior to the anterior lens capsule (**a, b**) and anterior lens capsule (**c, d**), (**b, d**) is the amplification of part of (**a, c**), respectively. The membrane superior to the anterior lens capsule united into one layer in some parts and separated into two or three layers in others.

However, these two disorders may occur in the same eye simultaneously. The patient reported here was found to have primary angle closure glaucoma as her primary problem, with true exfoliation as an incidental finding. True exfoliation was first distinguished from pseudoexfoliation by Dvorak-Theobald [17] in 1953, in which fibrillar material may deposit on the lens, zonules, and trabecular meshwork, which can lead to open angle glaucoma [18–21]. What is more, true exfoliation and pseudoexfoliation of the anterior lens capsule had once been found simultaneously in the same eye [16].

The anterior lens capsule was always one layer, with normal lens epithelium. Most importantly, no step was found in the anterior lens capsule, with a diameter of 5 mm, which was wider than that of the split area of the anterior lens capsule; this may mean that the so-called split membrane superior to the anterior lens capsule is not really generated from the anterior lens capsule, but from the aqueous humor

The Cause and Origin of True Exfoliation

The cause of true exfoliation remains obscure, mainly because this disorder has not been duplicated in an animal model. According to the information from clinical data, these processes possibly have an underlying causality from intense infrared radiation exposure. Traditionally it has been thought to be caused by heat or infrared-related changes in the anterior capsule. It has also been associated with trauma, intraocular inflammation and idiopathic causes [1–16]. Age-related changes in the lens may also play a role in true

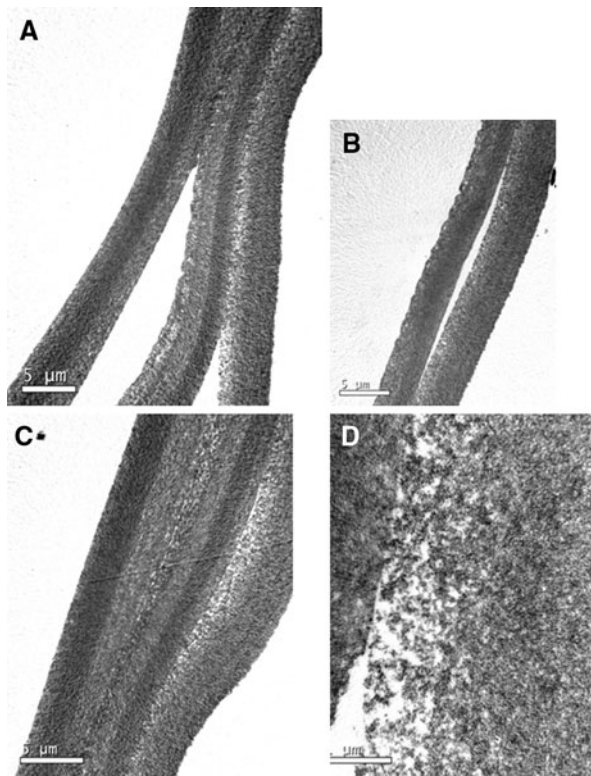


Fig. 5 Lens capsule true exfoliation under transmission electron microscopy. Transmission electron microscopy of the membrane superior to the anterior capsule revealed that the so-called split membrane is composed of two or three layers in itself (a, b). With a total thickness of 12.5–18 μm , each layer is approximately 5–6 μm (a–c), composed of homogeneous granular material that was arranged in a linear or band pattern. A cleavage plane was present where the layers had separated (d), which was the amplification of part of (a)

exfoliation of the lens because idiopathic true exfoliation in the absence of history appears more common in older patients [4, 6–8, 12].

Anderson and van Bockxmeer [14] and Seland [22] found that the electrophoretic patterns of the delaminated capsule were different from age-matched normal capsules, and hypothesized that capsular protein abnormality might play a role. The possibility of heat-activated proteolysis has also been suggested as a cause of capsular delamination; delamination of the anterior lamella might

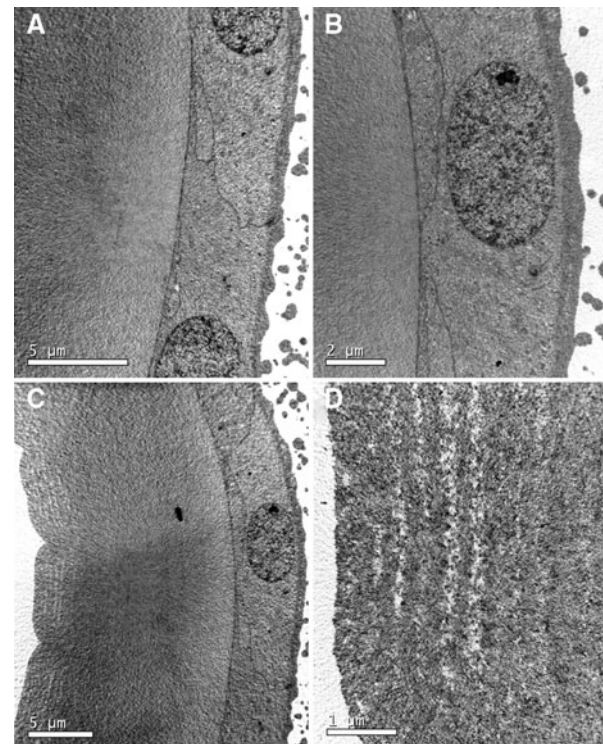


Fig. 6 The remaining capsule and epithelial cells under transmission electron microscopy. Transmission electron microscopy of the anterior lens capsule revealed a normal thickness of 16 μm and normal lens epithelial cells (a–c), composed of homogeneous granular material that was arranged in a linear or band pattern; (d) is the amplification of part of (c)

develop as a result of degeneration of the fibrillar constituents of the membrane or damage to the interfibrillar cement [2]. Brodrick and Tate [5] noted an ultrastructural change in the epithelium of the delaminated capsule, and suggested that their finding of degranulated and dilated endoplasmic reticulum with clumped nuclear chromatin might indicate a cellular abnormality as a potential underlying cause of the scrolling.

In this patient, there was no history of exposure to excessive heat, intense infrared radiation, uveitis, or trauma. This should be grouped into idiopathic true exfoliation in the left eye, although the authors could not ignore the fact

that the attack of angle closure glaucoma induced inflammation. The thickness, ultrastructure and lens epithelium of the remaining anterior capsule was higher than that of the age-matched normal capsules as reported previously, which should be approximately 8 μm for 65-year-old humans [22–24]. More interestingly, the delaminated membrane separated into two or three layers in itself, with a thickness of 5–6 μm in each layer, and a total thickness of 12.5–18 μm .

CONCLUSION

Lens capsule true exfoliation could occur in patients with primary angle closure glaucoma, with more than two layers in itself. The thickness and electron density of the remaining capsule under TEM is relatively normal when compared with age-matched capsules from senile cataract. There was no abnormality in the lens epithelial cell either.

ACKNOWLEDGMENTS

Zhiqi Chen, MD, Ruilin Wang MD, Fuzhi Deng, and Juan Huang assisted in the surgery and pathobiology process. Dr. G. Li is the guarantor for this article, and takes responsibility for the integrity of the work as a whole. This research was funded by the Science Foundation of Hubei Province, conferred to X.L. (no. 2008CDB214).

Conflict of interest. None.

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