Short communication

Lenticonus diagnosis in Alport’s syndrome: Anterior capsule apical angle calculation using Scheimpflug imagery

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ABSTRACT

Objective: We describe a simplified method to detect anterior lenticonus. Three eyes of 2 patients with anterior lenticonus, plus 16 eyes from 16 healthy controls underwent Scheimpflug imaging of their anterior segment with Pentacam. The anterior capsule apex angle was manually identified and automatically measured by AutoCAD.

Results: The mean angle was 173.06° (SD: 1.91) in healthy subjects, and 158.33° (SD: 3.05) in anterior lenticonus eyes. The angle obtained from patients was more than 3 SD steeper than those from healthy subjects.

Conclusions: The apical angle calculation method seems to discriminate well between normal eyes and eyes suspected of having anterior lenticonus.

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Diagnóstico del lenticono en el síndrome de Alport: cálculo del ángulo apical de la cápsula anterior del cristalino mediante imágenes Scheimpflug

RESUMEN

Objetivo: Se describe un método simplificado para detectar lenticiono anterior. Realizamos imágenes Scheimpflug del segmento anterior con Pentacam a 3 ojos de 2 pacientes con lenticiono anterior y 16 ojos de controles sanos. Se identificó el ángulo del ápex de la cápsula anterior y se midió con AutoCAD.

Palabras clave:
Lenticono anterior
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Introduction

Anterior lenticonus is an infrequent condition in which the anterior portion of the anterior lens and the underlying corneal protrude anteriorly due to a genetic defect in the synthesis of type IV collagen. It is frequently associated with Alport’s syndrome, a hereditary nephritis accompanied by ocular anomalies and neurosensory deafness.

The development of the protrusion gives rise to irregular astigmatism, myopia and high order aberrations which sometimes cannot be corrected with spectacles or contact lenses.

As changes in the environment are generally subtle, anterior lenticonus may not be detected in ophthalmological examination. Scheimpflug imaging technology is very useful to analyze anterior segment morphology. This paper presents 2 patients with lenticonus to describe a diagnostic by means of quantitative Scheimpflug analysis.

Case reports

Case 1

Male, 25, diagnosed with Alport’s syndrome due to kidney disease who visited due to progressive bilateral loss of vision. Best corrected visual acuity (BCVA) was 0.49 logMAR in right eye (RE) and 1 logMAR in left eye (LE). Refraction: −5.15 to 168° RE and −10.4 to 176° LE. Biomicroscopy suggested the presence of bilateral anterior lenticona.

Case 2

Male, 52, diagnosed with Alport’s syndrome, had 3 kidney transplants in addition to LE cataract operation several years ago, visited to request treatment for RE. BCVA in RE: 1.30 logMAR and in LE: 0.20 logMAR. Examination revealed anterior lenticonus with right posterior capsular opacity and left pseudophakia.

Scheimpflug anterior segment images were taken (Pentacam, Oculus, USA) of the 3 eyes (cases 1 and 2) and of 16 eyes of 16 healthy controls between 20 and 60 years old after pupil dilatation (2 drops of cycloplicic Coliricusi eyedrops at 1% by Alcon Cusi Laboratorios). All the subjects accepted the examination and signed informed consents. The study was reviewed and approved by the Ethics Committee of the Ramón y Cajal Hospital (Madrid) and carried out in accordance with the criteria established in the Helsinki declaration. A standard Scheimpflug image was selected (image 183-3). The images were exported and analyzed with the AutoCAD graphic design application (Audodesk, Inc., California, USA). The apex of the anterior capsule was identified and a tangent line projected manually from the capsule at each side. These lines made up an angle that was automatically measured by the AutoCAD application (Fig. 1).

The mean angle measurements were 173.06° (standard deviation [SD]: 1.91) in checkups and 158.33° (SD: 3.05) in anterior lenticona. The mean apical angle of the lenticona was more acute, over 3 SD, than that of controls.

Discussion

The above cases illustrate the usefulness of the Scheimpflug technology to detect anterior lenticona with a simple method that calculates the anterior capsule apex angle. Early diagnostic would enable rehabilitation of these patients’ vision and prevent and treat the amblyopia generated by irregular astigmatism in developing eyes.

Routine ophthalmological exploration was the only tool for detecting lenticone but does not reveal subcutaneous anomalies. With biomicroscopy it is possible to observe anterior chamber protrusion, which is sometimes opacified. Retinoscopy shows an “oil drop” image.

Wavefront analysis is of help in screening lenticone. Analyzing the difference between high order aberrations derived from the wavefront analysis of the high order aberrations map of corneal topography, it is possible to differentiate the corneal or lens origin. A markedly negative spherical aberration indicates anterior surface protrusion, although it requires morphometric measurements.

Lens anterior pole Scheimpflug images can show anterior lenticone, although qualitative analysis on its own is subjective and not very precise. Quantitative analysis of the protrusion provides more objective and earlier data. In a single case, Xu measures apex protrusion from a 4 mm long line that is manually drawn. This measure is over 3 SD larger in lenticone than in the normal population.

A different and simpler approach is proposed: the angle formed by the protrusion is measured, finding that it is over 3 SD more acute in lenticone than in healthy controls. This method seems less subjective and is based on the measures of 3 eyes of 2 patients, although it requires validation with additional cases.

However, this process has the drawback that it cannot be performed with Pentacam software, which means that the image has to be exported. Even so, after validation it would be easy to include said process.
It would be interesting to assess whether this method can detect pre-clinic anterior lenticone in patients diagnosed with Alport syndrome as kidney alterations and diagnostic usually appear before visual impairment.

New imaging technologies allow us to perform qualitative and quantitative analysis of the crystalline lens in order to detect and monitor its disorders. The present method would facilitate discriminating between normal lenses and anterior lenticone suspects before the anomaly became evident. In addition, it would allow us to assess the evolution thereof with successive examinations.

Conflict of interest

No conflict of interest has been declared by the authors.

REFERENCES