

Scheimpflug imaging in anterior megalophthalmos

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We report an anterior megalophthalmos case with decreased corneal thickness and show the findings using Scheimpflug imaging. A 25-year-old male was diagnosed with anterior megalophthalmos. In both eyes, enlarged corneal length was measured. Beside a comparatively good visual acuity, a thin but clear cornea, a fairly deep anterior chamber, and central lens opacity were found. Scheimpflug images were taken using Pentacam HR. Scheimpflug-based imaging can provide us new data at the examination of this syndrome affecting the whole anterior segment.

Key words: Anterior megalophthalmos, Scheimpflug imaging, topography

In case of simple megalocornea, the enlargement of the cornea is a unique finding. Mostly, it is a bilateral, non-progressive state, with a good visual acuity and common with-the-rule

astigmatism. Corneal diameter ranges between 13.0-16.5 mm, corneal thickness is normal, cornea is clear. The anterior chamber depth is increased. The lens-iris diaphragm positioned posteriorly, the intraocular pressure is at normal range.^[1-2]

The phrase “anterior megalophthalmos” was used by Vail^[1] for the first time to describe bilateral anterior segment enlargements, which was not accompanied by increased intraocular pressure. However, in case of anterior megalophthalmos, an enlarged lens-iris diaphragm can be found next to the megalocornea, and gonioscopy reveals that the ciliary body band is wider than the trabecular meshwork and scleral spur (=enlarged ciliary ring and crystalline lens).^[2] Besides the aforementioned, central mosaic corneal dystrophy, iridodonesis, iris stromal hypoplasia, phacodonesis caused by zonular stretching, ciliary body dysplasia, ectopia lentis, cataract, posterior embryotoxon, excessive mesenchymal tissue in the angle, glaucoma (not congenital),^[2-4] vitreoretinopathy predisposing to retinal detachment are well-known findings.^[5]

In literature, it is our study that reports an anterior megalophthalmos case for the first time, which was conducted using Scheimpflug camera. The interest of the case is the decreased corneal thickness that has not been mentioned before.

Case Report

Our aim was to describe a case of a 25-years-old male diagnosed with anterior megalophthalmos in both eyes and present the usefulness of Pentacam HR in such a case.

At birth, the lengths of both corneas of the proband were found specifically enlarged. At examination under anesthesia, buphthalmus was excluded with intraocular pressure measurement and optic disc examination.

Ultrasound biomicroscopy and topography were done at the age of 15. Pentacam HR measurements were done at the age of 25 of the patient. He had a visual acuity of 15/20 and 14/20 with myopic correction at the age of 25. Anterior and posterior keratometry map was done with Pentacam HR (Oculus, Wetzlar, Germany) in both eyes. Anterior keratometry values were 40.4/41.4 D on the right eye, 40.2/41.9 D on the left eye. Posterior keratometric data were -5.5/-5.7 D on the right eye, -5.8/-5.3 D on the left eye [Fig. 1]. At corneal aberration examination, the total RMS (root mean square) was 132.71 μm

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in the right eye and 134.26 μm in the left eye, the spherical aberration value was 4.51 μm and 5.16 μm .

At slit lamp examination, an optically clear cornea was detected. The horizontal diameter of the cornea was 17.0 mm in both eyes [Figs. 2-4] at the age of 1, 9, and at the age of 25, too. Corneal pachymetry maps were obtained with Pentacam HR using the Scheimpflug imaging method, which showed a central pachymetry value of 390 μm and a corneal thickness between 450-500 μm in the periphery [Fig. 5] in both eyes. Specular microscopy (EM-1000, Tomey) measured endothelial counts of 2450 cell/ mm^2 in both eyes.

The anterior chamber was found deep measured with ultrasound, 7.24 mm in the right eye and 7.47 mm in the left eye. Ultrasound biomicroscopy (HT 35-50 UBM system, Ophthalmic Technologies Inc.) (which was done at the age of 15) showed anterior chamber depth of 7.95 mm and 7.9 mm [Fig. 6]. With Pentacam HR, 8.04 mm anterior chamber depth was measured in the right eye, and 8.31 mm in the left eye, the volume of the chamber was 721 mm^3 in the right eye and 742 mm^3 in the left eye. The angle of the chamber measured with Pentacam HR is an average of 50.7 $^\circ$ and 54.9 $^\circ$ (between 34.5 $^\circ$ és 58.7 $^\circ$ degree in

both eyes) in the right and left eye [Fig. 5].

Significant iridodonesis was observed, but phacodonesis was not detected. Central opacity was observed in the lens in both eyes, of which ultrasound biomicroscopy images were taken [Fig. 6]. The thicknesses of the lens were 3.61 and 3.43 mm in the right and left eye, measured with ultrasound biomicroscopy.

Indirect gonioscopy detected wide ciliary band, dense pigmentation was observed on the trabecular meshwork in both eyes [Fig. 7].

The axial length was 28.4 mm in the right eye and 29.06 mm in the left, the vitreous index (vitreous length/axial length x100) was 61.8% in the right eye and 62.5% in the left eye. Intraocular pressure was within normal interval in both eyes. No fundus deviation was found.

Discussion

Only few readings have been published on anterior megalophthalmos in literature. Among the image scanning methods, only ultrasound biomicroscopy examinations have

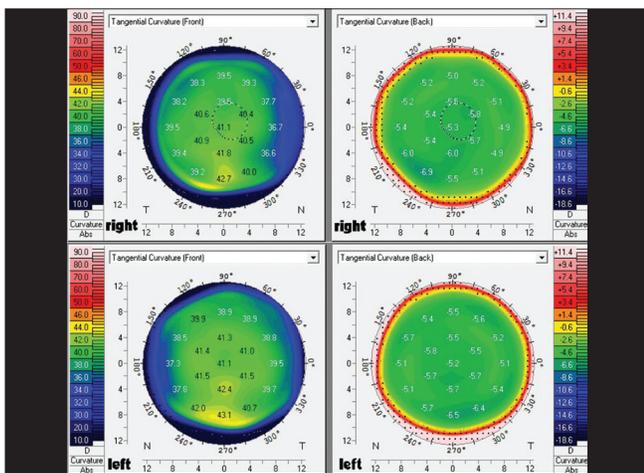


Figure 1: Anterior and posterior keratometry map obtained with Pentacam HR in both eyes. Image was taken at the age of 25

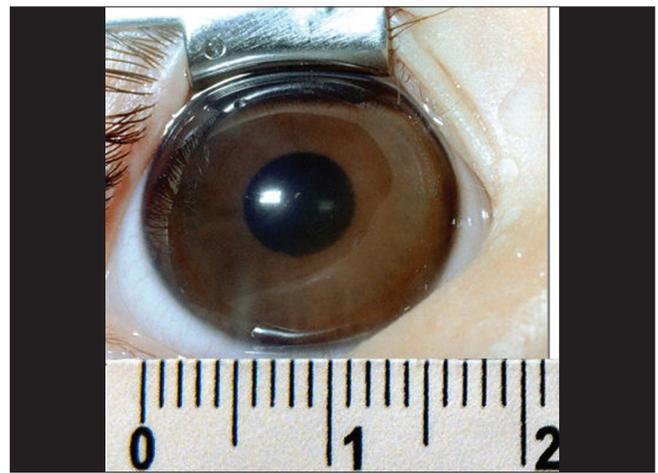


Figure 2: Anterior megalophthalmos at the age of 1. Corneal diameter is 17 mm



Figure 3: Bilateral anterior megalophthalmos. Image was taken at the age of 9. Significantly increased corneal diameter can be seen



Figure 4: Anterior megalophthalmos. Image was taken at the age of 3. Clear cornea and the significantly deep anterior chamber can be seen

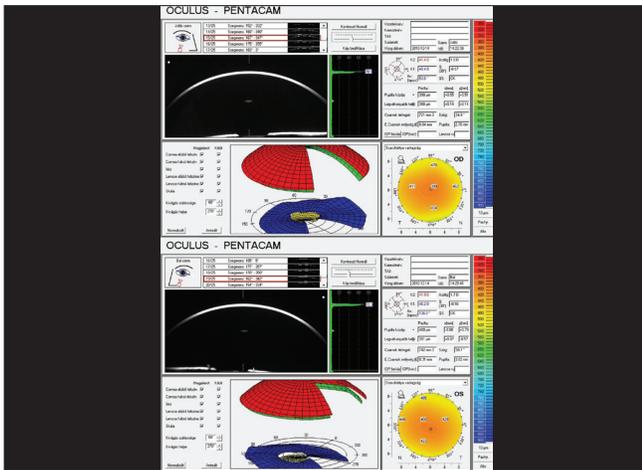


Figure 5: Scheimpflug picture of anterior megalophthalmos. Scheimpflug picture and corneal pachymetry map shows the cornea thinner than the average. The extreme depths of the anterior chamber on both eyes are clearly shown

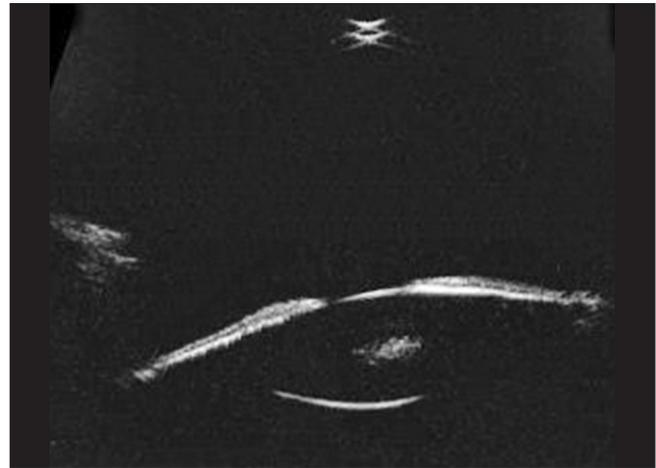


Figure 6: Ultrasound biomicroscopy picture in case of anterior megalophthalmos: The similar images taken of both eyes show the really deep anterior hall and central lens opacities

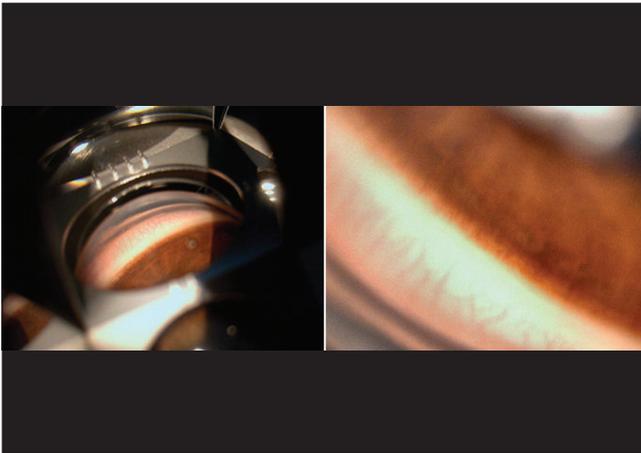


Figure 7: Wide ciliary ring can be observed on both eyes with indirect gonioscopy

been performed so far.^[3] During Scheimpflug imaging used by us, the lens of the device is tilted relative to the film plane, so the lens plane, film plane, and the plane of focus are not parallel to one another. The 3 planes intersect in a common straight line. Due to this configuration, anterior segment structures (lying in the same plane of focus) from the corneal surfaces to surface of the lens can be sharply focused. This type of imaging causes distortion of measurements in the photographs. The high-resolution Scheimpflug system used by us automatically corrects data of the anterior and posterior corneal surfaces for optical and geometric distortion, so all data presented by Pentacam HR are valid.

The anterior segment rotating Scheimpflug camera rotates along the optical axis of the eye. This device uses 475 nm monochromatic blue light for imaging, the camera captures 25, 50, or 100 scans in 2 seconds with 2760 measuring points. The software allows for automatic analysis of the anterior segment, anterior and posterior topography of the cornea, pachymetry, calculation of the chamber angle, volume, chamber height (anterior chamber depth), and analysis of the lens. Finally, the instrument creates a 3D model of the

anterior segment.

Scheimpflug analysis demonstrates clearly that the cornea is uniformly thinner than the normal and aberrometric values, such as spherical aberration are high. The depth of anterior chamber and also the angle of the chamber can be observed with Pentacam HR. We would like to demonstrate that all anterior segment parameters can be obtained with an easy, noncontact device.

In many cases, anterior megalophthalmos involves cataract, as in our case, too. Beside central lens opacities, our patient had relatively good and acceptable visual acuity, so no cataract surgery was needed. In these cases, the application of a cataract surgery and precise biometry is rather complicated.^[6-8] Beside cataract surgery, a few study reported photorefractive keratectomy^[9] performed in case of anterior megalophthalmos and penetrating keratoplasty conducted due to a developed bullous keratopathy.^[10]

In summary, anterior megalophthalmus is a rare disease affecting the whole anterior segment, in whose detailed description Scheimpflug imaging can play an important role.

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