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CASE REPORT

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Microcornea associated with myopia

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Abstract *Background:* The presentation of microcornea associated with axial myopia by means of modern examination techniques. *Methods:* Clinical investigations. *Results:* The horizontal diameter of the cornea was 8.00 mm on both sides. The average refractive power of the cornea was 39.27D/38.48D on the right/left sides by corneal topography. The central corneal thickness was 568 μm/559 μm on the right and left sides, respectively. The depth of the anterior chamber was 1.18 mm/1.14 mm and the origin of the irises was steep, as demonstrated by ultrasound biomicroscopy (UBM). The axial length was 26.42 mm/25.63 mm on the right/left sides. Endothelial morphology disclosed degeneration on both sides. *Conclusions:* The present case demonstrates that the clinical signs of microcornea are flat corneal surface, normal thickness, and degenerated endothelium. This disorder associated with axial myopia is an extremely rare ophthalmologic condition.

Keywords Microcornea · Pachymetry · Cornea topography · Axial myopia

Introduction

Microcornea is a congenital abnormality when the cornea and the anterior segment of the eye are smaller than normal. The horizontal diameter of the cornea does not reach 10 mm even in adulthood. The microcornea can be associated with a number of ophthalmologic and systemic symptoms [5]. However, in most cases the disease is associated with hypermetropia. Only few data are available to prove the coexistence of microcornea and axial myopia, and even the most recent one dates back 20 years [2, 3, 6, 8]. The purpose of the present study was to demonstrate the clinical signs and symptoms of microcornea with axial myopia by modern diagnostic methods.

Case presentation

A 64-year-old male patient underwent ophthalmologic examination because of primary angle closure glaucoma. The patient used to have good eyesight in his childhood and did not wear eyeglasses. He suffered from pulmonary emphysema. According to detailed family history, neither systemic nor ophthalmic alterations were present. In the course of eye examination his best corrected visual acuity was 20/600 on the right side (−9.0D sph-2.0 D cyl ax 80°), while on the left side it was 20/200 (−8.0D sph). On both sides, the cornea was clear and the horizontal diameter was 8.0 mm, and the vertical diameter 7.5 mm (Fig. 1a). The anterior chambers were shallow. The form and position of the pupil, as well as the structure of the iris, were normal in both eyes. Distinct nuclear opacity could be detected in both lenses. The structure of the vitreous body disclosed typical myopic fragmentation. In the course of the fundus examination severe myopic degeneration was discovered (Fig. 1b). The intraocular pressure on the right side was 18.0 mmHg and was 22.0 mmHg on the left side due to local latanoprost, timolol-pilocarpin combination. On ultrasound biomicroscopic (OTI Scan HF 35–50; Ophthalmic Technologies Inc., Toronto, Canada) examination the anterior chamber depth measured 1.18 mm on the right side, while it was 1.14 mm on the left side and the origin of

the irises was steep (Fig. 2a). The topography of the cornea (TMS-2N; Tomey, Erlangen, Germany) showed a regular surface on the right side, and an irregular surface on the left side. The average refractive power was 39.27 D on the right side, whereas on the left side it was 38.48 D (Fig. 2b).With ultrasound pachymetry (AL-1000, Tomey) the central thickness of the right cornea was 568 μm, whereas it was 559 μm on the left side. In the course of examination of the cornea with endothelial specular microscopy (EM-1000 Tomey) bigger, elongated cells with irregular structure were found on both sides. The axial length was 26.42 mm on the right side and 25.63 mm on the left side with ultrasound contact biometry (Ocuscan RxP, Alcon, Forth Worth, Tex., USA).

Discussion

Microcornea may occur sporadically, but autosome dominant and recessive forms of inheritance are also known. In general, it is associated with microphthalmos, but usually other ophthalmologic developmental anomalies are also detected; however, the cornea itself is histologically intact. Hypermetropic refractive error is very frequent, as well as the occurrence of cataract, and in 20% of cases glaucoma is present [2, 3, 5, 6, 8]. The refractive power of the cornea is generally normal or can be larger in some cases [8]. Microcornea can be associated with the very rare colobomatous macrophthalmia with microcornea syndrome; only three cases have published so far in the literature. This disease is inherited in an autosomal dominant pattern, with a complete penetrance which involves microcornea, typical iris and chorioretinal coloboma, and axial myopia. The macula is affected in most cases by uveal coloboma and myopic degeneration [1, 9].

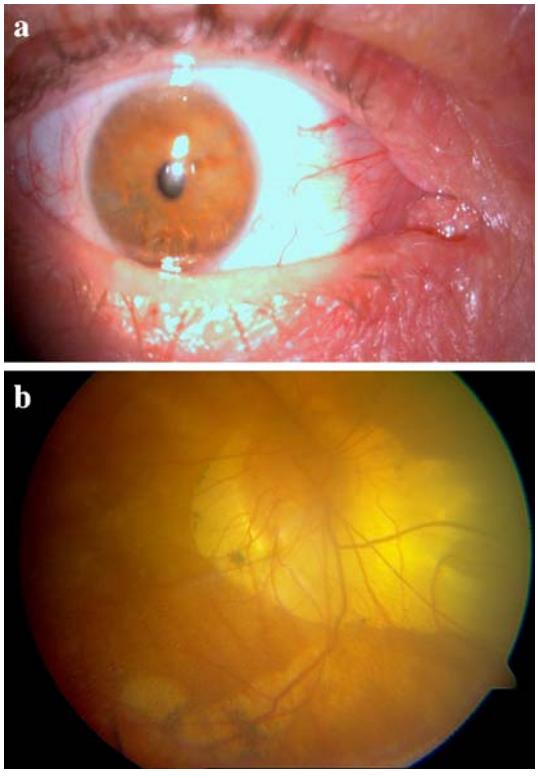

Fig. 1 Clinical appearance: note the smaller cornea with the horizontal diameter of 8.00 mm and vertical diameter of 7.5 mm (**a**). Myopic degenerations on the fundus are present (**b**)

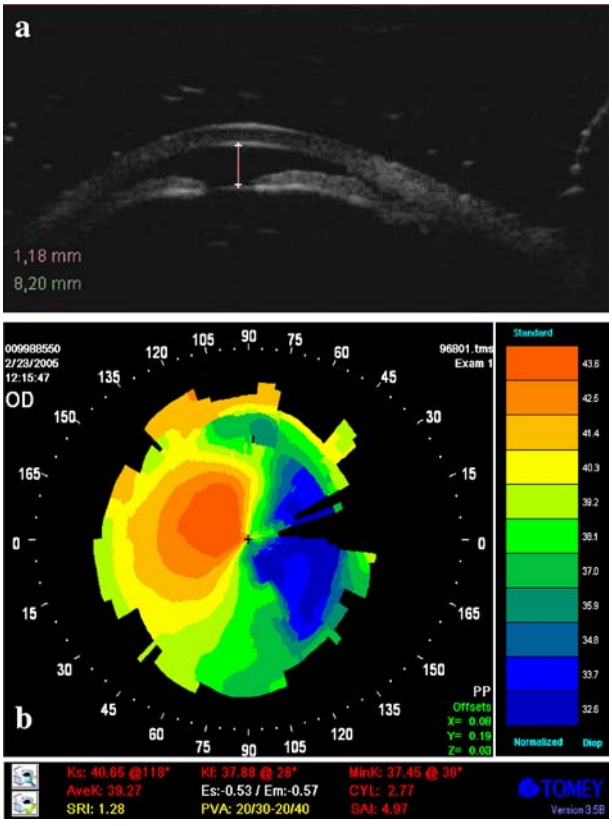

Fig. 2 Diagnostic methods: shallow anterior chamber is shown on the ultrasound biomicroscopical section of the anterior segment (**a**). Corneal topography: flat corneal surface (**b**)

In our case, a sporadic form occurred with smaller than normal corneal refractive power and myopic refraction of the eye.

In spite of the fact that the literature of the microcornea is limited, it is not entirely unified. According to the latest study dealing with the determination of the diameters of the cornea, the lower limit is 11.0 mm in the case of men and 10.7 mm in the case of women [7]. Inoue in his study makes an account of the presence of secondary Descemet's membrane in the case of microcornea [4].

This case demonstrated the clinical signs of microcornea associated with axial myopia as a rare ophthalmologic condition which is not necessarily of typical appearance. The clinical appearance of the present case differs from the colobomatous macrophthalmia with microcornea syndrome, because no colobomatous alterations and inheri-

tance pattern are present. It was found that the corneal surface was flat and the thickness of the cornea was normal. However, the endothelial layer disclosed serious degeneration, similar to bullous keratopathy, as reported previously [4]. Shallower anterior chamber and myopic refraction with myopic retinal degeneration were also present in this case. The angle closure glaucoma was successfully treated with local combined antiglaucomatous therapy.

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