

Anterior Megalophthalmos

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We describe a 36-year-old female who suffered from presenile cataract (nuclear sclerosis and posterior subcapsular opacity, with more-severe disease in the right eye than in the left) and report the use of some ophthalmologic examinations to evaluate the anatomic structures and visual functions of both eyes. Slit-lamp biomicroscopy showed increasing horizontal and vertical corneal diameters (14.0/13.5 mm) and iridodonesis. The pupils could not be fully dilated, and the lenses exhibited cataractous changes. The postlimbal depths were 1.772 mm (OD) and 1.690 mm (OS) (normal value, < 0.20 mm). The results of gonioscopy, specular microscopy, Goldmann visual field, and intraocular pressure evaluation were all within normal limits. Because the patient had bilateral megalocornea in the absence of glaucoma, a very deep anterior chamber depth, secondary effects of iridodonesis, and changes to cataractous lenses, anterior megalophthalmos was diagnosed.

This disease is rare. Secondary effects such as iridodonesis, miosis, atrophy of the iris stroma lens subluxation, and occurrence of cataractous lens should be kept in mind. (*Chang Gung Med J* 2005;28:191-5)

Key words: anterior megalophthalmos, presenile cataract, megalocornea, congenital glaucoma.

Megalocornea is a non-progressive, uniformly bilateral, congenital enlargement of the anterior segment in the absence of episodic or constant intraocular pressure elevation.⁽¹⁾ It can occur in 3 patterns: simple megalocornea unassociated with other ocular abnormalities; anterior megalophthalmos with megalocornea; and iris and angle abnormalities as well as buphthalmos in infantile glaucoma.⁽²⁾

Anterior megalophthalmos is the most-common familial form of megalocornea, which includes bilateral megalocornea, ciliary ring enlargement, and secondary effects of iridodonesis, miosis, atrophy of the iris stroma, and frequent occurrence of cataractous lenses.⁽³⁾ Clear corneas are of a normal thickness with well-defined limbi and horizontal diameters ranging from 12.5 to 18.0 mm.⁽³⁾

The pathogenesis of a simple megalocornea and

anterior megalophthalmos is unknown. There may be a large cornea (keratodysgenesis) and iris, and angle abnormalities (iridogoniodysgenesis), or a combination of these.⁽²⁾ Differentiating between anterior megalophthalmos and primary infantile glaucoma is very important, as in the latter case, surgical treatment is required to prevent blindness.

Herein we report the ophthalmologic and ocular biometry of a patient with anterior megalophthalmos.

CASE REPORT

A healthy 36-year-old woman suffered from blurred vision of the right eye. She visited our hospital for an eye examination in February 2002. After the examination, presenile cataracts (nuclear sclerosis and posterior subcapsular opacity, with more-

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severe disease in the right eye than in the left) and megalocornea (both eyes) were found. We used some ophthalmologic examinations including slit-lamp biomicroscopy, A-scan biometry (Sonomed A-1000 scanner, Sonomed Technology, gonioscopy, specular microscopy (TOMEY EM 1000, Tomey, Nagoya, Japan), keratometry (TOPCON KR-7000, Topcon Manufacturing Corp. of America, USA), Goldmann visual field, and a Perkins hand-held applanation tonometer to evaluate the anatomic structures and visual functions of both eyes. Slit lamp biomicroscopy of both eyes showed clear corneas without Descemet's membrane tear, increased horizontal and vertical corneal diameters (14.0/13.5 mm), and no iris transillumination, but iridodonesis was found (Figs. 1-2). Lenses exhibited cataractous changes. A-scan biometry revealed that the bilateral anterior chamber depths were OD 5.52 mm and OS 5.48 mm and the bilateral axial lengths were OD 26.37 mm and OS 26.48 mm. The results of gonioscopy (Fig. 3), specular microscopy, and Goldmann visual field evaluation were all within normal limits. Keratometry (KM) results were OD 39.87 x 15°/40.37 x 105° and OS 39.87 x 147°/40.37 x 57°. The KM readings of both eyes revealed flat corneas without significant corneal astigmatism. Intraocular pressures measured using the applanation tonometer were OD 13~18 and OS 15~18 mmHg. The pupil of either eye could not be fully dilated after pharmacologic mydriasis. The postlimbal depths by formulas of Meire and Delleman's study⁽⁴⁾ were 1.772 mm

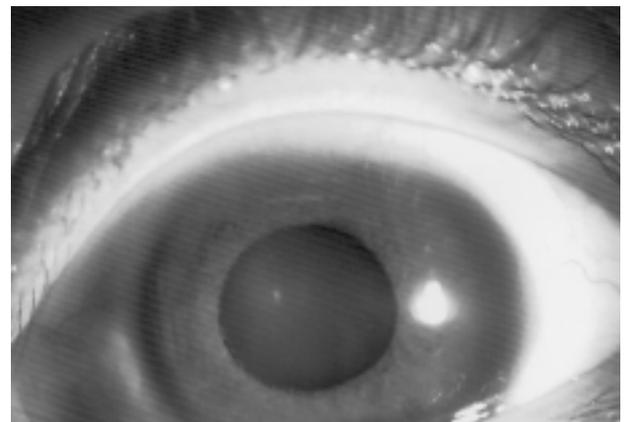
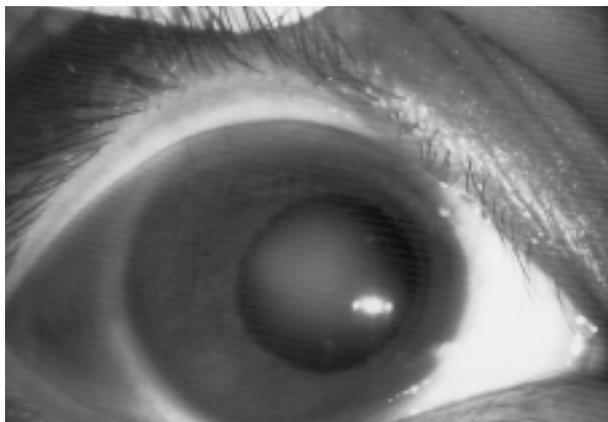
(OD) and 1.690 mm (OS) (normal value, < 0.20 mm) which indicated a posterior positioning of the iris and lens in both eyes. The corneal sizes of her son (9 years old) were within normal limits (horizontal corneal diameters of 12.0 mm (OD) and 11.5 mm (OS)), and no family history of enlarged corneas or other related ocular complications were detected. Because this female patient had bilateral megalocornea in the absence of glaucoma (Fig. 4), secondary effects of iridodonesis, cataractous lenses, a very deep anterior chamber depth, and increasing postlimbal depth, anterior megalophthalmos was diagnosed.

In order to improve her visual acuity, she received phacoemulsification and intraocular lens implantation (OD) in July 2002. During the operation, no definite zonular weakness area or phacodonesis was noted. Before surgery, the best-corrected visual acuity was 10/20 (OD). One month after surgery, the best-corrected visual acuity was 20/20 (OD).

DISCUSSION

Megalophthalmos is a term that refers to abnormally large eyes in the absence of glaucoma, with enlarged corneas and increased axial lengths.⁽⁴⁾ A series of 6 non-familial patients with developing lens luxation and retinal detachment was reported by Dufour et al. in 1971.⁽⁵⁾

A macrocornea indicates a horizontal diameter



Figs. 1 and 2 External photographs (OD and OS) revealing enlarged corneal size (with horizontal and vertical diameters of 14.0 and 13.5 mm, respectively) under a clear cornea without Descemet's membrane tear. The pupil of either eye could not be fully dilated after pharmacologic mydriasis.

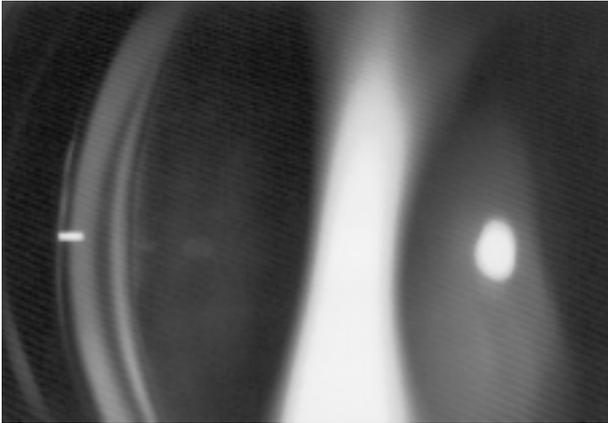


Fig. 3 Gonioscopy (OS) revealing a normal angle view.

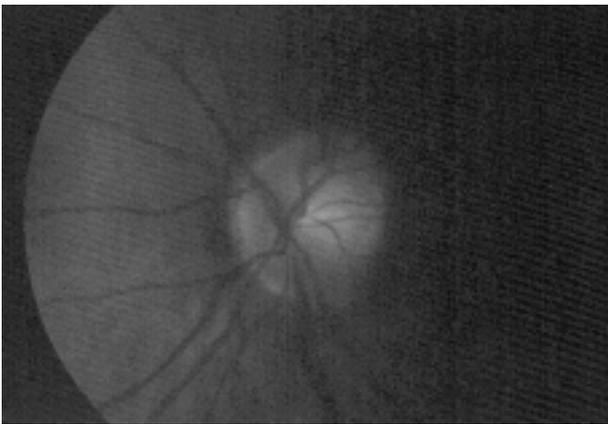


Fig. 4 Fundus photography (OS) revealing no definite glaucomatous optic neuropathy.

exceeding 12.5 mm.⁽³⁾ The major characteristics of megalocornea include: (1) a bilateral, non-progressive enlargement of a clear cornea; (2) a hereditary disorder which is almost always X-linked recessive; and (3) the absence of any evidence of glaucoma.⁽¹⁾ In comparison to megalocornea, eyes with anterior megalophthalmos have enlargement of the lens-iris diaphragm and ciliary ring in addition to the cornea.⁽³⁾

Megalophthalmos can be differentiated from congenital glaucoma by: (1) the clarity of the cornea and normal endothelial cell population densities with specular microscopy, whereas in congenital glaucoma, these densities are extensively diminished due to corneal distention;⁽⁶⁾ (2) normal intraocular pressure; and (3) a normal optic nerve. In our case, both eyes revealed: (1) clear corneas, enlarged corneal sizes, no

Descemet's membrane tears, and normal corneal endothelium densities; (2) normal intraocular pressure, iridodonesis, a high value for the postlimbal depth, and presenile cataract changes; and (3) a normal optic nerve.

Anterior megalophthalmos is a non-progressive, usually symmetric inherited condition (X-linked recessive, possibly being located on Xq21.3-q22). Although X-linked recessive inheritance is most common, all modes of inheritance (sporadic, autosomal recessive, and autosomal dominant) have been reported.^(1,7) In this case, because the family history was negative and the patient was a young healthy female, we suggest that this may have been a sporadic case.

Because of the enlargement of the ciliary ring and capsular bag, insertion of a standard posterior chamber intraocular lens is unsatisfactory because it is likely to decenter. To ensure a stable intraocular lens in the capsular bag, Dua and coauthors, in 1999, suggested suturing a 14.0-mm intraocular lens to the anterior capsule and posterior surface of the iris.⁽⁸⁾ The next year, Javadi and coauthors reported on 6 eyes of 4 patients in which a capsulorrhexis was performed; a standard intraocular lens (13.4 mm in length, with a 7.0-mm in width) can be safely implanted in the bag without complications.⁽⁹⁾ In our case, phacoemulsification and a standard intraocular lens (13.0 mm in length, with a 6.0-mm in width) implantation was performed, and no complications were noted in the following 2 months.

Anterior megalophthalmos is a rare disease. Secondary effects such as lens subluxation, iridodonesis, miosis, atrophy of the iris stroma, and occurrence of cataractous lens should be kept in mind. During cataract surgery, potential conditions such as vitreous loss, lens dislocation, etc. can influence the operative process.

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前位巨眼

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在此我們報告一位36歲女性罹患白內障,經儀器評估其雙眼結構及視覺功能後發現以下變化。首先,細隙燈下其眼角膜水平及垂直直徑增大(14.0/13.5 mm)並呈現虹膜震顫。其次,瞳孔並無法被藥物完全散大且水晶體呈白內障變化。輪狀部平面至水晶體之垂直距離為右眼1.772 mm及左眼1.690 mm(正常人為0.20 mm)。此外,隅角鏡,反射式顯微鏡,Goldmann視野計及眼壓計所測結果皆在正常範圍。由於病人呈現雙眼非青光眼性巨角膜,很深的前房以及併發虹膜震顫與白內障變化,故診斷為前位巨眼症。

此症是一罕見疾病。它所引發的病症如虹膜震顫,瞳孔縮小,虹膜基質萎縮,水晶體半脫位及最常見的白內障變化是我們須注意的。(長庚醫誌 2005;28:191-5)

關鍵字：前位巨眼症,白內障,巨角膜,先天性青光眼。

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