Sequential Bilateral Lenticulo-Corneal Touch in a Patient with Weill-Marchesani Syndrome

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Purpose: To report a case of Weill-Marchesani syndrome (WMS) with spontaneous sequential bilateral lenticulo-corneal touch, a previously unreported finding in this condition.

Case Report: A 26-year-old woman, a known case of WMS, presented with central lenticulo-corneal touch and corneal edema but normal intraocular pressure (IOP) and a patent laser iridotomy in her left eye. She underwent lensectomy, anterior vitrectomy and iris claw-fixated aphakic intraocular lens implantation. Seven days later a similar presentation developed in her right eye which underwent a similar procedure. Seven months postoperatively, best corrected visual acuity was 20/25, IOP was normal and the corneas were clear in both eyes; central corneal thickness was 555 and 573 µm in the right and left eyes, respectively. Ocular examinations were otherwise unremarkable.

Conclusions: Spontaneous lenticulo-corneal touch is a previously unreported finding in Weill-Marchesani syndrome.

Iran J Ophthalmic Res 2007; 2 (2): 161-163.

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INTRODUCTION

Ectopia lentis refers to displacement of the crystalline lens which may be congenital, developmental or acquired. Trauma is the most common cause in acquired forms while non-traumatic ectopia lentis is most commonly associated with Marfan syndrome, homocystinuria, aniridia, congenital glaucoma and less frequently with Ehlers-Danlos syndrome, hyperlysinemia, sulfite oxidase deficiency and Weill-Marchesani syndrome (WMS).[1]

WMS is a rare hereditary connective tissue disorder characterized by short stature, brachydactyly, joint stiffness, a broad skull and characteristic eye abnormalities including shallow orbits, microspherophakia, high myopia, ectopia lentis and glaucoma.[2] Despite the clinical homogeneity, both autosomal dominant and recessive inheritance patterns have been reported.[3,4] The characteristically small and spherical lens and the zonular laxity predispose the eye to pupillary block and angle-closure glaucoma. Therefore, prophylactic laser iridotomy is prudent in this setting. Mean age of lens subluxation in patients with WMS is 18 years and the displacement is usually inferior.[5] Herein we report a case of WMS with sequential bilateral lens dislocation into the anterior chamber with central corneal touch but normal intraocular pressure (IOP) and patent iridotomies.

CASE REPORT

A 26-year-old woman complained of photophobia and decreased vision in her left eye of two days' duration. She had short stature (143 cm height), brachydactyly, joint stiffness and a broad skull in favor of WMS. Ophthalmologic examination revealed best corrected visual acuity (BCVA) of 20/40 with -13.75 -4.50@155˚ in her right eye and 20/800 with -14.00 -4.50@10˚ in the left. In the right eye the cornea was clear and the anterior chamber was shallow. In the left eye the cornea was edematous at the site of lenticulo-corneal touch, the anterior chamber was nearly flat and the lens was microspherophakic (Fig. 1). Laser iridotomies were patent in both eyes. IOP was 14 mmHg and 10 mmHg in the right and left eyes respectively. Fundoscopic examination revealed normal optic discs with 0.2 cup/disk ratio and pink neuroretinal rims. The lenticulo-corneal touch did not improve with cycloplegia and supine position; therefore the patient underwent lensectomy, anterior vitrectomy and iris claw-fixated aphakic intraocular lens (Artisan, Ophtec BV, Groningen, the Netherlands) implantation in the left eye. Seven days later, a similar presentation developed in her right eye which underwent the same procedure.

Seven months after surgery, BCVA was 20/25 in both eyes and IOP was 14 mmHg in the right eye and 12 mmHg in the left. Both corneas were clear with central thickness of 555 µm in the right eye and 573 µm in the left. Funduscopy revealed an attached retina, normal appearing discs and posterior poles in both eyes without any retinal breaks.

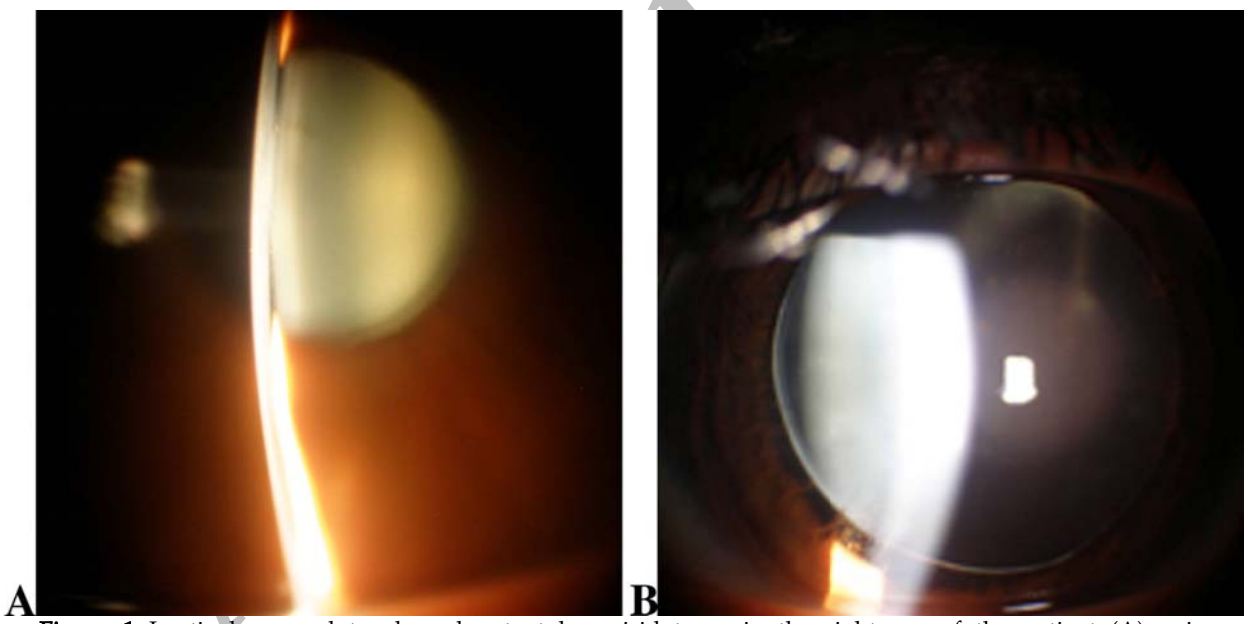

Figure 1 Lenticulo-corneal touch and patent laser iridotomy in the right eye of the patient (A); microspherophakia, anterior lens displacement and central corneal edema (B).

DISCUSSION

The normal crystalline lens is almost spherical at birth with an equatorial diameter of 6.5 mm and anteroposterior diameter of 3.5 to 4.0 mm. The equatorial diameter increases to 9.0 mm at age 15 years and maintains this size thereafter.[6] In microspherophakia, the lens is small and spherical; the exact cause is unknown but alterations in secondary lens fibers have been suggested. Another theory attributes microspherophakia to the lack of tension in rudimentary zonular fibers inhibiting normal lens development, leading to the spherical shape.[1]

In patients with WMS, the zonules are characteristically longer and looser than normal. This can contribute to microspherophakia and induce pupillary block.[3]

In WMS, the crystalline lens is usually dislocated inferiorly; anterior displacement and flat anterior chamber with a patent iridotomy and normal IOP has not yet been reported. Harasymowycz et al[7] reported a case of WMS developing flat anterior chamber after trabeculectomy which did not resolve after anterior chamber reformation and tight closure of the trabeculectomy flap. The exact mechanism of this phenomenon is not determined but may be due to the characteristic anatomical abnormalities including narrow angles, microspherophakia, zonular laxity and ciliary body hypoplasia.[8] Lens dislocation into the anterior chamber must be added to the current indications for lensectomy in this syndrome which include, progressive subluxation with pupil bisection, monocular diplopia and visual acuity less than 20/70 for distance and J5 for near.[5]

Surgical complications due to difficult capsulorrhexis and an excessively mobile lens are not uncommon. However, the outcomes of lensectomy in WMS have been improved with recent advances in microsurgical techniques. The absence of adequate capsular support may mandate implantation of an anterior chamber intraocular lens (IOL), iris-fixated IOL, posterior chamber iris- and scleral-sutured lenses or simply leaving the eye aphakic.[9] Behndig[10] described the successful use of capsular tension ring (CTR) and lens implantation in a case of WMS. However one should consider the risk of complete dislocation of the capsular bag-CTR-IOL complex into the vitreous cavity in eyes with extremely weak zonules. Harasymowycz et al[7] reported the successful implantation of trans-scleral fixated IOLs in both eyes of two brothers with WMS. Considering the reported complications of scleral fixated IOLS[11] we used iris claw-fixated aphakic IOLs which seemed safer than other types of IOLs.[9,12,13]

In conclusion, spontaneous lenticulo-corneal touch and flat anterior chamber in patients with WMS without signs of pupillary block seems to be a newly reported finding in this rare syndrome; lensectomy and iris claw-fixated IOL implantation may be considered in this setting.

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