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Narrow Angles in High Myopia

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CASE PRESENTATION

A 27-year-old white woman with high myopia (spectacle correction of -8.50 D OU) was referred for an evaluation of narrow angles. She originally presented to her local ophthalmologist with complaints of blurred vision of gradual onset; she had last been checked for glasses 2 years earlier. She reported a single episode when she awoke during the night and found her vision to be "glazed over" with no associated pain or nausea. When she awoke the following morning, her vision was fine, and she experienced no further episodes. She also reported a history of head trauma from a fall while in the military, but she denied direct trauma to her eyes. The patient's past medical history was only significant for nephrolithiasis and sinus disease, and she was using no medications. Her family history was unremarkable. Her highest recorded IOPs were 21 mm Hg OD and 20 mm Hg OS.

On examination in our clinic, the patient's visual acuity was 20/40 OD (20/25 with pinhole) and 20/30 OS. Her IOPs measured 18 mm Hg OD and 16 mm Hg OS, with a central corneal thickness of 586 µm OD and 579 µm OS. The slit-lamp examination was notable for bilaterally shallow anterior chambers (just greater than two corneal thicknesses centrally) and phacodonesis in both eyes (Figure 1); otherwise, it was normal. Gonioscopy showed appositional closure throughout most of the angle in both eyes. Compression opened the angles to the scleral spur and created peripheral iris concavity with a central elevation overlying the lens. The undilated fundus examination revealed healthy discs with cup-to-disc ratios of 0.4 bilaterally and normal-appearing retinas.

The patient was of mildly short stature with normal and proportionate limbs and extremities.

Humphrey visual field testing (using a standard 24-2 Swedish interactive threshold algorithm; Carl Zeiss Meditec, Inc., Dublin, CA) was normal, as were optic disc and retinal nerve fiber layer imaging (by confocal scanning laser ophthalmoscopy and spectral domain optical coherence tomography [OCT], respectively). These tests confirmed the absence of glaucomatous damage.

Axial length, as measured by partial coherence interferometry, was 23.7 mm OD and 23.3 mm OS. Based on the slit-lamp examination and imaging with anterior segment OCT, we estimated the anterior chamber to be approximately 1.5 mm deep centrally in both eyes.

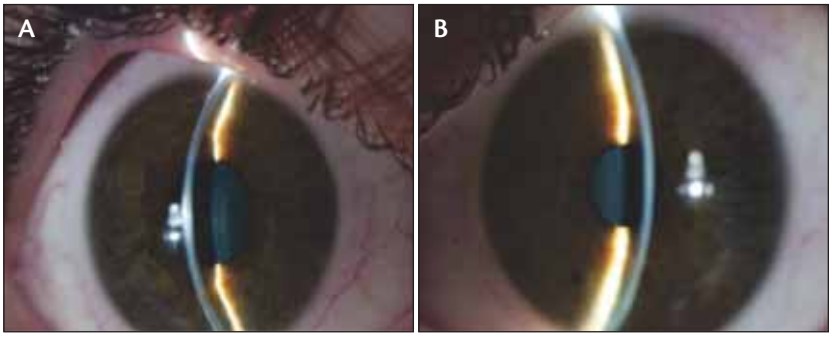

Figure 1. The slit-lamp examination of the patient's right (A) and left (B) eyes demonstrates shallow anterior chambers.

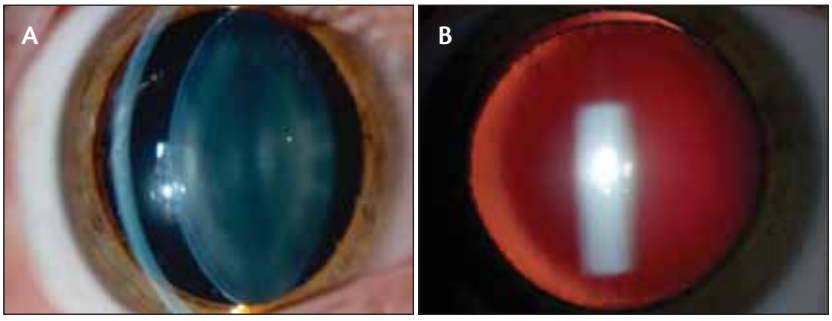

Figure 2. The slit-lamp examination after pupillary dilation shows shallow anterior chambers with increased sphericity of the crystalline lens (A) and visibility of the lens equator (B).

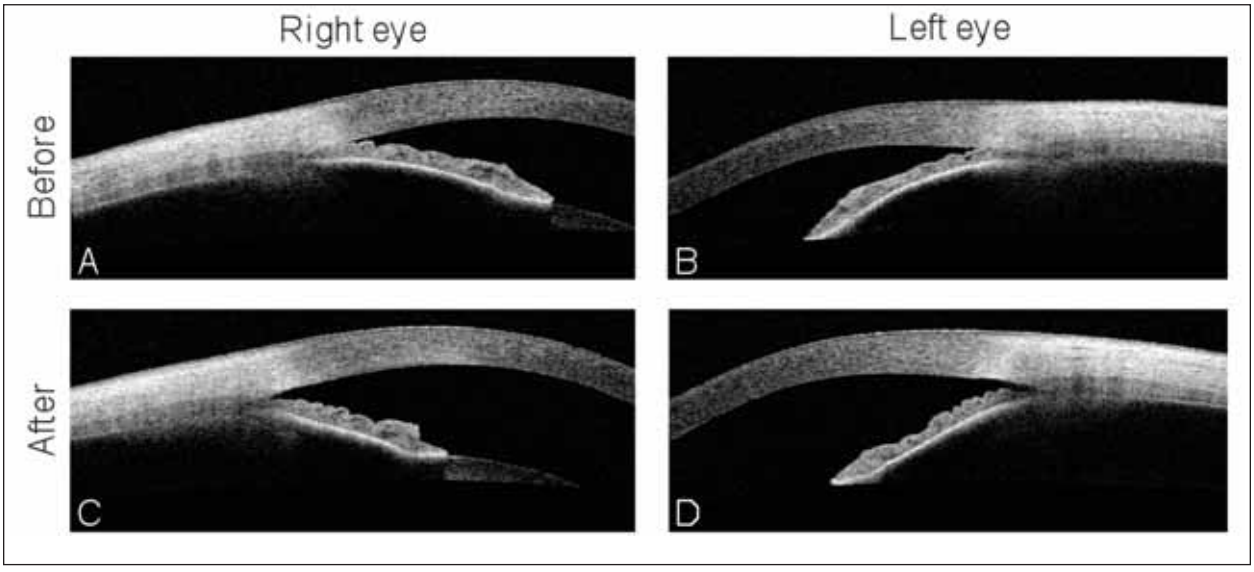

Figure 3. Anterior segment OCT of the right and left eyes shows appositional closure of the angles (A and B), with opening of the angles to the scleral spur after laser peripheral iridotomies (LPIs; C and D).

Eventual pupillary dilation revealed thick, rounded crystalline lenses (in the anteroposterior dimension), their equators visible within the pupillary margins (Figure 2). Scheimpflug imaging supported our clinical impression of the lenses' dimensions, each with a measured thickness of nearly 5 mm and a diameter of nearly 8 mm. No choroidal effusion was visible on ultrasound biomicroscopy.

DISCUSSION

This case demonstrates the uncommon presentation of narrow angles in a highly myopic young patient. Narrow angles are typically present in older patients, particularly hyperopes who have short axial lengths, where a growing crystalline lens or cataract begins to crowd the anterior chamber and promote relative pupillary block. The age and refractive error of this patient should immediately raise the possibility of an atypical cause for angle narrowing.

Reported cases in the literature of angle closure in the setting of high myopia (excluding those due to inflammation or contracting membranes) include a number of possible etiologies such as primary pupillary block, plateau iris, malignant glaucoma after scleral buckling, and syndromic lenticular abnormalities.[1] In young patients, primary pupillary block is much less frequent. More common are secondary causes of narrow angles, including plateau iris, iris cysts, retinopathy of prematurity, and lenticular abnormalities.[2] Rarely, angle closure with a large myopic shift can be induced by medications such as miotics or topiramate.[3]

In this case, the history and examination findings point to spherophakia as the cause of angle narrowing. Spherophakia is a rare, bilateral abnormality of the lens due to abnormally weak zonules. They limit the radial growth of the developing lens, leading to the formation of a relatively spherical lens and resultant high lenticular myopia. With pharmacologic pupillary dilation, the lens equator and zonules may be seen to varying degrees. A significant reduction in the equatorial lens diameter evokes the term *microspherophakia*, but the anteroposterior thickness of the lens is greater than normal. Because the zonules are weak, the lens may be loose, which manifests as irido- or phacodonesis, and it may shift anteriorly, which further induces myopia and shallows the anterior chamber.

Angle closure in spherophakia usually results from pupillary block induced by the anteriorly displaced lens and its increased anterior curvature.[3] This mechanism can be exacerbated by miotics and alleviated by pupillary dilation and cycloplegia, giving rise to the term *inverse angle closure*. Disruption of the lens zonules may also cause subluxation or complete dislocation of the lens into the anterior chamber. As with any case of pupillary-block angle closure, LPI is indicated and likely will be curative. Any "phacomorphic" component from an anterior lens, however, may still pose a risk for chronic angle closure.

Spherophakia and lens subluxation are classically seen in Weill-Marchesani syndrome (associated with short stature, brachydactyly, brachycephaly, and stiff joints),[3] Marfan syndrome (associated with tall stature, arach-

nodactyly, joint hyperextensibility, chest-wall deformities, cardiac defects, and megalocornea),[4] and homocystinuria (associated with Marfanoid habitus and mental retardation).[5] Although this patient was of mildly short stature, she had normal-appearing fingers and no known systemic abnormality. Thus, hers likely represents one of few reported cases of isolated spherophakia.[6-10] Recent efforts have identified some of the genetic mutations associated with spherophakia or lens subluxation in both syndromic and isolated forms.[11-13] Genetic testing should be offered to any patient with these findings.

If angle closure persists after LPI or there is advanced lens subluxation, extraction of the crystalline lens may be necessary. Just as in cases of trauma or pseudoexfoliation, surgery carries a high risk of complications, including zonular dehiscence and vitreous loss. Numerous options for lensectomy exist, including pars plana lensectomy or intracapsular cataract extraction with the placement of an anterior chamber or posterior chamber IOL. Another option is extracapsular extraction by phacoemulsification with possible placement of the IOL in the bag, provided sufficient zonular support exists and the surgeon recognizes the risk of late dislocation of the IOL.[8-10] The optimal approach would depend on the individual case but is beyond the scope of this discussion. In cases where glaucoma persists despite lensectomy, appropriate medical and surgical IOP-lowering therapy is necessary.

CLINICAL COURSE

This patient was diagnosed with high lenticular myopia due to isolated spherophakia and secondary appositional angle closure without ocular hypertension or detectable glaucoma. She was successfully treated with LPIs to completely open her angles (Figure 3). She declined a genetic evaluation. Months later, her IOP remains normal, and she continues to be asymptomatic with refractive correction. We advised her of the importance of continued follow-up and cautioned her about the possibility of future problems that might arise from further subluxation of her lenses. ❑

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