Acute Angle Closure in the Fellow Eye as a Complication of Prone Positioning After Vitreoretinal Surgery

Facedown positioning is a routine procedure after some vitreoretinal surgical procedures. We report a case of acute angle closure in the fellow eye during the postoperative phase of this procedure.

Report of a Case. A 56-year-old woman had a 2-month history of decreased vision and mild discomfort in the left eye. She had a history of penetrating injury at the age of 8 years that led to traumatic cataract and corneal scarring in her left eye. Two years before we saw her, she had undergone uncomplicated combined cataract extraction and lens implantation with penetrating keratoplasty elsewhere.

There had been a satisfactory convalescence, but the visual acuity OS remained counting fingers at 1 m. Amblyopia was therefore suspected.

At our first examination of her, the visual acuity was 6/9 OD and hand motion in the left eye. Full ophthalmological workup revealed a graft rejection and a subtotal retinal detachment with a peripheral retinal tear at the 6-o’clock position in the left eye. Intraocular pressure was 22 mm Hg OD and 18 mm Hg OS. In the right eye there was early nuclear cataract formation, but otherwise all findings were normal. No signs of pseudoexfoliation were noted and the cup-disc ratio was 0.3. The patient was admitted and the graft rejection was successfully treated with intensive topical steroids.

After the corneal graft inflammation settled, the patient underwent a left pars plana vitrectomy with endolaser and silicone oil filling under retrobulbar anesthesia. No systemic atropine was used. Postoperatively the patient was positioned facedown. The retina was flat under silicone oil on day 1 and the pressure was 40 mm Hg in the eye that had undergone surgery. Systemic treatment with oral dorzolamide hydrochloride reduced the pressure to 27 mm Hg. On day 3 the patient complained about severe pain in the right (fellow) eye. The visual acuity had dropped to 6/60 OD. On examination, acute angle closure with very shallow anterior chamber and angle grade 0 (Schaffer) was found. The intraocular pressure was 45 mm Hg despite continued treatment with dorzolamide hydrochloride, 250 mg 4 times a day, since day 1 after surgery and additional intravenous 20% mannitol (300 mL over 1 hour).

Right eye YAG laser iridotomies were performed and the intraocular pressure dropped as the anterior chamber deepened. Pressures remained within normal limits after withdrawal of all systemic and topical treatment. The visual acuity returned to 6/12 OD.

Gonioscopy 2 months later showed a grade 2 (Schaffer) anterior chamber angle in the right eye with small patent laser iridotomies at the 11- and 2-o’clock positions. The axial length was measured at 22.79 mm. A reliable A-scan was not possible in the left eye because of the silicone oil filling.

Comment. The prone position test for narrow-angle or angle-closure glaucoma was introduced in 1968 and was found to be more sensitive and safer than the darkroom test or pharmacological provocation tests. Angle closure is thought to evolve secondary to pupillary block as the lens moves anteriorly in prolonged prone position. Facedown positioning after vitreoretinal surgery, especially after repair of full-thickness macular holes, is effectively an extended prone position provocation test.

Interestingly, the acute angle closure occurred in the fellow eye and not in the eye that had undergone surgery, probably because the eye that was operated on was pseudophakic, leaving the anterior segment less crowded. Another factor is probably the silicone oil that is lighter than water and in a prone position will move anteriorly in prolonged prone positioning. It is therefore recommended that assessment of the risk for angle closure in both eyes be included in the preoperative evaluation, especially in at-risk patients (such as those patients of southeast Asian, Chinese, or Inuit origin; or those patients with a positive family history, hyperopia, short axial length, or microphthalmos).

Where occultable angles are found during gonioscopy, a prone position provocation test preoperatively may be useful. In high-risk cases, prophylactic laser iridotomy may be considered or vitreoretinal surgery without postoperative prone positioning could be attempted.

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Long-term Fundus Changes Due to Fundus Albipunctatus Associated With Mutations in the RDHS Gene

Fundus albipunctatus (FA) is a type of congenital stationary night blindness with an autosomal recessive inheritance pattern. The fundus of affected patients has a characteristic appearance of numerous small yellow-white dotlike lesions at the level of the retinal pigment epithelium.
Marmor reported that the dotlike lesions increase in number over the years. Because his study had been carried out before mutations in the gene encoding retinal dehydrogenase type 5 (RDH5) were identified in patients with FA, the molecular genetic abnormalities of his patients had not been studied. In the present study, we identified a novel compound heterozygous mutation in the RDH5 gene in a patient with FA and investigated medical records to identify changes in fundus findings during a long-term follow-up period.

**Report of a Case.** A 21-year-old woman was first seen at age 6 years, when she was aware of night blindness. Her corrected visual acuity was 20/15 OU, and her visual fields were full. Numerous yellow-white dotlike lesions were observed at the level of the RPE in her fundus bilaterally. Since then, she had undergone periodic examinations for 15 years. Although the basic pattern of dotlike lesions did not change, there was subtle variation in their distribution and particularly in their density. For example, in the inferonasal area, the numbers of dotlike lesions appeared to have increased (Figure 1). Conversely, in the nasal area, they appeared to have decreased. No abnormality had been noted in the macular area during the 15 years of observation. Results of molecular genetic analysis revealed novel compound heterozygous mutations with nucleotide 928C to GAAG (Leu310 to GluVal) and Arg167His (CAC to CGC) in the RDH5 gene (Figure 2). For the molecular genetic study, proceedings followed the tenets of the Declaration of Helsinki and were approved by the Ethical Committee for Medical Research of Hirosaki University School of Medicine, Hirosaki, Japan.

**Comment.** One of the 2 mutations that this patient harbors is a 4–base pair (GAAG) replacement of a single nucleotide (C), such that the leucine at amino acid 310 is replaced by glutamate and valine. A homozygous pattern of this mutation has been known to be the most common cause of FA in Japan. Therefore, it seems likely that this novel combination of the mutations as a compound heterozygous pattern is also a primary cause of FA.

Although the origin of the yellow-white dotlike lesions is still uncertain, it has been speculated that abnormal materials were produced as the result of the deficiency of RDH5. Although the number and density of lesions are known to gradually increase with age, no one has previously reported on the decreasing number of lesions during a long time period, as was shown in our patient. These findings may suggest important clues for understanding the chemical properties, formation and degradation, and distribution of yellow-white dotlike lesions in FA. It is also interesting that dotlike lesions are not usually seen in the macular area, where
particularly high lysosomal enzyme activity has been reported.5

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Discordant Retinitis Pigmentosa in Monozygotic Twins

Retinitis pigmentosa (RP) is a diverse group of retinal dystrophies characterized by night blindness, progressive constriction of the visual field, and bone spicule deposition in the peripheral retina.1 All forms of inheritance have been described, including X-linked, autosomal dominant, autosomal recessive, and mitochondrial, but approximately 42% of cases are simplex (sporadic) with no known family history.1 An unusual case of monozygotic twin sisters discordant for simplex RP is reported here.

Report of a Case. Twin A was first seen by us at age 22 years with bilateral vitreous cells and cystoid macular edema (CME). Visual acuity was 20/70 OD and 20/40 OS. She had a moderate initial response to periorcular and oral steroids and to oral acetazolamide. Findings from an extensive uveitis workup were unrevealing, and she was otherwise healthy with no evidence of malignancy or systemic autoimmune disorder. She subsequently developed continued visual loss, nyctalopia, visual field constriction, arteriolar narrowing, and pigmented retinopathy with peripheral bone spicules consistent with a diagnosis of RP (Figure 1). Results of electroretinographic (ERG) studies were very