CASE REPORT

Macular Corneal Dystrophy Type 2 and Cataract in Siblings With Systemic Hypertension

Macular corneal dystrophy is an autosomal recessive disorder characterized by irregular macular opacities in diffuse cloudiness of the stroma. The abnormal deposits in the cornea reportedly stain blue with Alcian blue and colloidal iron. The disorder is divided into 2 types, according to keratan sulfate metabolism; type 1 is accompanied by lack of keratan sulfate in the serum and cornea, and type 2 has normal levels. Thonar and associates described 16 patients with macular corneal dystrophy, ranging in age from 22 to 67 years, who had no cataract.

To our knowledge, cataract in patients with systemic hypertension and macular corneal dystrophy has rarely been reported. We recently examined a man and his sister with systemic hypertension who had bilateral cataract and corneal opacities.

Case Reports

The pedigree for our patients shows a consanguineous relationship between the patients’ parents (Fig 1).

Case 1. A 49-year-old man was first seen in our clinic in 1988 complaining of blurred vision in both eyes. At age 27 years, the patient had undergone bilateral intracapsular cataract extraction at another hospital. Preoperatively, the patient’s visual acuity had been 0.1 in both eyes. Three months after the cataract extraction, he obtained good visual acuity (0.5 in the right eye).
eye and 0.3 in the left). The patient had been treated for systemic hypertension for 10 years.

On ophthalmic examination, his visual acuity was 0.3 in the right eye and 0.02 in the left eye. His intraocular pressure was 20 mm Hg in both eyes. Irregular macular opacities in diffuse cloudiness of the stroma, without neovascularization, were found in both eyes (Fig 2). The lens could not be seen in either eye. Both fundi appeared normal.

In 1992, his corneas exhibited increased cloudiness, and his visual acuity decreased to 0.02 bilaterally. At age 54 years, the patient underwent penetrating keratoplasty in the right eye in January 1992 and in the left eye in February 1992. Histopathologic findings of the excised cornea showed the presence of abnormal materials that stained blue with Alcian blue (Fig 3) and colloidal iron (Fig 4).

The postoperative course was uneventful. Clear grafts were seen in both eyes. His visual acuity recovered to 0.8, right eye, and 0.5, left eye. No recurrence of corneal opacity was noted during a follow-up period of 4 years. The keratan sulfate level in the serum, determined by enzyme-linked-immunosorbent assay (ELISA), was 309.3 ng/mL (normal, 120 to 500 ng/mL).

**Case 2.** A 49-year-old woman, the sister of patient 1, complained of blurred vision in both eyes when she visited our clinic in 1992. At age 27 years, the patient had been diagnosed with corneal opacity and cataract in both eyes at another eye clinic. She had been treated for systemic hypertension for 20 years.

On ophthalmic examination, her visual acuity was 0.1 in both eyes. The intraocular pressure was 15 mm Hg in both eyes. Corneal clouding, without neovascularization, and cataract were visible in both eyes (Fig 5). The fundus appeared ophthalmoscopically normal. Normal responses were found on bright-flash electroretinogram.

The patient underwent penetrating keratoplasty, extracapsular cataract extraction, and intraocular lens implantation in the right eye in June 1993 and in the