Medical therapy for bilateral uveal effusion syndrome in nanophthalmos

Uveal effusion syndrome (UES) is a rare disease of idiopathic exudative detachment of the choroid, ciliary body, and retina, characterized by the accumulation of fluid escaping from the choriocapillaris into nearby free spaces.[1] Brockhurst[2] described this disorder as related to nanophthalmos and scleral abnormality and indicated that it might be caused by the congestion of the choroidal venous system, resulting from the compression of the vortex veins by the thick sclera.

Surgical management has been shown to be successful for the treatment of UES. Various surgical techniques, such as vortex vein decompression[3] and partial- or full-thickness sclerectomy,[4] have been established to relieve choroidal venous congestion and decrease resistance to transscleral outflow. Uyama et al.[5] divided UES into 3 subgroups based on the axial length and scleral abnormalities and reported that surgical management was effective only for subtypes with abnormal sclera.

In this report, we describe a case of bilateral UES in a patient with nanophthalmic eyes with abnormal sclera who was successfully treated with nonsurgical management.

A 64-year-old male visited our clinic with a 2-month history of decreased visual acuity in both eyes. The patient presented with a medical history of diabetes mellitus and sigmoid colon cancer with liver metastasis. He stated that his vision had been poor since childhood because of high-grade hypermetropia.

His best-corrected visual acuity was 20/500 OD and 20/100 OS; intraocular pressure was 9 mmHg in both eyes. Slit-lamp examination showed a shallow but quiet anterior chamber and moderate nuclear sclerotic cataract in both eyes. Both eyes also showed inflammation in the anterior vitreous, which was scored as grade 2+ according to the Standardization of Uveitis Nomenclature Working Group guidelines.[6] Fundus examination revealed serous retinal detachment (SRD) in the inferonasal and temporal quadrants of the right eye and subretinal fluid (SRF) in the macular region of the left eye. Multiple pigmentary changes throughout the fundus periphery were also noted (Fig. 1A and 1B). Optical coherence tomography (OCT) revealed severe macular edema with retinal distortion in the right eye

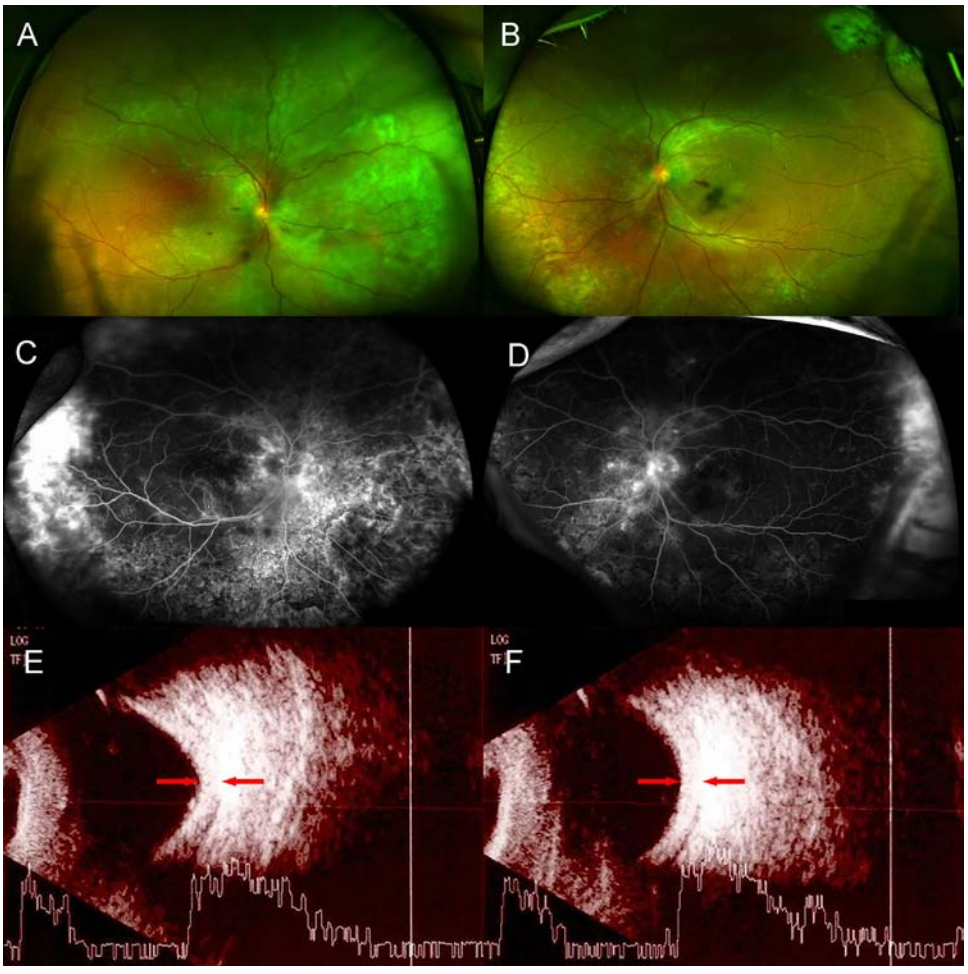

Fig. 1—Ultrawide-field fundus photography, ultrawide-field fluorescein angiography, and B-scan ultrasonography at initial presentation. (A, B) Fundus photography showing serous retinal detachment with pigmentary changes. (C, D) Fluorescein angiography reveals multiple punctate leakage in both eyes and pooling of dye in the areas of serous retinal detachment in the right eye. (E, F) B-scan ultrasonography shows the thickened sclera in both eyes.

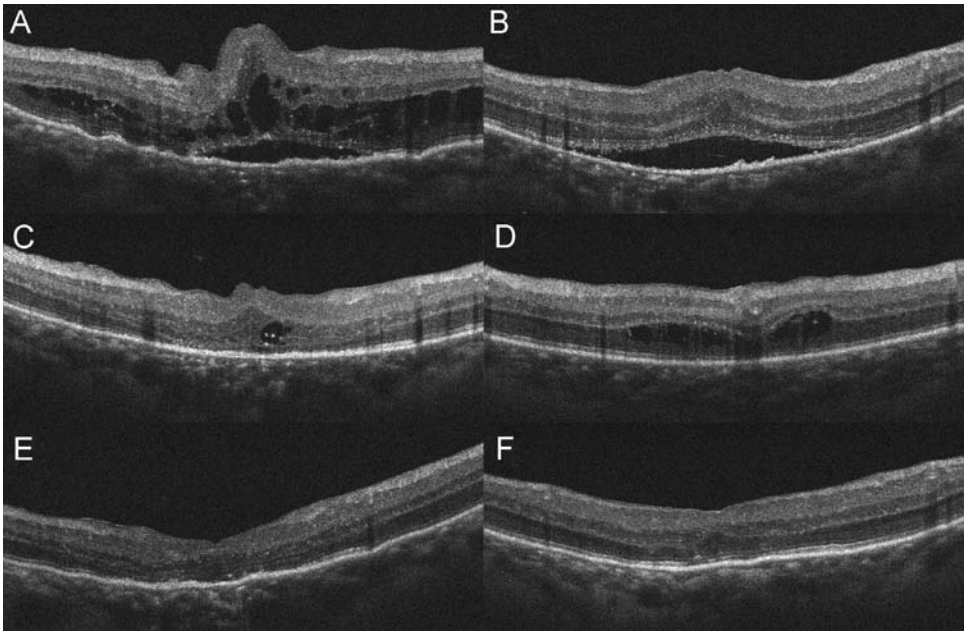

Fig. 2—Serial optical coherence tomography images. Optical coherence tomography shows severe macular edema with subretinal fluid in the right eye (A) and in the left eye (B) at the first visit. (C, D) At 2 weeks after medical therapy, the macular edema has decreased and subretinal fluid has been absorbed. (E, F) After 2 months of additional treatment, further improvement of macular edema is noted. Absence of the foveal pit and disruption of the photoreceptor layer are also noted.

and shallow SRF in both eyes (Fig. 2A and 2B). Wide-field fluorescein angiography revealed multiple punctate areas of leakage in both eyes and pooling of dye in the areas of SRD in the right eye (Fig. 1C and 1D). The spherical equivalents were +17.38 D in the right eye and +15.50 D in the left eye. Axial lengths measured by A-scan ultrasonography were 18.53 mm in the right eye and 18.37 mm in the left eye. B-scan ultrasonography demonstrated a grossly thickened sclera in both eyes (Fig. 1E and 1F).

The patient refused surgical treatment because he already had terminal cancer. Hence, we decided to manage him conservatively. He received latanoprost (0.005% twice a day) and oral acetazolamide (250 mg twice a day). After 2 weeks, OCT revealed decreased macular edema in the right eye and resolution of SRF accumulation in both eyes (Fig. 2C and 2D). However, he reported nausea, vomiting, weakness, and dizziness; therefore, we discontinued oral acetazolamide. Additional treatment with topical bromfenac (0.1% twice a day) was initiated. Two months later, OCT revealed further improvement of macular edema in both eyes (Fig. 2E and 2F). The SRD resolved completely, and retinal pigment with typical "leopard spot" changes remained in both eyes (Fig. 3A and 3B). At the last visit, 6 months after the medical therapy, the patient had no recurrence of the effusion. His vision, despite clinical resolution of the disease, had not improved, probably because of diffuse photoreceptor disruption and absent foveal pits, as seen on OCT, and clinically significant cataract.

UES is an extremely rare disease of unknown etiology characterized by uveal effusion and SRD.[4] Histologically abnormal sclera of nanophthalmic eyes is thought to have contributed to the pathogenesis of UES. Elagouz et al.[4] reported that abnormalities of the sclera, including reduced scleral protein permeability, reduced scleral

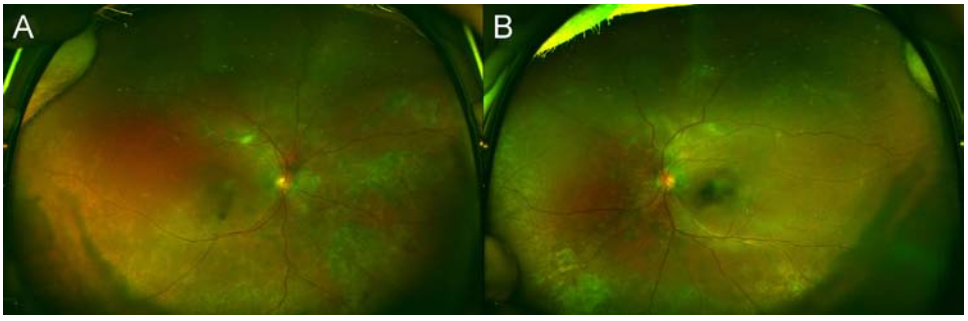

Fig. 3—Ultrawide-field fundus photography at 2 months after initial visit. (A, B) Fundus photography shows the resolution of serous retinal detachment with multiple pigmentary changes throughout the fundus periphery.

hydraulic conductivity, vortex vein compression, increased choroidal vessel permeability, chronic choroidal inflammation, and chronic hypotony, could cause UES.

The standard treatment for UES has been surgery. Surgery is aimed at resolving choroidal venous congestion by direct or indirect vortex vein decompression[1,3,7] and relieving scleral rigidity by relaxing scleral tension.[5,8] However, the isolation of the vortex vein itself is very difficult, and decompression is technically complicated to perform.[1] Significant complications such as vein rupture and considerable bleeding are also unavoidable. Sclerectomy and sclerotomy[4,8] procedures remain invasive and technically demanding as well. The response to surgery is also variable. Our report makes it apparent that SRF absorption in UES, despite scleral abnormality, could be obtained with medical therapy alone.

The exact mechanism underlying the observed resolution of SRF accumulation remains unclear, but several speculations can be raised. Weinreb[9] demonstrated that topical prostaglandin administration could reduce scleral collagen levels by increasing scleral metalloproteinase levels. Therefore, it could modulate transscleral fluid movement and enhance scleral macromolecular permeability. Furthermore, acetazolamide, a carbonic anhydrase inhibitor, seems to decrease macular edema by stimulating the pump mechanism of the retinal pigment epithelium.[10,11] The effects of nonsteroidal anti-inflammatory drugs could be attributed to their anti-inflammatory effect against choroidal inflammation and effusion. Kumar et al.[12] indicated that nonspecific choroidal inflammation might be the underlying cause of UES.

Few reports have documented the medical treatment of UES. Andrijevic Derk et al.[13] used oral carbonic anhydrase inhibitors and topical prostaglandin analogues in 3 cases and demonstrated apparent resolution of chorioretinal detachment in 2 cases. We present a further case that confirms that marked resolution of SRD in UES with nanophthalmic eyes could be obtained with medical therapy alone. Although a case–control study would be needed to rule out spontaneous resolution, it seems infeasible to perform because of the extremely rare incidence of UES.

In conclusion, medical therapy can be of value in the treatment of UES before invasive surgical treatment. Further studies on a larger number of patients with different clinical severities of UES are warranted to validate the efficacy of medical therapy.

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Optical coherence tomography angiography imaging of Purtscher retinopathy

Purtscher retinopathy is an occlusive microvasculopathy associated with trauma and was first described by Purtscher.[1] It usually results from head trauma or chest compression. A similar condition as a result of nontraumatic cause is referred to as "Purtscher-like retinopathy." The diagnosis is primarily clinical and includes unilateral or bilateral sudden vision loss with fundus features, including cotton wool spots, intraretinal hemorrhage, and pathognomonic Purtscher flecken (retinal whitening