

Mobile lens–induced angle closure glaucoma and rubeosis iridis in Sturge Weber syndrome

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Dear Sir,

I am Dong Hyun Lee from the Department of Ophthalmology at the Pusan National University Yangsan Hospital, Yangsan, Korea. Herein, I present the development of a lens-induced angle closed glaucoma and iris neovascularization in the patient with Sturge Weber syndrome (SWS) with open angle glaucoma. Informed consent was obtained from a patient before each procedure. The study was conducted in accordance with the tenets of the Declaration of Helsinki.

A 65-year-old man presented to our hospital with severe ocular pain, photophobia and a conjunctival injection of the left eye. The best corrected visual acuity was light perception and intraocular pressure (IOP) was 51 mm Hg in the left eye. The slit lamp examination revealed a deep anterior chamber and the gonioscopy showed visible trabecular meshwork corresponding to Scheie grade II. The fundus was invisible due to mature cataract (Figure 1). A huge choroidal mass was detected on ocular ultrasound (AVISO, Quantel Medical, France) (Figure 2A). A purple colored port-wine hemangioma was presented in the left face (Figure 2B). A calcified lesion in the frontal and occipital lobes, atrophy of

the left hemisphere (Figure 2C) and a choroidal mass (Figure 2D) were found in the computed tomography of the brain. He did not have any systemic co-morbid conditions as diabetic mellitus, hypertension and cardiovascular disease. Based on the above findings, we diagnosed the patient as SWS with secondary open angle glaucoma.

The IOP was controlled under 20 mm Hg for 6mo after the anti-glaucoma treatment with brimonidine tartrate 0.15% (Alphagan®-P, Allergan, USA), latanoprost 0.005% (Xalatan®, Pfizer, USA) and brinzolamide 1%/timolol 0.5% (Elazop®, Alcon, USA). However, the IOP increased to 38 mm Hg again in spite of maximal medical anti-glaucoma treatments. Very shallow anterior chamber and invisible trabecular meshwork (TM) resulting from lens subluxation and an iris neovascularization were detected (Figure 3A, 3B). Angle-closure glaucoma was superimposed on open angle glaucoma. The IOP decreased to 20 mm Hg again after laser iridotomy by double frequency Nd: YAG laser (PASCAL, Optimedia, USA) with anti-glaucoma eye drops. Two months later, laser iridotomy moved the lens increasingly forward (Figure 3A). The IOP was up to 60 mm Hg and severe ocular pain was developed. A dislocated lens into the anterior chamber was shown on ultrasound biomicroscopy (AVISO, Quantel Medical, France) (Figure 3A, 3B). We performed a lensectomy and vitrectomy because of the development of a concomitant malignant glaucoma. A big choroidal hemangioma, ischemic retinal change adjacent to choroidal lesion, retinal vascular sheathing and optic disc atrophy were observed at the time of surgery. One week after the surgery, IOP became 12 mm Hg.

SWS is a congenital disorder having influence on the cephalic venous microvasculature [1-3]. The ocular complications in SWS derive from vascular abnormalities of the conjunctiva, episclera, retina and choroid, including glaucoma, conjunctival, choroidal and retinal hemangiomas and exudative retinal detachment [1]. The prevalence of glaucoma, the most common ocular complication, was reported in 30% -71% of SWS patients [1,4]. Generally, glaucoma in SWS might be related with developmental anomalies of trabecular outflows, increased episcleral venous pressure, hemangioma in iris and angle or with massive

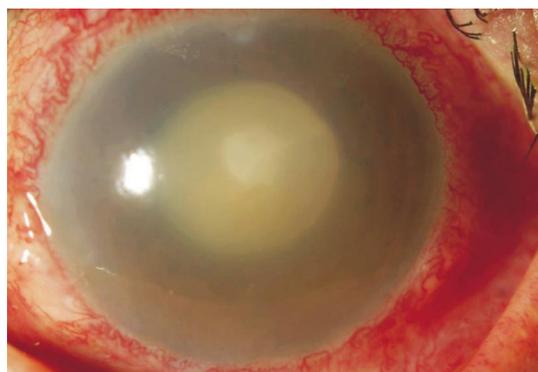


Figure 1 The photograph shows an edematous cornea due to increased IOP and a mature cataract at the presentation.

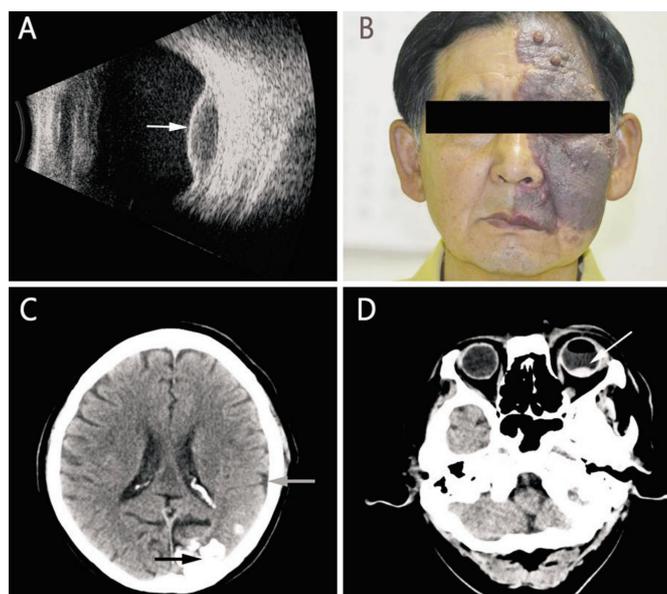


Figure 2 Representative images of the patient A: The ultrasonograph shows a choroidal hemangioma (white arrow); B: The dark color port-wine hemangioma is seen on the left face; C: The computed tomography of brain shows a cerebral atrophy (gray arrow) and left occipital and cortical calcifications (black arrow); D: A contrast enhanced computed tomography shows a well-defined mass with enhancement (white arrow).

exudates originating from choroidal hemangioma, especially in the side of facial hemangioma involving eyelid and conjunctiva [4]. Lens-related ocular complications and iris neovascularization were rarely reported in SWS patients[5,6]. Our patient showed a large retinal ischemic lesion overlying a choroidal hemangioma. A choroidal hemangioma has been reported in up to 40% of SWS patients [7]. Extensive exudative retinal detachment was developed in the large choroidal hemangioma which induced severe retinal ischemia, iris neovascularization and a neovascular glaucoma, although the anterior segment ischemia has been a rare complication in SWS [8]. However, our presented patient had a mature cataract and an iris neovascularization without exudates. Previous study of perfusion magnetic resonance image scan reported that the venous blood flow impairment

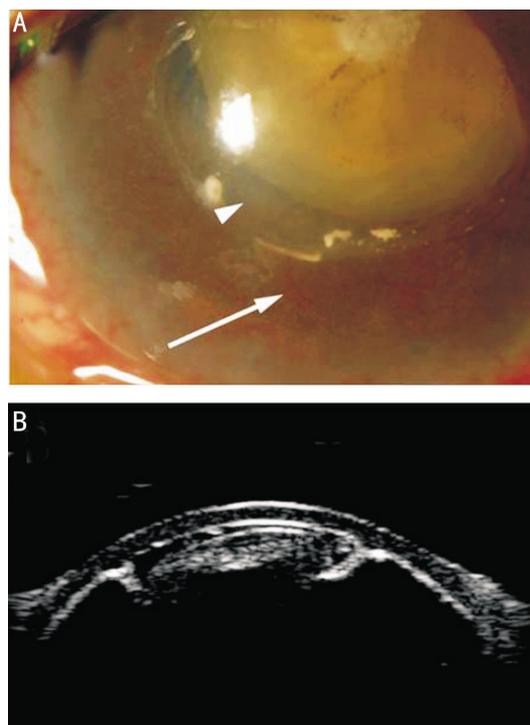


Figure 3 Photograph and ultrasound biomicroscopy of anterior segment A: The photograph shows that corneal edema decreased after anti-glaucoma treatment. Dislocated lens and zonular dialysis (arrowhead) developed after laser iridotomy. Neovascularization of the iris was detected (arrow). B: Ultrasound biomicroscopy shows that the crystalline lens dislocated into the anterior chamber.

and a perfusion defect in the cortex adjacent to a huge hemangioma were found, and the cortical ischemia resulted in secondary vascular proliferation in the brain of the SWS patient[7]. This finding supported that a severe ischemic status could develop in the posterior segment due to the large choroidal hemangioma, which might have induced the anterior ischemia resulting in iris neovascularization, mature cataract and zonular weakness in our patient.

Another cause of lens and zonule complications in SWS might be an embryological anomaly of the neuroectoderm[1]. This developmental anomaly could contribute to the malignant glaucoma in our patient due to the anterior dislocation of the mobile lens with zonular weakness. The IOP sufficiently decreased after lensectomy in this case although it was reported as ineffective in the treatment of increased IOP associated with lens dislocation in SWS[6].

In conclusion, a huge choroidal hemangioma could induce an ischemic change in the overlying retina, which was likely the main cause of secondary iris neovascularization and mature cataract. Angle closure glaucoma caused by lens dislocation and zonular laxity could develop in the patient with SWS.

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