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A Case of Acute Angle-closure Glaucoma Secondary to Posterior Scleritis in Patient with Sturge-Weber Syndrome

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Background: Sturge-Weber syndrome has been known to be frequently associated with facial cutaneous angioma and ipsilateral glaucoma. However, as far as we know, no cases accompanied by acute angle-closure glaucoma have been reported in patients with Sturge-Weber syndrome.

Case: A 14-year-old boy with unilateral acute angle-closure glaucoma secondary to posterior scleritis associated with Sturge-Weber syndrome is described.

Observations: Slit-lamp examination revealed diffuse episcleral venous hemangioma in the right eye. With ultrasound biomicroscopy, a forward shift of the lens-iris diaphragm, a swelling of the ciliary body, and an anterior rotation of the ciliary processes with annular choroidal effusion were detected. The patient responded well to treatment with systemic corticosteroids and cycloplegics.

Conclusions: In our patient, inflammatory changes of the sclera, including swelling of the ciliary body, choroidal effusion, an anterior rotation of the ciliary processes at the scleral spur, and swelling of the lens, leading to closure of the anterior chamber angle, were suggested to be the major mechanisms of intraocular pressure elevation. **Jpn J Ophthalmol 2002;46: 74–77**

Key Words: Acute angle-closure glaucoma, choroidal effusion, scleritis, Sturge-Weber syndrome.

Introduction

Sturge-Weber syndrome represents encephalotrigeminal angiomatosis, and is known to be frequently associated with facial cutaneous angioma and glaucoma.[1,2] The glaucoma secondary to Sturge-Weber syndrome occurs ipsilateral to the hemangioma in newborns, children, or adults and is characterized as chronic and mild elevation of intraocular pressure. As the possible pathogenesis of the glaucoma in Sturge-Weber syndrome, an abnormal angle structure[3] and elevated episcleral venous pressure[4] have been involved, although the precise etiology of the glaucoma has not been elucidated. However, as far as we know, no report of a case with acute angle-closure glaucoma secondary to posterior scleritis in Sturge-Weber syndrome has been published.

Herein, we describe a rare case of unilateral Sturge-Weber syndrome with ipsilateral acute angle-closure glaucoma secondary to posterior scleritis. The patient's glaucoma was well controlled by cycloplegics and topical and systemic corticosteroids and no surgical treatment was required.

Case Report

A 14-year-old-boy was referred to our hospital for sudden onset of ocular pain and visual loss in his right eye associated with severe headache and nausea. He had no history of systemic diseases such as juvenile rheumatoid arthritis, rheumatic fever, or other immunogenic disorders. He had a port wine stain on his face, which involved the region of distribution of the first and second divisions of the right trigeminal nerve. His best-corrected visual acuity was 0.01 OD and 1.0 OS at the initial examination.

Received: February 2, 2001
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Published by Elsevier Science Inc.

0021-5155/02/$–see front matter
PII S0021-5155(01)00463-4

Intraocular pressure was 48 mm Hg OD and 10 mm Hg OS. Ophthalmic examination revealed no abnormalities in the left eye. Slit-lamp examination of his right eye showed a slightly dilated pupil with corneal and conjunctival edema and diffuse episcleral venous hemangioma with subconjunctival hemorrhage (Figure 1). The anterior chamber did not have either flare or cells. Anterior chamber depth was 2.09 mm OD and 3.60 mm OS, and lens thickness was 4.26 mm OD and 3.49 mm OS, respectively. Gonioscopic examination revealed complete occlusion of the angle OD and open angle OS. Fundus examination identified a localized choroidal detachment in the temporal periphery and a slightly hyperemic disc with 0.5 cup-to-disc ratio in his right eye. Fluorescein angiography showed leakage of the retinal veins overlying the choroidal detachment, although no signs suggested a choroidal hemangioma. In the examination of the right eye by ultrasound biomicroscopy, a forward shift of the lens-iris diaphragm, a swelling of the ciliary body and an anterior rotation of the ciliary processes with annular choroidal effusion were disclosed (Figure 2). Computed tomography revealed a slight proptosis, thickening and anterior shift of the lens, choroidal detachment, and thickening of sclera in the right eye (Figure 3), but no abnormal intracranial regions.

Based upon the above clinical findings, acute angle-closure glaucoma secondary to posterior scleritis was suggested to be associated with Sturge-Weber syndrome. He was treated with acetazolamide (Diamox®, Takeda, Osaka) 750 mg three times a day, topical instillations of timolol maleate (0.5% Timoptol®; Banyu, Tokyo) twice daily, dolzolamide chloride (1% Tolsopt®; Banyu) three times a day, and cyclopentolate chloride (1% Cyplesin®; Santen, Osaka) and betamethasone sodium phosphate (0.1% Rinderon®; Shionogi, Osaka) three times a day. Four days later systemic prednisolone (Predonine®; Shionogi) 30 mg twice daily was added to the above medications. After systemic administration of the corticosteroid, the intraocular pressure (IOP) decreased in the right eye. Twelve days later, ophthalmic examinations revealed significant recovery. Visual acuity in his right eye had increased to 1.5, the anterior chamber became deeper (3.81 mm), lens thickness became thinner (3.59 mm), choroidal detachment disappeared, and cup-to-disc ratio became smaller (0.3). However, a relatively low degree of peripheral anterior synechia (PAS) remained within the upper quadrant angle.

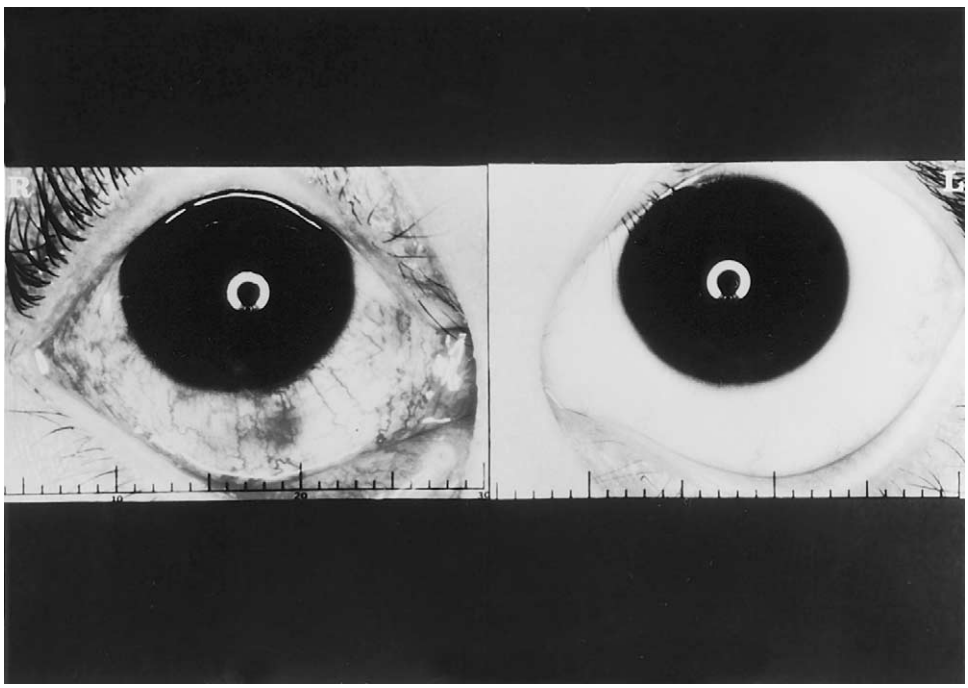

Figure 1. Slit-lamp micrograph of right and left eyes at initial examination. Right eye showed slightly dilated pupil with corneal and conjunctival edema and diffuse episcleral venous hemangioma with subconjunctival hemorrhage. Left eye showed no abnormality.

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Vol 46: 74–77, 2002

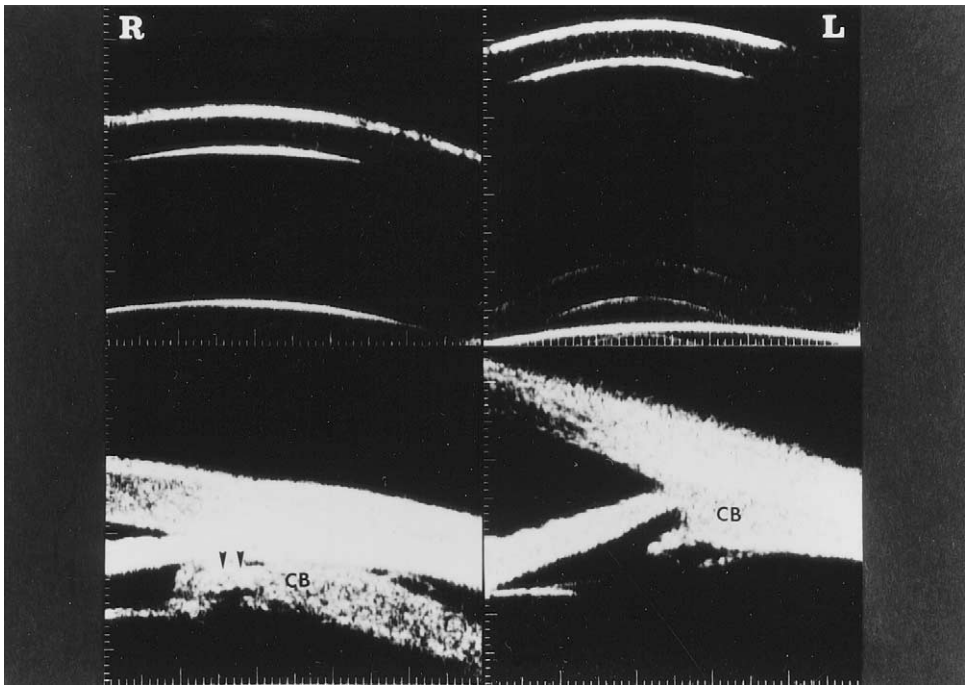

Figure 2. Biomicrograph of right and left eyes at 9 o'clock position. Ultrasound biomicroscopy disclosed forward shift of lens-iris diaphragm, swelling of ciliary body and anterior rotation of ciliary processes (arrowheads) with annular choroidal effusion. CB: ciliary body.

Discussion

Secondary glaucoma in patients with Sturge-Weber syndrome has been characterized by the presence of an abnormal angle structure resembling congenital glaucoma with anterior insertion of the iris root and a thick uveoscleral meshwork, in addition to elevated episcleral venous pressure.[5] Therefore, it was suggested that (1) a choroidal hemangioma leading to a shallow anterior chamber and subsequent angle-closure and (2) hypersecretion of aqueous by a choroidal hemangioma may be involved in the development of secondary glaucoma. As far as we know, elevation of IOP is usually chronic or subacute, but not acute. However, in our present case, acute elevation of his IOP had been recognized. Therefore, we considered that, in addition to the active glaucoma state caused by Sturge-Weber syndrome, some undiagnosed inflammatory changes of the sclera (posterior scleritis) including swelling of the ciliary body, choroidal effusion, anterior rotation of the ciliary processes at the scleral spur and swelling of the lens, facilitated the closure of the anterior chamber angle. In fact, the glaucoma in our patient responded well to systemic corticosteroids and cycloplegics. Because there has been no reported case of posterior scleritis associated with Sturge-Weber syndrome so far, any relationship between posterior scleritis and Sturge-Weber syndrome is unclear. This could be a simply coincidental case.

Posterior scleritis is an uncommon condition[6] known to have an acute onset such as unilateral signs of diffuse anterior scleritis, proptosis, and limitation of eye movements secondary to scleral thickening, together with an exudative retinal detachment. In addition, angle-closure glaucoma has been noted as one of the severe complications[7,8] of posterior scleritis. The mechanism of the development of glaucoma has been suggested to include angle-closure glaucoma caused by choroidal effusion or PAS formation and neovascular glaucoma.[9]

In terms of treatment in Sturge-Weber syndrome, goniotomy or trabeculotomy should be considered in newborns and in the presence of an abnormal angle appearance. In older patients, medical therapy should be tried, although many patients respond poorly and surgery often becomes necessary. However, trabeculectomy is frequently unsuccessful because a sudden uveal effusion with detachment of the choroid can occur in up to 24% of cases when the anterior chamber is penetrated during the procedure.[10] Our patient did not require any surgical intervention and responded rapidly to treatment of the posterior

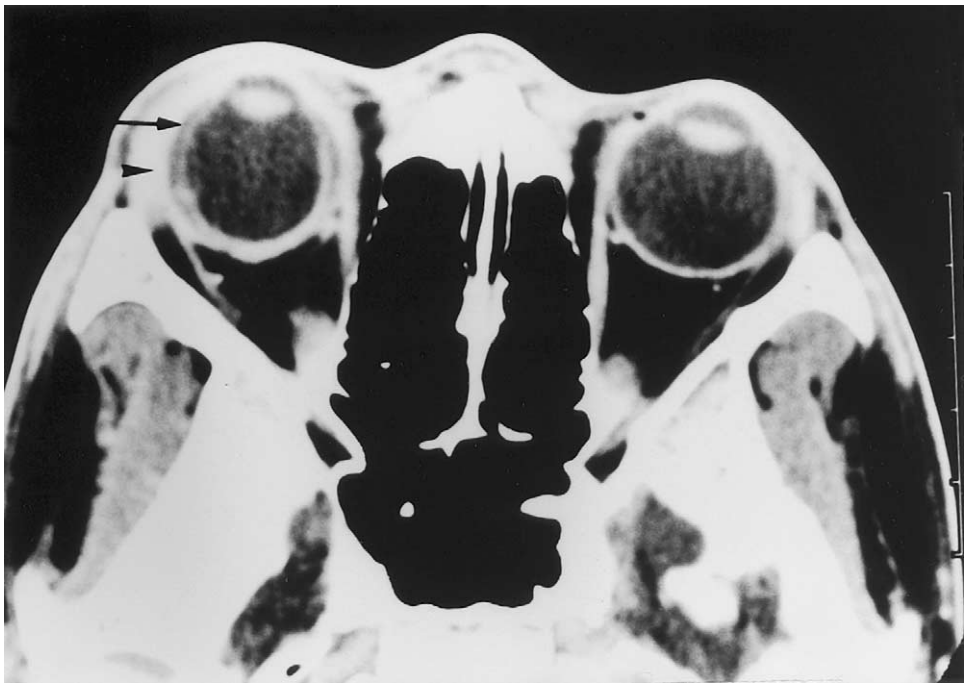

Figure 3. Computed tomograph (CT) of head. CT image revealed slight proptosis, thickening and anterior shifting of lens, choroidal detachment (arrow), and thickening of sclera (arrowhead) in right eye. There are no abnormal intracranial regions.

scleritis. There has been no sign of recurrence for at least 6 months after remission.

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