

An Incidental Dyke-Davidoff-Masson Syndrome Accompanying Sturge-Weber Syndrome: Case Report

Sturge-Weber Sendromuna Eşlik Eden Tasadüfi Dyke-Davidoff-Masson Sendromu

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ABSTRACT A 24-years-old female patient has been followed for glaucoma secondary to left sided cutaneous vascular nevus on her face. Having the previous diagnosis of Sturge-Weber syndrome (SWS), she had right nasal hemianopsia and almost total loss of visual field in the left eye associated with left glaucomatous optic atrophy. She had undergone cranial magnetic resonance imaging (MRI) for right nasal hemianopsia and for evaluation of intracranial involvement of SWS. Cranial MRI revealed enlargement of frontal sinus, elevation of the left orbital roof and left cerebral hemiatrophy. These imaging findings were interpreted as Dyke-Davidoff-Masson syndrome (DDMS). We report what we believe to be the second case in the literature of SWS occurring together with DDMS.

Key Words: Sturge-Weber Syndrome; glaucoma

ÖZET 24 yaşında kadın hasta, yüzünün sol tarafındaki kütanöz vasküler nevüse bağlı glokom nedeniyle takip edilmekte idi. Daha önce de Sturge-Weber sendromu (SWS) tanısı olan hastanın, sağ nazal hemianopsi ve sol glokomatöz optik atrofiye bağlı sol gözde neredeyse total görme alanı kaybı mevcuttu. Sağ nazal hemianopsiyi ve SWS'nin kafa içi tutulumunu değerlendirmek için beyin manyetik rezonans görüntülemesi (MRG) yapıldı. Beyin MRG'sinde frontal sinüste genişleme, sol orbital tavanda yükselme ve sol serebral hemiatrofi görüldü. Bu görüntüleme bulguları Dyke-Davidoff-Masson sendromu (DDMS) olarak yorumlandı. Bu olgu raporuyla, literatürde bildirilen SWS ve DDMS'nin ilişkili olduğu ikinci olguyu sunmaktayız.

Anahtar Kelimeler: Sturge-Weber Sendromu; glokom

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Sturge-Weber syndrome, otherwise called encephalo-trigeminal angiomas, is a rare neurocutaneous syndrome, characterized by facial capillary malformation (nevus flammeus or port wine stain), ipsilateral leptomeningeal venous angiomas and ocular abnormalities.¹ The most common clinical features include progressive seizures, ipsilateral glaucoma, contralateral hemiparesis, hemiatrophy and hemianopia.² Magnetic resonance imaging can demonstrate choroid plexus enlargement and cerebral atrophy, while computed tomography also shows cortical calcification.^{1,3}

Dyke-Davidoff-Masson syndrome is a disorder characterized by rare epilepsy syndrome characterized by seizures, facial asymmetry, contralateral hemiparesis or hemiplegia, and mental retardation. This disorder was

first described by Dyke, Davidoff and Masson in 1933.⁴ The specific imaging findings to support the diagnosis are cerebral hemiatrophy, ventricle enlargement, and shift to the affected side, with compensatory skull changes.

We report the second case in the literature, in which SWS and DDMS are associated.

CASE REPORT

24 years old female with a previous diagnosis of Sturge-Weber syndrome (SWS) was followed for glaucoma in the left eye. Her visual acuities were 20/25 and 10/200 in the right and left eyes respectively. Intraocular pressures were measured as 17 mmHg in the right eye without medication and 19

mmHg with latanoprost 1×1 eye drop. Biomicroscopic examination was normal in the right eye while dilated conjunctival vessels were observed in the left eye (Figure 1). On fundus examination, the right optic disc appeared slightly pale while glaucomatous optic atrophy with a total cup was observed in the left eye (Figure 2). Neuro-ophthalmologic examination demonstrated relative afferent pupillary defect and complete loss of color vision in the left eye. Visual field examination revealed right nasal hemianopsia and generalized depression in the right eye (Figure 3). Cranial MRI was performed to evaluate the intracranial involvement of the disease. The imaging findings including leptomeningeal venous angiomatosis, enlargement of transmedullary veins, periventricular veins and choroid plexus, en-

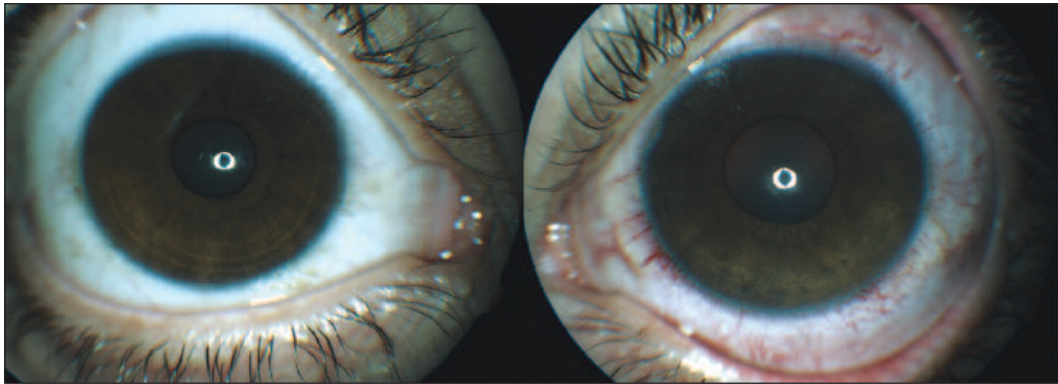


FIGURE 1: Biomicroscopic examination of the right and left eyes.

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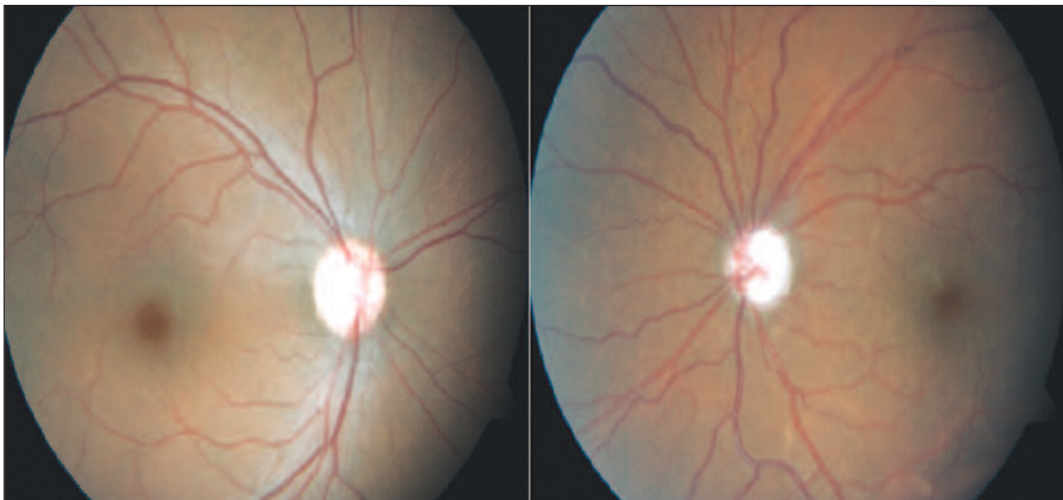


FIGURE 2: Fundus examination of the right and left eyes.

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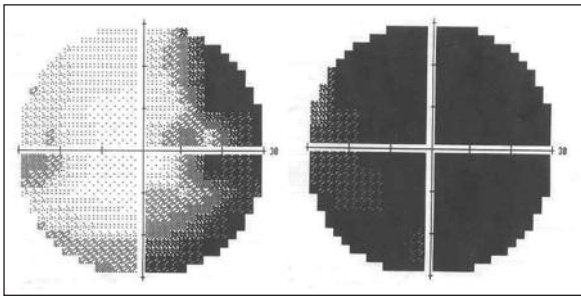


FIGURE 3: Visual field examination of the right and left eyes.

largement of paranasal sinuses which is more prominent in the right frontal sinus, elevation of the left orbital roof, left diplopic calvarial expansion and left cerebral hemiatrophy were observed (Figure 4). MR Susceptibility weighted imaging (SWI) revealed the enlarged abnormal cerebral veins connected to the periventricular veins (Figure 5).

The radiologic diagnosis was Sturge-Weber syndrome and Dyke-Davidoff-Masson syndrome (DDMS).

DISCUSSION

Sturge-Weber syndrome is a sporadic disease which belongs to a group of disorders named as phacomatoses.^{3,5} The classical triad is characterized by a facial capillary malformation (port-wine stain), ipsilateral leptomeningeal venous an-

giomatosis and ocular abnormalities.^{1,6} The most common ocular manifestation of SWS is glaucoma.⁷ The abnormal cortical venous drainage causes cerebral abnormalities involving cerebral atrophy, and cortical calcification.³ Magnetic resonance imaging can demonstrate enlarged choroid plexus and cerebral atrophy. Specifically, T1 weighted gadolinium post contrast MRI and MR Susceptibility weighted imaging (SWI) can demonstrate focal cortical atrophy, contrast-enhancing leptomeningeal angiomas, and abnormal cerebral veins, and is currently an imaging standard for clinical SWS diagnosis.

Dyke-Davidoff-Masson syndrome is a rare disorder characterized by seizures, facial asymmetry, contralateral hemiparesis or hemiplegia and mental retardation. The pathogenesis is associated with decreased arterial vascular supply, which may include absence of one internal carotid artery or posterior cerebral artery or hypoplasia of cerebellar arteries.⁸ The diagnosis mostly depends on the specific radiologic findings including cerebral hemiatrophy, enlargement of frontal sinus, elevation of the orbital roof, and skull changes including diplopic calvarial expansion, elevation of the petrous ridge. In addition to the vascular origin which is mostly congenital, various causes of cerebral injuries including trauma, infection and ischemic states, may lead to the acquired type of the disease.^{7,9}

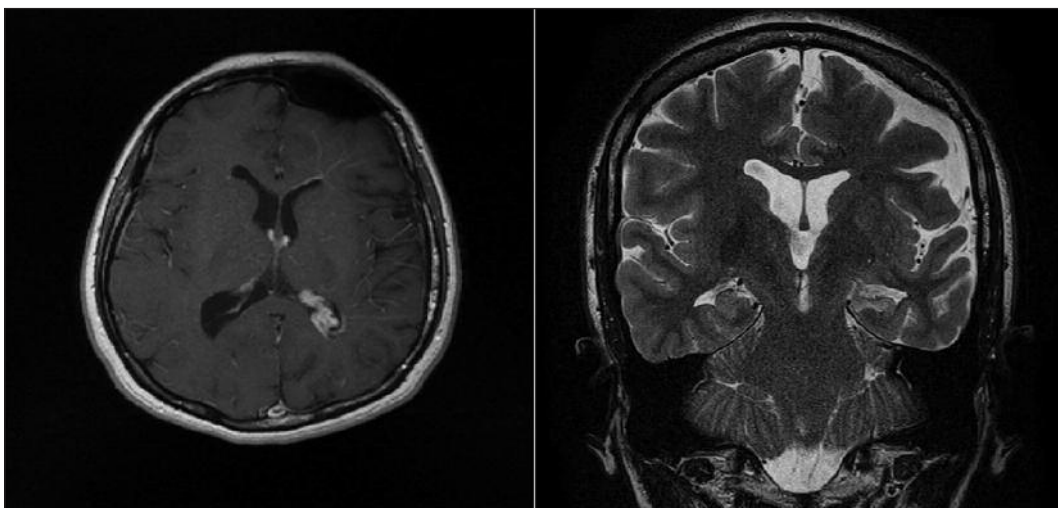


FIGURE 4: T1 post-gadolinium MRI (a) and coronal T2 weighted MRI (b) demonstrate enlargement of left frontal sinus, left cerebral hemiatrophy, and enlargement left choroid plexus.

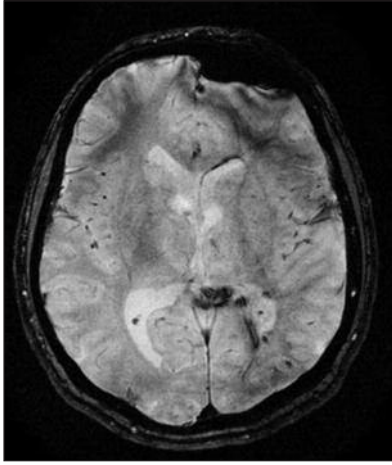


FIGURE 5: On the MR Susceptibility weighted imaging (SWI) the enlarged abnormal cerebral veins connected to the periventricular veins are observed.

The differential diagnosis the SWS patients either with or without DDMS may be provided by a careful radiological examination. The case presented in this paper is the second reported case of SWS accompanied by DDMS.³ A case with capillary malformation of the face (without SWS diagnosis) and DDMS has also been reported in a pediatric patient.⁵ Unlike the classical presentation of DDMS, our patient denied any history of seizure or hemiparesis.

Although being suggested as an arterial insufficiency disorder, DDMS can also occur with venous insufficiencies including SWS, as in the presented case.

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