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Case Report

A CASE OF STURGE-WEBER SYNDROME WITH SUDDEN DETERIORATION IN PREGNANT LADY WITH COVID-19

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ABSTRACT

Sturge-Weber syndrome has been included in the neurocutaneous disorders that is characterized by hamartomas involving the brain, skin, and eyes. Facial port-wine stain is characteristic. It involves the first branch of the trigeminal nerve and the embryonic vasculature distribution in this area leading to several ocular complications of the anterior segment with involvement of the eyelids and conjunctiva. Diffuse choroidal hemangiomas can occur in the posterior segment of the eyes. However, glaucoma is the most frequent ocular comorbidity with a prevalence rate ranging from 30%–70%. We report a case report of Sturge–Weber syndrome in a pregnant patient with facial port wine stain and seizures since birth, glaucomatous atrophy on fundoscopy and brain atrophy in MRI.

Key words: Sturge-Weber syndrome, Glaucoma, Choroidal hemangiomas, Port-wine stain.

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INTRODUCTION

Sturge-Weber syndrome (SWS) is a rare, congenital neuro-oculocutaneous disorder. Its manifestations are varied and include unilateral port-wine birthmark (PWB), intracranial leptomeningeal angioma, hemianopia, glaucoma, choroidal hemangioma, hemiatrophy of brain, hemiparesis, progressive seizures, and cognitive impairment.[1] SWS can be classified as trisymptomatic when the skin, eye, and central nervous system (CNS) are affected; bisymptomatic when the skin and CNS or the skin and eye are affected; and monosymptomatic when the skin or the CNS is affected.[2] The typical triad consists of facial capillary malformation (port wine stain or nevus flammeus), an ipsilateral vascular anomaly in the brain (leptomeningeal hemangioma), and ocular hemangioma.[3]

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Ocular manifestations of SWS include conjunctival, episcleral, and choroidal hemangiomas. Choroidal hemangiomas may be present in up to 71% of patients with SWS and may be circumscribed or diffuse.[4] Retinal vascular abnormalities are rare and include vascular tortuosity, arterio-venous malformations.[5] Glaucoma presents in 30-70% of patients with SWS.[4] Approximately 60% of patients present with glaucoma at birth and 40% manifest glaucoma later in life.[6] The incidence of glaucoma increases when the PWS involves the eyelid.[7] It presents most often ipsilateral to the PWS but can also manifest bilaterally.[8]

The embryologic basis of SWS has been reported to be related to an impaired development of the cell precursors in the neural crest during the first embryological trimester, leading to the characteristic malformations observed in the central nervous system, skin, and eyes.[9] Diagnosis is easily performed when the classical clinical signs of SWS are present, consisting of unilateral facial PWS along the first branch of the trigeminal nerve,

hemiatrophy, progressive seizures, contralateral hemiparesis, mental deficiencies, hemianopia, and ipsilateral glaucoma.[10]

Case report

A 24 year old primigravida with 34 weeks period of gestation (POG) was admitted in gynaecology department with pregnancy induced hypertension. On ultrasound IUGR was detected. Patient complained of facial and periorbital puffiness for which the patient was referred to the Ophthalmology department. The patient has history of port wine stain (PWS) and bilateral ocular pain for 2 years. The patient also gave history of having seizure episodes since birth for which she is currently not taking any treatment.

Figure 1. Port wine skin discolouration on left side of face along with bilateral upper and lower eyelid before COVID 19

Figure 2. Fundoscopy showing advanced Glaucomatous changes

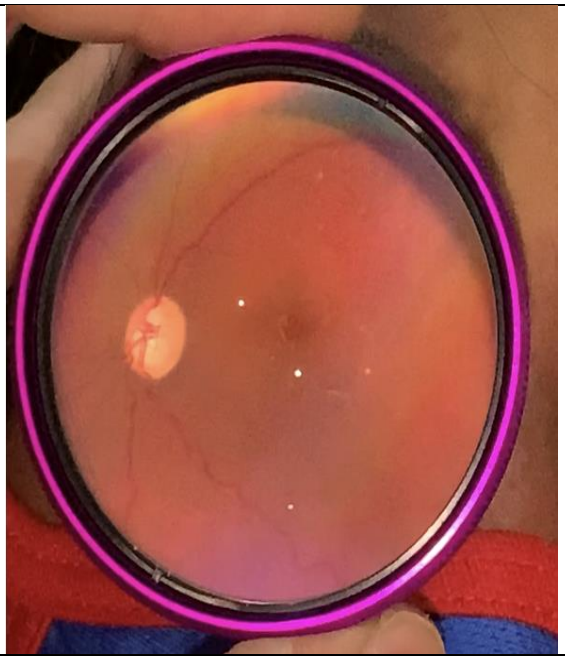


Figure 3. Indirect Fundoscopy showing Retinal detachment

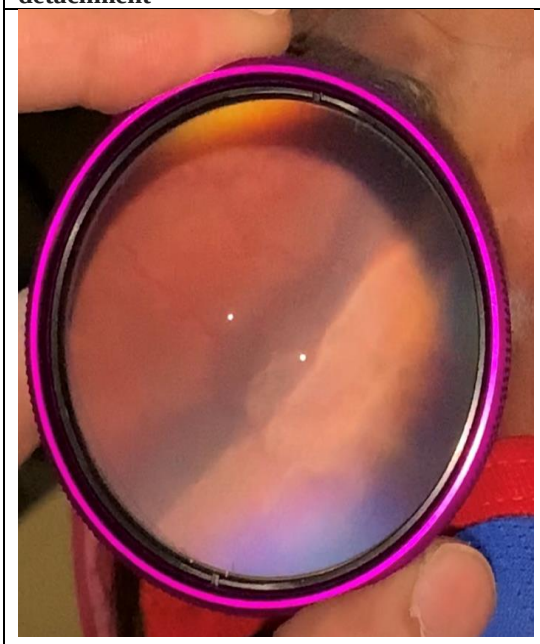


Figure 4. MRI brain showed atrophy of right temporo-occipital area and gyriform hypo intensity on T2 images

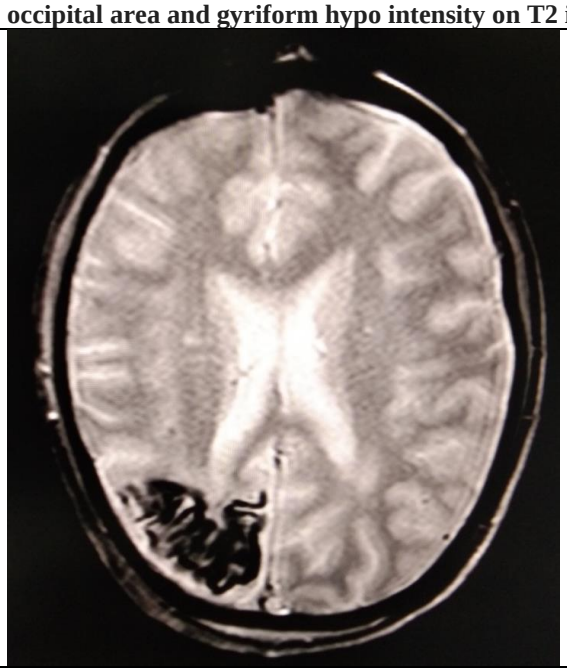
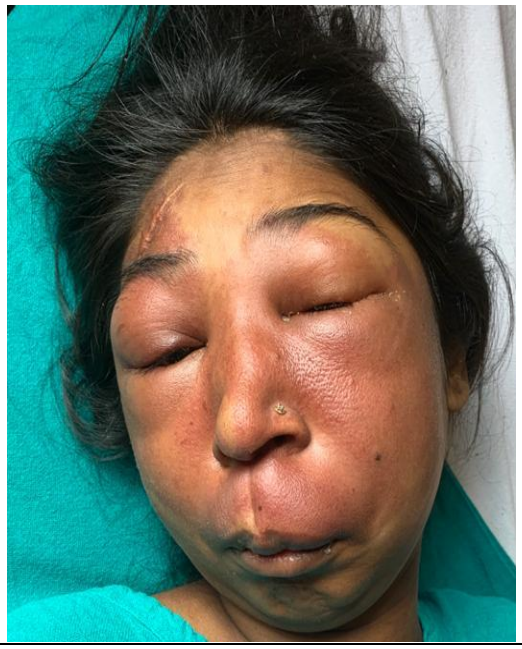


Figure 5. Deterioration of the patient 3 days after testing positive for COVID 19



On examination, port wine skin discolouration on left side of face along with bilateral upper and lower eyelid involvement (Figure 1) was present before COVID. On ocular examination, the patient had BCVA of 6/18 in right eye and finger counting at one metre in left eye with relative afferent pupillary defect (RAPD) in left eye. IOP was 25 mm of Hg and 46 mm of Hg in right eye and left, respectively. On fundus examination, bilateral distinct disc margin with cup disc ratio (CDR) of 0.8:1 right eye and 0.9:1 in left eye was seen(Figure 2), suggestive of bilateral glaucomatous optic atrophic changes. Tortuosity of vessels was noted in supero temporal area which upon being traced ended into retinal detachment (RD) (Figure 3) suggesting choroidal hemangioma in left eye. B-scan, MRI brain and neurological examination for further evaluation of patient was advised. MRI brain showed atrophy of right temporo-occipital area and gyriform hypo intensity in same area on T2 images (Figure 4) suggestive of Sturge-Weber syndrome. The patient was started on IOP lowering non-teratogenic drugs.

Follow-up of patient was done after 3 days. Patient had sudden deterioration of SWS with PWS edema(Figure 5) and abruptio placentae associated with fetal distress. COVID status was positive. The patient was taken up for emergency LSCS and baby shifted to NICU after delivery. The patient was shifted to COVID ward for further management.

Discussion

Pregnant patient with Sturge-Weber syndrome associated with glaucomatous changes, choroidal hemangioma and RD are rarely seen together. In literature, no evidence of sudden deterioration of SWS sign is present and the case was reported with safe delivery in SWS. In this case, the patient with positive COVID-19 status might have developed vascular abnormalities due hypercoagulable state causing deterioration in hemangioma, abruptio placenta and fetal distress due to which emergency LSCS was done. Such simultaneous occurrence of fundus abnormalities have not been reported in the literature before.

Acknowledgement

None.

Conflict of interest

The authors declare that they have no conflict of interest.

Statement of Humans and Animal Rights

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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