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Case Report

Sturge-Weber Syndrome: A Case Report

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Abstract: Sturge-Weber syndrome (SWS) is a rare, congenital, neuro-oculo-cutaneous disorder which is characterized by port-wine stain (facial nevus), glaucoma, seizures, hemiparesis, intracranial calcification and mental retardation. In the present case, a 17-year-old male patient presented with a port wine stain on the left side of the face, glaucoma of the left eye, seizure and weakness of right sided weakness of body.
Keywords: SWS - Sturge-Weber syndrome

INTRODUCTION

Sturge- Weber-syndrome or encephalo-trigeminal angiomatosis is a rare nonhereditary developmental condition. It is characterized by angiomatosis of face with a variable distribution sometimes matching the dermatomes of one or more divisions of trigeminal nerve [1, 2]. SWS is believed to be caused by the persistence of vascular plexus around the cephalic portion of the neural tube. This plexus develops during the sixth week of intrauterine development but normally undergoes regression during ninth week [2].

Schirmer provided the first detailed description of SWS in 1860, Sturge further described SWS-related dermatological, ophthalmic and neurological manifestations in 1879, and Weber reported radiological alterations seen in these patients in 1929 [3]. This disorder occurs in 1:50,000 live births [4]. Both sexes are affected equally and no racial predilection is seen [5].

The main characteristic features of this disorder are unilateral facial nevus, seizures and mental retardation and glaucoma [6]. The facial nevus is present at birth and tends to be unilateral and involve forehead and scalp. The nevus may also be evident over the lower face, trunk, and in the mucosa of the mouth and pharynx. Seizures are typically focal, tonic-clonic and contralateral to the side of the facial nevus. The seizures are associated with a slowly progressive hemiparesis in most of cases. Buphthalmos and glaucoma of the ipsilateral eye are a common complication [7].

CASE PRESENTATION

A 17-year-old male patient presented with history of seizures and weakness of right half of body. Seizure initially started on right side of the body then become generalized which was associated with post ictal confusion and urinary incontinence. The patient's past medical history revealed that he had developed a convulsive disorder at the age of 2 year for which he was taking medication.

General examination showed a port wine stain on the left side of the face (figure 1), while eye examination showed dilated blood vessels in the left eye (figure 2) and fundus examination shows Glaucomatous optic atrophy in left eye (fig 6). Oral examination showed unilateral gum hyperplastia (figure3), unilateral hyperplastic lesions on the left side of the maxilla (figure 4).

Fig-1: Port wine stain and hemihypertrophy on the left side of the face.

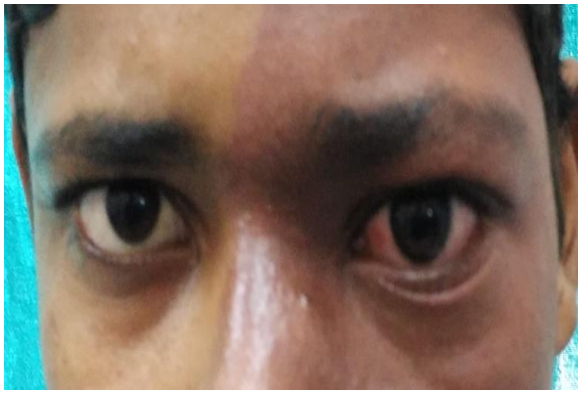

Fig-2: Dilated blood vessels in the left eye

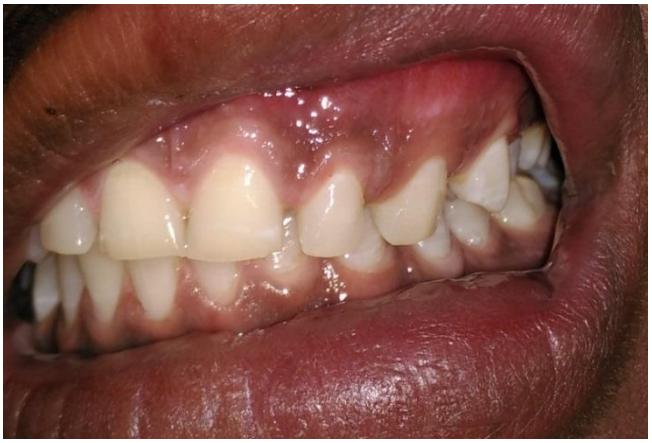

Fig-3: Unilateral gum hyperplastia.

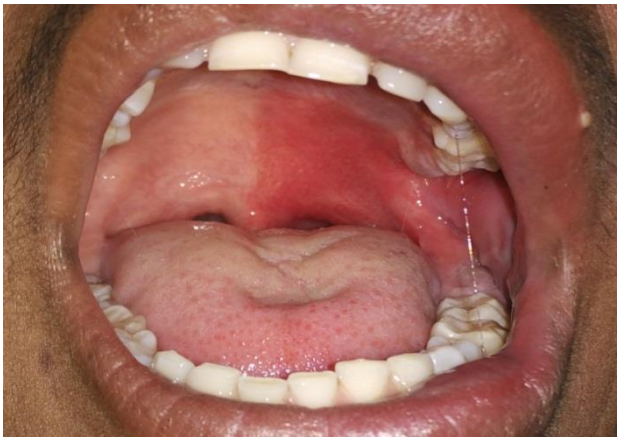

Fig-4: Hyperplastic left soft palate

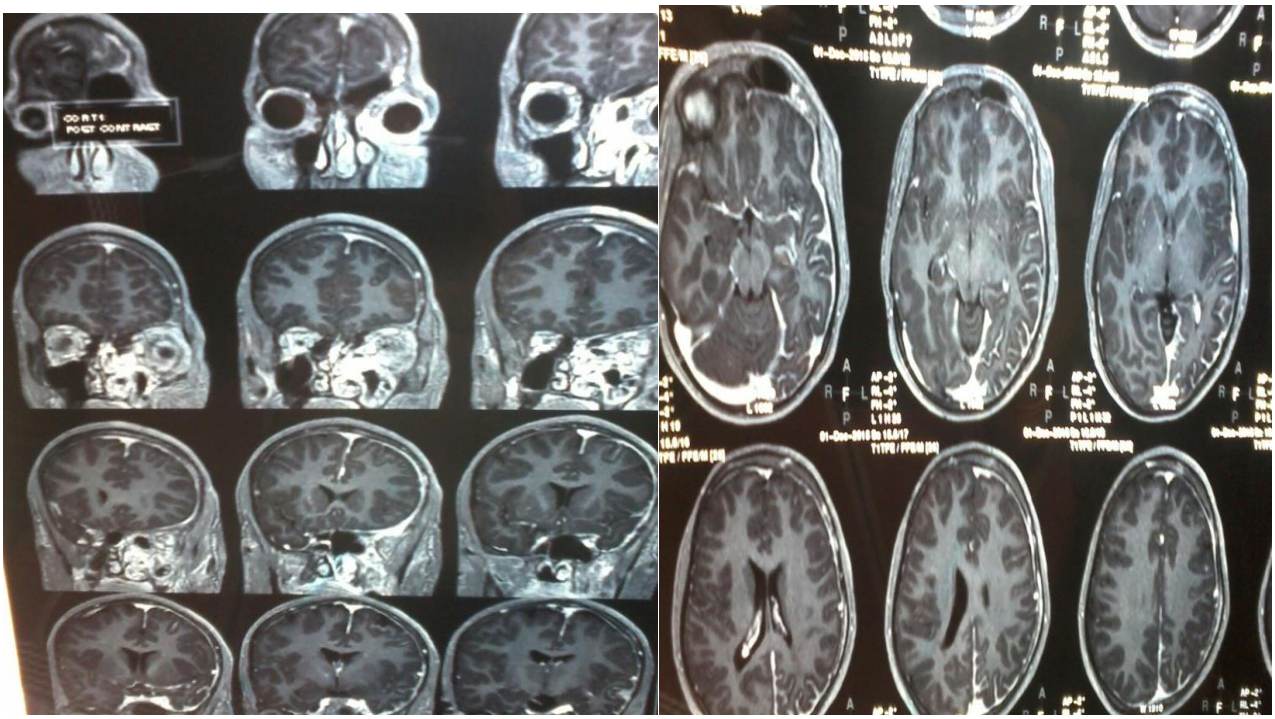

Fig-5: MRI showed

Abnormal signal intensity areas in left occipital and temporoparital lobeshowing hypointense signal on T1 and T2 and Blooming on GRE sequence.It is associated with adjacent cortical atrophy and prominent sulcul spaces.

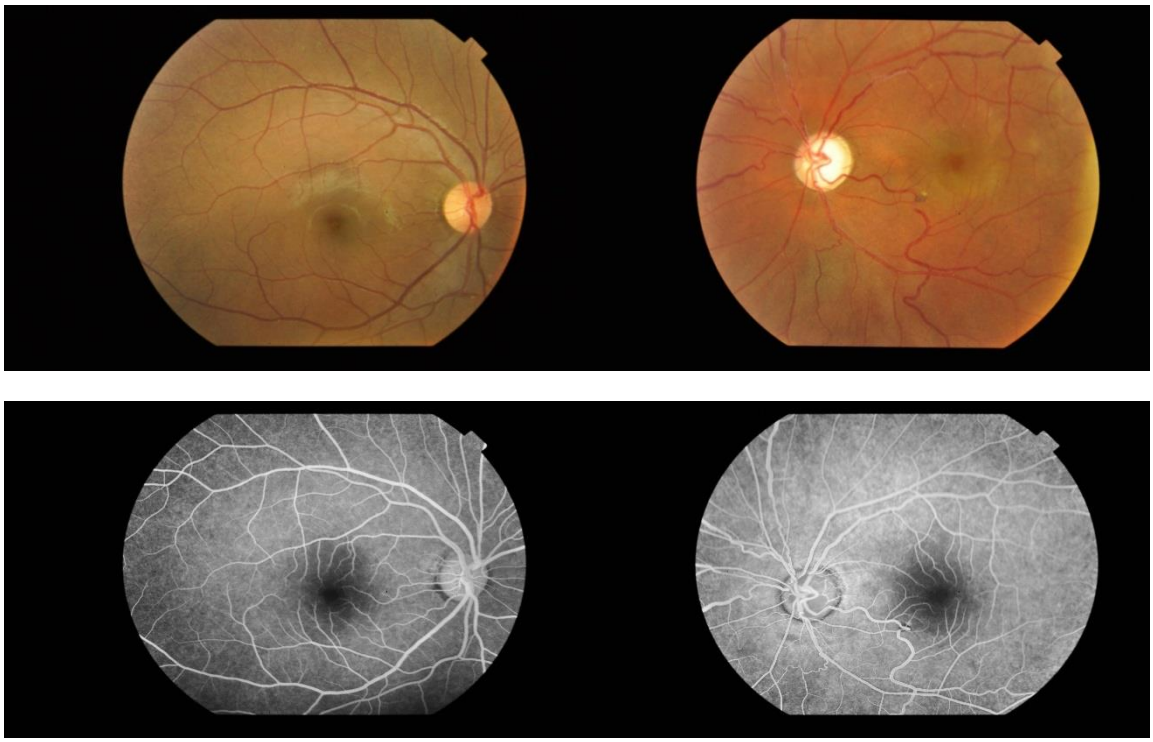

Fig-6: Left eye fundus examination shows Glaucomatous optic atrophy
Right eye fundus examination within normal limits

MMSE score was 28 and Blood investigations were normal. Based on the history, and on clinical, ophthalmological and radiological findings, diagnosis of Sturge Weber Syndrome was made.

DISCUSSION

SWS is referred to as complete when both CNS and facial angiomas are present and incomplete when only one area is affected without the other. The Roach Scale is used for classification, as follows [8] :

Type I - Both facial and leptomeningeal angiomas; may have glaucoma
Type II - Facial angioma alone (no CNS involvement); may have glaucoma
Type III - Isolated leptomeningeal angioma; usually no glaucoma.

According to the above criteria, our case is complete Type I SWS case.

Table 1 Clinical manifestations of SWS and manifestations seen in our case.

No.	Clinical Features	Incidence (%)	Present case
1.	Epilepsy	80	+
2.	Port-Wine Stain	76	+
3.	Abnormal Radiographic Findings	63	+
4.	Mental Retardation	54	-
5.	Oral Manifestations	38	+

6.	Hemiparesis	37	+
7.	Ocular Manifestations	37	+

In study of Sujansky et al, the age of onset of seizures was (0-23 years) and the relationship between the seizures and developmental delay was established. In those with and without seizures, the prevalence of developmental delay was (43% vs. 0%) [9]. Early onset of seizures and poor response to medical treatment, bilateral cerebral involvement and unilateral severe lesions were indicative of a poor prognosis [10]. In this case patient developed seizure at age of 2 year and seizure respond well to the treatment. This patient did not have development delays and higher functions were normal which was unusual finding.

The differential diagnosis included Rendu-Osler-Weber syndrome, angio-osteodystrophy syndrome, Maffucci syndrome and Klippel-Trenaunay-Weber syndrome [11].

Treatment and prognosis depends upon the nature and severity of clinical features. Presence of port wine stain can cause deep psychological trauma to patient and development of personality is affected in almost all patients [11]. Port wine stains can be improved by dermabrasion, tattooing, and flash lamp pulsed dye lasers [2]. Antiepileptics with folic acid supplementation given to prevent seizure. Physical therapy advised for paralysis or weakness while eye drops or surgery to treat glaucoma and Psychological counseling of parents were done.

During follow up, at the 1-month and 2-month and 4-month follow-up visits, there was a significant improvement in weakness of right side of body with no history of seizures.

CONCLUSION

Sturge-Weber syndrome presents with a large number of clinical manifestations and early diagnosis is difficult. Early diagnosis is necessary to avoid future complications. Expert psychological counselling of patients and their parents is necessary.

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