Brief Reports

Sturge-Weber Syndrome

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Sturge-Weber syndrome (SWS, encephalotrigemnial angiomatosis), is characterized by port-wine stain of the upper face, contralateral partial or secondary generalized seizures, contralateral hemiparesis and ipsilateral intracranial calcifications(1). In this communication, we are reporting a case with some uncommon features. We could not find a similar report in Indian literature.

Case Report

A 12-year-old girl born to non-consanguinous parents came with history of nevi on face, neck and trunk since birth; recurrent attacks of parethesias and weakness of left half of body since the age of 1½ years and diminishing vision for last 8 years. On examination she had bilateral facial nevi more extensive on right side (*Fig. 1*); nevi on neck and trunk; port wine stain on both sides of the tip of tongue, right side of the floor of the oral cavity and on right of the hard palate. Eye examination showed bilateral port wine stain of upper eyelids (*Fig. 1*), heterochromia and glaucoma (intraocular tension 31.3 and 50.8 mm Hg in right and left eye, respectively by Schiotz tonometer). Her developmental history, intelligence and systemic examination were normal. *X*-ray skull showed typical rail road track calcification in right occipital area. Computerized tomography showed right parieto-occipital calcification with underlying cortical atrophy on contrast studies. The rest of the family members were normal.

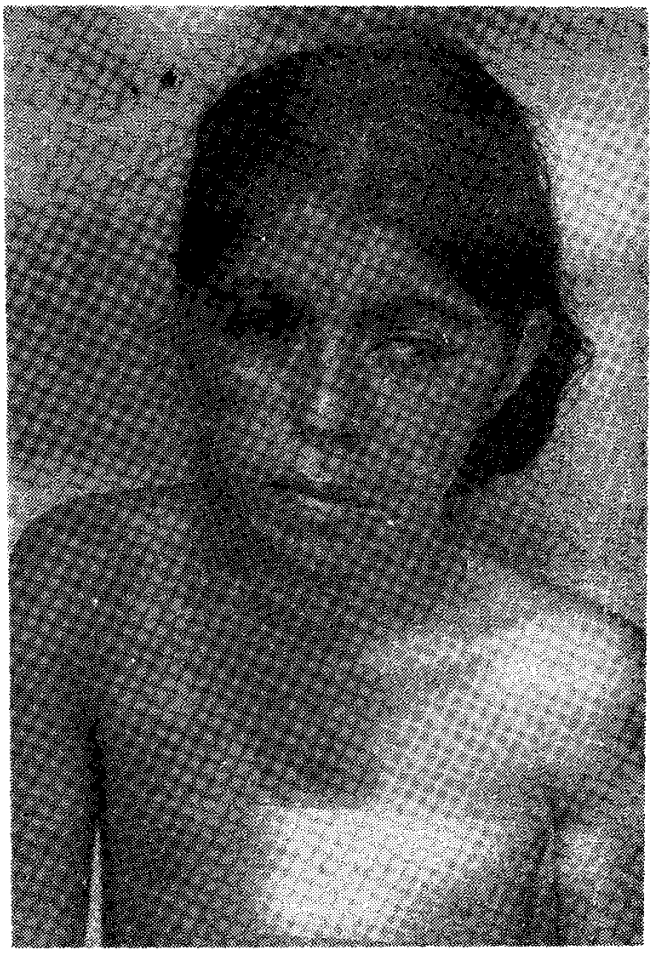

Fig. 1. Bilateral facial nevus.

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Discussion

Sturge(2) described a patient with syndrome and Weber(3) published a *X*-ray showing unilateral sclerosis in a similar patient. The interesting features of this case have been shown in *Table I*. In SWS, the basic defect is usually unilateral and rarely does the cutaneous abnormality extends beyond the distribution of the trigeminal nerve. It is intriguing to note the frequent presence of cutaneous malformation in the distribution of the ophthalmic division of the trigeminal nerve and concordent presence of the cerebral malformation in the occipital lobe, which is explainable embryologically(7,9).

The reported incidence of convulsions in SWS is 55-97%. There is no relation between the type and severity of the seizures with the extent of the cutaneous involvement. Both the cerebral hemispheres are involved in 15%(9). Central nervous involvement has not been reported if the nevus is entirely below the palpaberal fissure(7-8). Patients with leptomeningeal angiomatosis, who do not have the skin lesions are considered *forme fruste* of SWS or separate entity(10).

X-ray examination of the skull may reveal the characteristic calcification of meningeal angiomatosis before seizures have occurred, but rarely before two years. Computed axial tomography can establish early diagnosis but these procedures need not to be carried out if port wine stain does not involve the ophthalmic branch of the trigeminal nerve.

The ectopic melanosis is thought to be due to the developmental defects in neural tube and underlying mesodermal elements which may arrest normal migration of neural crest-derived melanoblasts(11).

The risk of eye abnormalities is more in those with nevus flammeus in the cutaneous distribution of the both first and second branches of trigeminal nerve. Nearly, 60% of those with glaucoma develop the condition prior to 2 years of age. Therefore, there is a need for regular supervision by an ophthalmologist(1).

Despite the frequent inclusion of SWS with the neurocutaneous genodermatoses, there is no evidence that the disorder is inherited.

TABLE I—*Uncommon Features of Index Case and Their Reported Incidence in Literature*

Clinical features	Incidence (%)	
Bilateral facial nevus	37	(4)
Nevus on trunk and neck	36	(5)
Oral mucosa lesions	25	(5)
Bilateral glaucoma	30	(6)

Figures in parentheses indicate reference numbers.

REFERENCES

1. Alexander GL, Norman RM. The Sturge Syndrome. Bristol England, John Wright and Sons Ltd, 1960.
2. Sturge W. A case of partial epilepsy, apparently due to lesion of one of the vasomotor centers of the brain. Clin Soc London Tr 1879, 12: 161-167.
3. Weber FP. Right-sided hemihypotrophy resulting from right-sides congenital spastic hemiplegia, with a morbid condition of the left side of the brain revealed by radiograms. J Neurol Psychopath Aug 1922, 3: 134-139.
4. Uram M, Zubillaga C. The cutaneous manifestations of Sturge-Weber Syndrome. J Clin Neuroophthalmol 1982, 2: 145-148.
5. Yingkun F, Yinchang Y. Sturge-Weber Syndrome: A report of 22 cases. Chin Med J 1980, 93: 697-708.
6. Clibis GW, Tripathi RC, Tripathi BJ. Glaucoma in Sturge-Weber Syndrome. Ophthalmology 1984, 91: 635-649.

7. Chao DH. Congenital neurocutaneous syndromes of childhood. III Sturge-Weber disease. J Pediatr 1959, 55: 635-649.

8. Jacob AH. Vascular malformation. *In:* Pediatric Dermatology, Eds. Schachner LA, Hansen RC. Vol 2. New York, Churchill Livingstone, 1988, p 1047.

9. Paller AS. The Sturge-Weber Syndrome. Pediatr Dermatol 1987, 4: 300-304.

10. Crosley CJ, Binet EF. Sturge-Weber syndrome. Clin Pediatr 1978, 17: 606-609

11. Bendl BJ, Bashir R, Dowling ADC. Sturge-Weber Syndrome. Cutis 1983, 31: 286-294.

Dietary Intake Amongst 'Well To Do' Adolescent Boys and Girls in Delhi

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It is well known fact that prosperity of a nation depends upon the quality of its human resources. The young women who are at the brink of womenhood, constitute the most crucial segment of our population from the point of view of the 'quality' of our future generation. Adolescence is a period of peak growth for boys and girls(1). Food and nutrient needs are proportionately higher during the growth spurt; adolescent students have been reported to be suffering from protein-calorie malnutrition because of less intake(2). The food intake of teenagers in the developed countries has been reported to vary and to be inadequate(3). The common causes of malnutrition among adolescents in the poor community are lack of food or less access to food and inadequate knowledge about dietary requirements(4). But what happens amongst the 'well to do' group. In India, adequate information on the dietary intake amongst adolescents belonging to 'well to do' group is not available. The present study was conducted to fill this gap in the knowledge.

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Material and Methods

The present study was conducted in a public school of Delhi, which catered to the urban elite. The tuition fee per child per month was Rs. 400/-. The school was selected by using purposive sampling keeping in view the operational feasibility. All the students studying in VII to XII standards were included for the detailed study The girls having menstrual period on days of study were excluded. For each student, information on the following parameters was collected like–age, sex, family income, family size, height, weight and dietary habits. All the students were categorized in two main groups as per age–Group A, students aged 13-15 years and Group B between 16-18 years.

The dietary intake of each student was recorded by an experienced dietician using