Faridpur Med. Coll. J. 2013;8(1): 44-45

Case Report

"Sturge-Weber syndrome"- A Case report

A Halim[1], MT Alam[2], RC Barman[3]

Abstract :

Sturge-weber syndrome is a disease characterized by capillary or cavernous haemangioma (Port-wine stain) along the cutaneous division of Trigeminal nerve. There is venous haemangioma in subjacent leptomeninges, which may spread causing atrophy of cortex. The patient Md. Zobair Hossain, 11 years old boy, nondiabetic, nonhypertensive presented to us on 20.05.2011 with the complaints of repeated bleeding from a swelling over the outer aspect of right eye ball for 1 month, weakness of left half of body for 1 year and repeated convulsion for 7 years. Diagnosis was confirmed by CT scan of brain. Though treatment is unsatisfactory, he was advised for laser therapy for cutaneous lesion and anti-convulsant drug for epilepsy.

Key words: Sturge-weber syndrome, cuntaneous haemangioma, left sided hemiplegia, epilepsy, intracranial calcification.

Introduction :

Sturge-weber syndrome is a capillary haemangioma along the cutaneous distribution of ophthalmic division of trigeminal nerve. There is venous haemangiorma in subjacent lepto-meninges which causes atrophy of cortex[1]. It may be sporadic or inherited as autosomal dominant disorder. Underlying brain damage is a cause of infantile hemiplegia, mental retardation and epilepsy. Lesion is on the face in dermatological distribution. There is formation of angiomatous nodule.

Case Report:

Master Zubair Hossain, 11 years old boy presented to us with the complaints of repeated bleeding from right facial nodule for 1 month, weakness of left half of body for 1 year and repeated convulsion for 7 years. On examination there is reddish discolouration of right side of face along the cutaneous distribution of ophthalmic division of trigeminal nerve with a dark nodule formation in outer part of right orbit which bleed on touch (Figure 1). Neurological examination reveals that wasting of left upper and lower limbs, muscle power 4/5, reflexes exaggerated with extensor plantar response, sensory function intact. Right half of body is normal. Examination of eye including fundus reveals no abnormality. Investigations show; X-ray skull - Tramline calcification, CT scan of brain - large mass of calcification in right fontal, temporal and parietal lobes with small areas of enhancement around calcification.

Figure 1: Capillary haemangioma in cutaneous nerve distribution.

1. Dr. Abdul Halim, MBBS, FCPS (Medicine), Assistant Professor, Department of Medicine, Noakhali Medical College, Noakhali.

2. Dr. Md. Towhid Alam, MBBS, FCPS (Medicine), Associate Professor, Department of Medicine, Faridpur Medical College, Faridpur.

3. Dr. R C Barman, MBBS, DTCD, Assistant Professor, Department of Respiratory Medicine, Faridpur Medical College, Faridpur.

Address of correspondence :
Dr. Abdul Halim, MBBS, FCPS (Medicine), Assistant Professor, Department of Medicine, Noakhali Medical College, Noakhali.
Email:ahalimcmc24@gmail.com Mobile:+88-01712109620

Discussion:

This syndrome is a sporadic disorder and consists of a constellation of symptoms and signs including a facial nevus (Port-wine stain), seizures, hemiparesis. Stroke like episodes, intracranial calcification, and in many cases mental retardation. It occurs sporadically with a frequency of approximately 1/50,000 live birth[2].

The condition is thought to result from anomalous development of the primordial vascular bed in the early stages of cerebral vascularization. At this stage, the blood supply to the brain, meninges and face undergoes reorganization, while the primitive ectoderm in the region differentiates into the skin of the upper face and the occipital lobe of the cerebrum.

The facial nevus is present at birth, tends to be unilateral and always involve the upper face and eyelid. Seizures develop in most patients in 1st year of life. They are typically focal, tonic-conic of contralateral to the side of facial nevus. The seizures may become refractory to anti-convalsants and are associated with a slowly progressive hemiperesis. Neurodevelopment appears to be normal in first year of life. Mental retardation occurs in 50% in later childhood. Management of Sturge-Weber syndrome is multi-faceted and is aimed at seizure control, and identification and management of behavioral and learning problems[3]. For patients with well-controled seizur and normal or near normal development, treatment consists of anti-convulsants, survillance of complications including glaucoma, buphthalmos and behavioural abnormalities. Flash-Lamp-pulsed laser therapy often provides excellent clearing of the port-wine stain, particularly if located on the forehead.

If the seizures are refractory to anti-convulsant therapy, especially in infancy to Fast, 1-2 years and arise from primarily one hemisphere, most centres advise a hemi-spherectomy[4]. Because of risk of glaucoma, regular measurement of intra-ocular pressure with a tonometer is indicated. In this patient he responded well to anti-conuulsant drug. He has some learning problem as he developed left sided hemiplegia. For bleeding site we advised for laser therapy.

Conclusion:

Sturge-Weber syndrome is a capillary haemangioma along the cutaneous distribution of ophthalmic division of trigeminal nerve. There is venous haemangioma in subjacent lepto-meninges which causes atrophy of cortex. Seizures develop in most patients in 1st year of life and are associated a slowly progressive hemiperasis. Mental retardation occurs in 50% in later childhood. Patients need survillance for detection of complications including glaucoma, buphthalmos, behavioural abnormalities.

References :

1. Abdullah ABM. Short cases in clinical medicine. 4th edition, New Delhi: Elsevier; 2010.p.365.

2. Sahin M. Neurocutaneous syndromes. In: Kliegman RM, Stantous B F, Schor NF, Behrman RE, Editors. Nelson text book of paediatrics. 19th edition. Churchill Livingstone, Edinburgh: Elsevier, saunders; 2011.p.2051.

3. Youmeu. Neurological Surgery, 5th edition. Oxford, England: Butter-worth-Heinemann;1996.p.813.

4. Wilkins RH. Neurosurgery. 4th edition. Churchill Livingstone, Edinburgh: Elsevier; 2000.p.2038.