

Case Report

PREGNANCY WITH STURGE WEBER SYNDROME PRESENTING AS STATUS EPILEPTICUS

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ABSTRACT

Sturge Weber Syndrome (SWS) is associated with venous malformation that may affect eye (infantile onset glaucoma), skin (portwine facial stains/PWS) & CNS (mental retardation, neurological symptoms and convulsions). The incomplete monosymptomatic Sturge Weber Syndrome may present only with CNS symptoms. Very few cases of SWS associated with pregnancy (Dolcart and Bhat 1995) have been reported in literature. The effect of pregnancy on the neurological symptoms of SWS is not known. We report a case of Sturge Weber Syndrome with pregnancy, presenting as status epilepticus, a life threatening condition. Adverse effect of the disease on pregnancy and the need for follow up of these patients is discussed herewith.

Key Words: *Sturge Weber Syndrome, Seizures, Pregnancy*

CASES

This patient G2 A1 named PA was brought to casualty on 24.2.2012 with history of amenorrhoea 6 months having continuous tonic & clonic convulsions, frothing from mouth and respiratory compromise. Her pulse was 110/min., B.P. was 110/80mmHg. Bilateral crepts were present. Uterus size was about 18wks. PV exam. revealed fetus lying in vagina. She was given IV 5 mgm. Midazolam to control fits & intubated and shifted to ICU. She delivered a stillborn fetus in casualty. H/O seizures since the age of 10-12 years was revealed by the relatives. There was no H/O taking antiepileptic drugs. She was put on IV phenytoin (12 ampoules in 100ml normal saline over 30 mins.), IV mannitol. Her seizures were controlled after adding thiopental. She was off ventilator on 29.2.2012. Tab. phenytoin 200 mgm. HS, tab. Carbamazepine 200mgm TID and antibiotics were given to the patient. Investigations - CBC, LFT, KFT, S. Electrolytes reports were normal. CT Scan of head revealed hyperdense parenchymal & cortical gyriform calcification at left frontal and left basal ganglion region and dystrophic calcification suggestive of Sturge Weber Syndrome. No facial PWS was seen. Patient was discharged on 10th day with advice to continue antiepileptic drugs and follow up in Neurology OPD & Eye OPD every 6 months.

DISCUSSION

The incidence of SWS is estimated as 1 per 50,000. SWS is also called encephalotrigeminal angiomatosis. The angiomas involve the leptomeninges and skin of the face typically in the ophthalmic (V1) and maxillary (V2) distributions of the trigeminal nerve. The cutaneous angioma is called a portwine stain (Taddeucci *et al.*, 2005). The incidence of SWS is 3% in patients having facial PWS. Risk of SWS is more if PWS is in 1st. branch of trigeminal nerve distribution area (Hennedique *et al.*, 2008). SWS is caused by embryonal blood vessels and their secondary effects on surrounding brain tissue. A vascular plexus develops in the sixth week around the cephalic part of the neural tube under ectoderm destined to become facial skin. Normally this vascular plexus regresses around 9th week of gestation. Failure of this regression results in residual vascular tissue, which forms the angiomata of the leptomeninges, face & ipsilateral eye. SWS is termed as complete when both CNS and facial angiomas are present and incomplete when only one area is affected. Roach scale is used for classification as follows (Roach 1992)

Type I - Both facial and leptomeningeal angioma present, may have glaucoma.

Type II - Facial angioma only, No CNS involvement; may have glaucoma.

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Type III – Only leptomeningeal angioma (CNS involvement); usually no glaucoma (Aydin *et al.*, 2008)

Our case belonged to Type III, as she presented only with seizures. Only 24 cases of type III have been reported presenting as migraine like symptoms (Taddeucci *et al.*, 2005). Neurological manifestations of SWS syndrome Seizures (75-90%), maybe refractory. Onset of seizures range from birth to 23 years age. 75% have onset before 1 year. Seizures result from cortical irritability caused by cerebral angioma, through mechanisms of hypoxia, ischemia and gliosis. Episodes of status epilepticus are dangerous in an already compromised vascular system. Stroke like episodes, headache (in 77%) may occur. Ocular manifestations include glaucoma (48%). Out of these 61% had onset of glaucoma before 24 months of age. Decreased vision and blindness can occur. Skin manifestation Facial portwine stains (PWS). Not all people with a PWS have SWS. The incidence of SWS is 8-33% in those patients having PWS. The incidence of SWS is high if PWS involves V1 distribution of trigeminal nerve (Takeoka 2010). Neuroimaging Maximum extent of disease may require a combination of structural and functional neuroimaging, since mismatch may exist among neuroimaging modalities. Each modality may demonstrate abnormalities not detected by the other. This is especially important in the identification of the epileptogenic region when considering surgery for refractory seizures.

CT Scan

Shows typical gyriform calcifications or tram-track calcifications (Figure 1). The gyriform calcification in CT and plain film is due to altered vessel wall permeability causing leakage of calcium phosphates or calcium carbonates with subsequent secondary crystallization within perivascular parenchyma. Other findings include brain atrophy, ipsilateral choroid plexus enlargement & abnormal draining veins. CT Scan is more sensitive than MRI for detecting cortical calcifications.

MRI

Although MRI does not show calcifications, gadolinium enhancement may show pial angioma; therefore MRI may permit early diagnosis of SWS.

PET

Offers functional (metabolic activity) data to determine the full extent of involvement of brain parenchyma. Used for presurgical evaluation.

F-18 FDG (Fluoro deoxyglucose)

Measures F-18FDG metabolism in brain tissue to determine its metabolic function.

SPECT

Hyperperfusion in early stages and hypoperfusion in late stages.

Management Medical

Antiepileptic drug should be used for seizures, for glaucoma medications which decrease production of aqueous fluid (beta antagonist eye drops) or which promote outflow of aqueous fluid (adrenergic eye drops, miotic eye drops) can be used. Portwine stains can be treated by laser therapy.

Surgery

It is indicated when functional neurological deficits occur, intractable seizures or evidence of progressive cortical damage is noted. Focal cortical resection, hemispherectomy, corpus callosotomy, vagal nerve stimulation can be done depending on extent and location of lesion. Glaucoma surgery may be beneficial if medications are unable to control intraocular pressure. Trabeculectomy, goniotomy etc. can be considered. For portwine stains pulsed laser therapy can be done.

Prognosis

In patients with seizures, over half of cases develop severe learning disabilities in later childhood with need for special education. Factors suggesting a progressive course of disease are –Increased seizure

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frequency and duration, increased focal or diffuse atrophy or increase calcifications in neuroimaging, development of hemiparesis and deterioration of cognitive functions. Glaucoma can cause blindness. Diagnosis is difficult in type III SWS because of no cutaneous lesions (PWS). In our case there was no

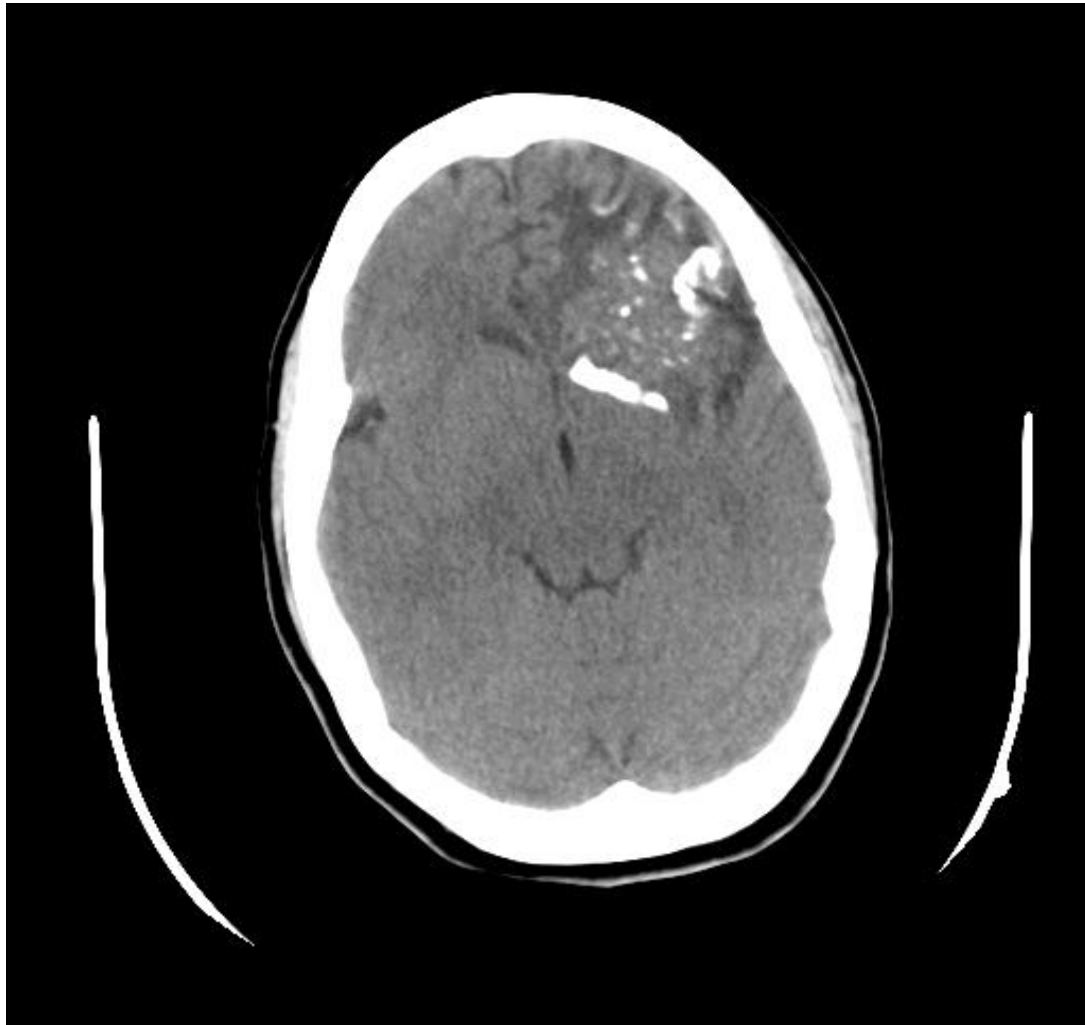


Figure 1: CT Scan showing gyriform calcification

neurological deficit also, hence the diagnosis could only be made after CT scan. Cerebral venous malformations are considered relatively benign during pregnancy and they infrequently bleed during labor or in postpartum period. Cerebral arteriovenous and arterial haemangiomas are more dangerous and may rupture with blood pressure fluctuations and valsalva efforts during labor. In SWS the leptomeningeal angiomas are predominantly venous and deep arterial malformations are uncommon. Whether the hormonal or haemodynamic changes associated with pregnancy were responsible for the 'status epilepticus' condition as in our case at admission can only be speculated. A case of severe rt.sided hemiplegia with hemianopia and aphasia followed by seizures in 3rd trimester of pregnancy has been reported in which recovery occurred gradually in postnatal period (Chabriat *et al.*, 1996). The diagnosis of (SWS) should be kept in mind in patients presenting with seizures if it is associated with facial PWS and even with no facial PWS. When a newborn has a facial PWS reaching V1 ,ophthalmologic

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examination must be performed in the 1st month of life and neuroimaging at 6-12 months of life or earlier if neurologic signs are seen. These patients need close follow up of progression of their neurological disease. Regular fundus examination and intraocular pressure monitoring should be done and treated timely to avoid optic atrophy and blindness.

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