Sinus Headache And Migraine Overlap In A Case Of A Sturge-Weber Syndrome

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Citation

Abstract
Sturge Weber Syndrome (SWS) is a congenital syndrome of unknown cause and is characterized by a triad of port-wine stain, glaucoma, and central nervous system involvement. Migraine is a known feature of SWS. Sinus headache and migraine symptoms can overlap resulting in difficulty in diagnosis and treatment. A case of SWS having overlap of migraine and sinus headache is presented and diagnostic and treatment modalities are discussed.

INTRODUCTION
Sturge-Weber Syndrome (SWS) is a neuro-cutaneous disorder that classically presents with a facial port-wine stain, in the ophthalmic division (V1) of the trigeminal nerve, associated with an ipsilateral lepto-meningeal angioma and glaucoma. Patients often develop progressive neurological problems including difficult to control seizures, migraine, stroke-like episodes, mental retardation and hemiparesis.

Deviated nasal septum and chronic sinusitis can be associated with headache. Migraine on other hand can have associated nasal symptoms. This overlap of nasal complaints can result in difficulty in diagnosis and treatment. The problem is further compounded if the patient carries a high surgical risk. We report a patient suffering from SWS with sinus headache scheduled for surgery.

CASE REPORT
16-year-old female patient, known case of SWS, with port-wine stain and glaucoma presented with a history of recurrent headache, localized over the fore-head for two years, associated with nausea. There was no history of vomiting, photophobia, epilepsy or loss of vision. The patient also gave history of nasal blockage, post nasal drip and frequent need to clear the throat. The patient was treated for migraine with some relief, and was referred to otorhinolaryngology department for her nasal symptoms.

Her general physical examination revealed red-pinkish discoloration of left side of the face involving forehead, cheek and upper lip in the region of V1, V2, V3 divisions of trigeminal nerve. Neurological examination was normal. Otorhinolaryngological examination revealed DNS to right. Ear and throat examination was normal.
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Figure 1
Figure 1: Clinical photograph of the patient showing port-wine stain involving the left half of the face in V1, V2, V3 distribution of the trigeminal nerve.

Her haematological, biochemical and coagulation profile was normal. CT scan head and PNS revealed DNS to right and mucosal hypertrophy of the left maxillary and both ethmoid sinuses. Osteo-meatal complex were blocked on both sides (Fig 2).

The patient was diagnosed as a case of SWS with complete trisymptomatic syndrome with chronic sinusitis and sinus headache and surgery was planned. Her pre-anesthetic evaluation was unremarkable and she was taken up for surgery under local anesthesia. Septoplasty, bilateral middle meatus antrostomy and anterior ethmoidectomy was

Figure 2
Figure 2: CT scan PNS showing the mucosal hypertrophy of maxillary and ethmoidal sinuses, DNS to right side and mucosal hypertrophy of left maxillary sinus.

Calcification was seen on the left parieto-occipital region with enhancement and prominence of choroid plexus in ipsilateral lateral ventricle (Fig 3).

Figure 3
Figure 3: CT scan Head showing calcification in the left parieto-occipital region with enhancement and prominence of choroid plexus in ipsilateral lateral ventricle.
performed. She had an uneventful recovery. Her headache improved both in intensity and frequency. She is on regular follow-up for CNS lesion and glaucoma.

DISCUSSION

The exact cause of Sturge Weber Syndrome is unknown, however it is postulated that failure of regression of embryonic blood vessels results in residual vascular tissue, which forms the angioma of the lepto-meninges, face and ipsilateral eye.

The venous dilations on the face, also called as port wine stain, are the hallmark of the disease. It is present in about 96% of patients and is visible at birth. SWS occur only when the port wine stain involves the area of V1 and V2 branches of the trigeminal nerve distribution, as was also noted in our patient.

The incidence of glaucoma ranges from 30-71%, which can develop at birth or later at any age. Glaucoma is produced by mechanical obstruction of the angle of the eye, elevated episcleral venous pressure or hyper secretion of fluid by either the choroid angioma or ciliary body. The similar mechanism is responsible for buphthalmos. Decreased vision and blindness can result from untreated glaucoma as the increased intraocular pressure can lead to optic nerve damage.

The incidence of epilepsy in these patients is 75-90%. Seizures result from cortical irritability caused by cerebral angioma, through mechanisms of hypoxia, ischemia and gliosis. The onset of seizures prior to the age of two years suggest a greater chance of refractory epilepsy and mental retardation as these patients have extensive involvement of the trigeminal nerve distribution, as was also noted in our patient.

Sujanksy and Conradi reported that developmental delay occurred in 58% patients, however the rate was higher (71%) of those with seizures and less (6%) of those without seizures. Moreover patients with a later onset of seizure had a lower incidence of developmental delay. The incidence of neurological deficit also increases with age and affects 65% adults indicating the progressive nature of the disease.

CT scan head may show the characteristic double lined gyroriform pattern of calcification paralleling cerebral convolutions, also known as railroad sign. These calcifications are usually not detectable before one year of age and may not be seen for several years. Other findings include brain atrophy, ipsilateral choroids plexus enlargement and abnormal draining veins.

Headache in SWS is usually secondary to vascular disease giving symptoms of migraine headache and occurs with frequency of 28% to 60%. The prevalence of migraine in children younger than 10 years is 31% with SWS, much greater than 5% prevalence in the general population.

The evolution of a migraine attack begins when the nervous system is confronted with a single triggering event or a synergistic consequence of multiple risk factors. If the migraine process continues and reaches a critical threshold, there is activation of the trigeminovascular system. Activated trigeminal afferents release vasoactive peptides; that mediate vasodilatation and neurogenic inflammation of cranial vessels.

Sinus headache is considered secondary to inflammatory pathologic conditions. Since there is overlap of the symptoms it may be difficult to diagnose the two conditions. If the dominant symptom is an episodic headache or facial pain over the sinus cavities, nasal symptoms are in the midst of the migraine-associated symptoms such as nausea and photophobia, migraine is likely. The presence of the prodrome or aura is also highly suggestive of migraine. If, however the patient complaint is sinus pressure and congestion, but there is no escalation to moderate to severe headache, and there is nasal obstruction and purulent discharge, rhino-sinusitis should be the primary consideration.

A CT scan PNS is helpful to diagnose the extent of the disease but not the severity of headache. Shields reported a lack of correlation between the degree of sinus-area pain and the presence and severity of CT-observed pathologic conditions in the sinuses, thus supporting clinical symptoms rather than the imaging for establishing the diagnosis of rhino-sinusitis.

Levine reported that, if headaches were associated with other diagnostic criteria for rhino-sinusitis such as facial pressure or congestion, a positive outcome in terms of headache relief occurred in approximately 75% of the cases reviewed. If headaches were the only indication for surgery, however, a positive outcome was noted in less than 20% cases. Hence a detailed history, physical and radiological examination is
helpful in evaluation of the patient. However given the high prevalence of these disorders, it is valuable to keep in mind that they often exist co-morbidly.

SWS patients can have intubation difficulties due to angiomata of lip, oral cavity, tongue, larynx and trachea and soft, non-styled, well-lubricated tracheal tubes should be used to prevent haemorrhage. Straining, bucking and obstructed airways during induction or emergence may increase intraocular pressure, blood pressure and intracranial pressure, latter two can cause intracranial haemorrhage if the patient has associated haemangiomatous lesions of brain.

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