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Case Report

Asymptomatic Sturge-Weber syndrome patient with extradural-intradural Ewing sarcoma

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ABSTRACT

Sturge Weber syndrome (SWS), also called as encephalotrigeminal angiomatosis is an uncommon congenital neurological disorder and frequent among the neurocutaneous syndromes specifically with vascular predominance. Symptoms and signs depend on the extent and location of the venous dysplasia. In its complete trisymptomatic form, SWS is physically characterized by port-wine stains (PWS) over the trigeminal area, leptomeningeal angiomas usually over the parieto-occipital region, and eye abnormalities. We report a case of 19 year old gentleman asymptomatic Roach type II SWS patient with Ewing sarcoma presenting as extradural-intradural spinal tumour.

Keywords: Sturge Weber syndrome, Ewing's sarcoma, PNET spinal tumour

INTRODUCTION

Sturge-Weber syndrome (SWS), or encephalotrigeminal angiomatosis, is a rare, sporadic disorder that occurs with a frequency of approximately 1/50,000 births.[1] The syndrome is characterized by a facial PWS in the ophthalmic distribution of the trigeminal nerve.[2] An intracranial vascular anomaly (leptomeningeal angiomatosis [LA]) involving the occipital and posterior parietal lobes may affect both cerebral hemispheres. The result is hemiparesis, hemianopsia, seizures, mental retardation, and glaucoma.[3] Schirmer was first one to describe it nearly a century ago (1860) in association with facial angioma and buphthalmos. Later in 1879 while reporting a case of 6½ year old girl, William Allen Sturge acclaimed these features in accordance with neurological findings and termed it as SWS.[4] There is a risk of 10e50 % for the involvement of brain if a child is born with Port-wine birthmark (PWB) on the forehead or the upper eyelid. When the upper and lower eyelids are involved in port-wine birthmark, the risk of developing glaucoma becomes as high as 50%.[5] The frequency of occurrence of SWS is between 1:20,000 and 1:50,000. Amongst these port-wine birthmark is the most common with the occurrence of 0.3% in live births.[4] In 1992, Roach categorized SWS variants into three types: Type I: individual has a facial PWS, leptomeningeal angioma, and may have glaucoma. Type II: individual has a facial PWS, no leptomeningeal angioma, and may have glaucoma and type III: individual has LA, no facial PWS, and, rarely, glaucoma.[6]

The aim of this article is to report a case of asymptomatic SWS with Extradural-intradural lumbar Ewings sarcoma and add to the existing literature.

CASE REPORT

A 19 year old gentleman came to neurosurgery OPD of Subharti Medical college, Dehradun with complaints of Low back ache radiating to right leg×5 months mild to moderate in severity. Progressively increasing insidious onset swelling in the right flank×5 months, painful on touching. Weakness of right leg started with slipping of chappals from right foot without awareness with knee bending to difficulty in weight bearing on right leg, followed by weakness in left leg 2 months back and to cause difficulty in weight bearing and walking, making

patient bed ridden. Reduced sensations present over right leg started 2 months back involving right foot and leg and progressively thigh and left leg below knee within 1 month period. Incontinence of urine present since 1 month.

On examination the E4V5M6, Pupil B/L 2 mm reacting, all cranial nerves intact B/L. Port Wine stain present on the left side of face involving ophthalmic and maxillary division of trigeminal nerve (Figure 1). Rest of the general examination was normal. Table 1 showed neurological findings in the patient.

Table 1: Neurological findings.

Variables		Right	Left
Tone	Upper limbs	Normal	Normal
	Lower limbs	Hypotonia	Hypotonia
Power	Hip	2/5	4/5
	Knee	2/5	4/5
	Ankle	1/5	2/5
	Upper limb	5/5	5/5
Sensation	Reduced	L3 to S1	L5 to S1
Deep tendon reflex	Biceps	Normal	Normal
	Triceps	Normal	Normal
	Knee	Absent	Absent
	Ankle	Absent	Absent
	Babinski	Flexion	Flexion

CECT L-S spine (Figure 2) was suggestive of two lobulated heterogeneously lobulated T2 hyperintense lesion with cystic and haemorrhagic component along right lateral intraspinal extramedullary location extending transforaminally to involve right Psoas muscle and second lesion along right paraspinal muscle. Patient was taken up for surgery and L2 to L4 complete laminectomy with tumour decompression done under general anesthesia. Intraoperatively tumour was soft, grayish, highly vascular, extradural with intradural extension, extending to left paraspinal muscles.

Histopathology (Figure 3) was suggestive of malignant small round cell tumour, consistent with Ewing's sarcoma.

MRI brain (Figure 4) was suggestive of tram-track gyral cortical calcification in fronto-temporal region and in right parietal region. Calvarium and frontal sinus enlargement with ipsilateral choroid plexus enlargement. Multiple prominent vascular channels in the medullary white matter representing collateral pathways for the venous drainage which drain into enlarged sub-ependymal veins into the prominence venous channel along the pial surface.

PET-CT in Figure 5 showing tram-track calcification.

On fundus examination the right vasculitis with ischemic retinopathy.

Post-operatively patient was started on VEC regimen of chemotherapy and kept under follow up for further management.

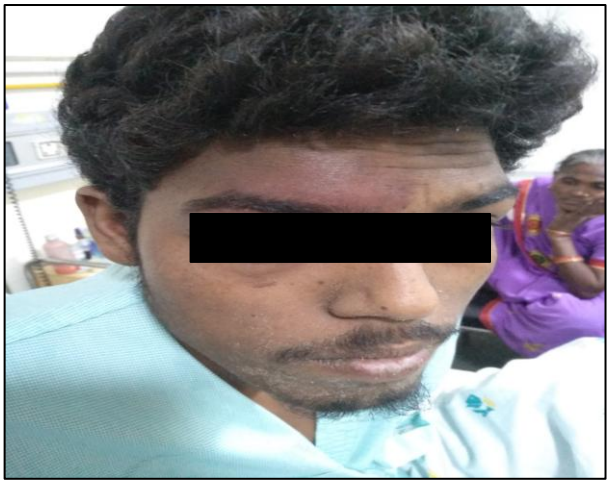

Figure 1: Port Wine stain present on the left side of face.

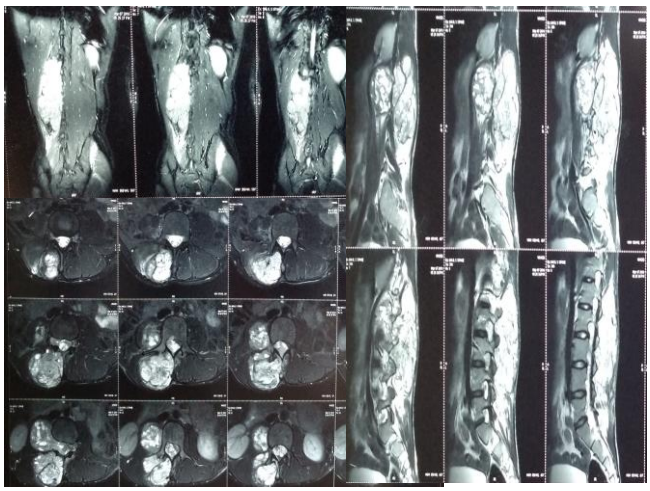

Figure 2: CECT L-S spine showing large extradural tumor with intradural extension.

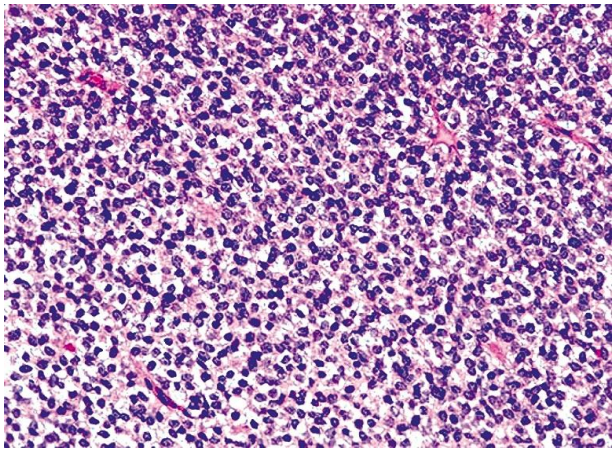

Figure 3: Malignant small round cell tumour.

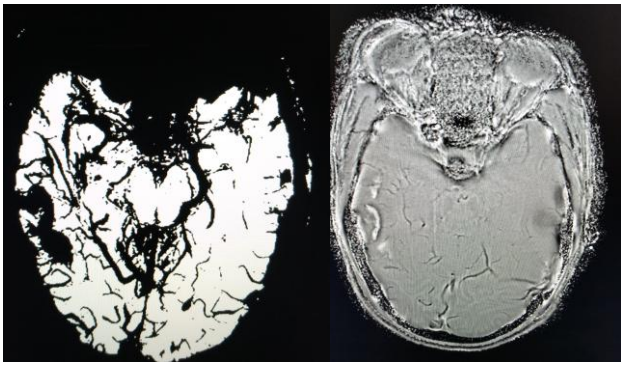

Figure 4: SWI of tram-track calcification in fronto-temporal region.

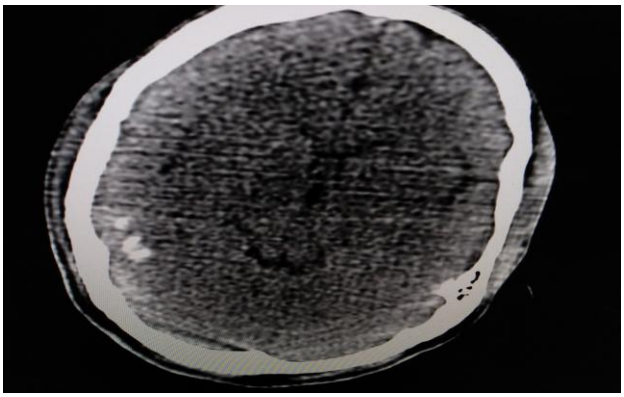

Figure 5: Tram-track calcification.

DISCUSSION

Bioxeda et al in 1993 studied 121 patients with PWSs and found 88% of stains were predominantly distributed over maxillary branch of trigeminal nerve.[7] In 1999, Inan and Marcus reported the presence of port-wine nevus up to 90% on right side of face.[8] The 50% patients showed lesion extension over midline and 33% showed bilateral involvement of the lesion. SWS is a developmental anomaly of embryonic origin because of malformation in mesodermal and ectodermal development. It results due to failure of regression of a vascular plexus around cephalic portion of neural tube which is destined to become facial skin. This vascular plexus normally forms at 6th week of intrauterine life and regresses by 9th week. Failure of its regression results in residual vascular tissue which forms angiomas of leptomeninges, face and ipsilateral eye.[9] Due to this there is abnormal flow of blood which in turns causes vasomotor phenomenon, resulting in ischemia, gliosis, atrophy, and calcification of the cortical tissues.

Imaging

Conventional skull radiographs are usually advised for visualizing the calcifications in frontal lobe OPG showed osteohypertrophy of the involved side.[4] Lateral cephalic and PA view skull failed to show any calcifications in the frontal lobe. The recommended imaging modality to diagnose SWS is magnetic resonance imaging (MRI).[4] Magnetic resonance imaging of the brain was performed and high-resolution T1eT2 weighed serial sections obtained in the sagittal and axial planes. Contiguous fast FLAIR images were also obtained in the coronal plane on 1.5 T scanner using a dedicated 8-phased-array surface coil. Additional diffusion weighted images were obtained in the axial plane. T1-weighted serial sections were also obtained in the sagittal, axial and coronal planes after injection of intravenous contrast. In present case SWI MRI showed tram track calcification characteristic of SWS.

Ocular examination

The choroidal hemangioma causes increased secretion of aqueous humor which in turns causes increased intra ocular tension and causes buphthalmos and glaucoma.[10] Glaucoma, exudative retinal detachment (ERD) and heterochromia iridis may be present in 30-70% as ophthalmic manifestation.[11] In the present case the authors report the right vasculitis with ischemic retinopathy under ophthalmoscopy.

Management

There is enough evidence-based studies suggesting laser treatment for PWSs in early infancy. Anticonvulsant medications in combination with aspirin can be given for management of neurologic consequences. Surgical interventions are often needed in glaucoma as in association with SWS this doesn't respond to medical therapy. Photodynamic therapy and intra-vitreous bevacizumab is an upcoming treatment modality for diffuse choroidal hemangioma.[11] Correction of gingival enlargement can be done with gingivectomy and lasers as they have additional advantage of homeostasis and minimum damage to oral tissues.[13] In the present case treatment plan which was carried out consisted of treatment of extradural tumour extending to intradural, which was debulked surgically and histopathology confirming Ewings sarcoma, post-operatively was started on chemotherapy regimen.

CONCLUSION

SWS presents with varied clinical features. There is no case reported upto date showing the occurrence of extra-osseous Ewings sarcoma with SWS. Available latest treatment modalities and thorough knowledge of this syndrome is very important in management of this condition. It is a multidisciplinary team work to deliver proper medical care to the patient. Complete understanding regarding the entire disorder should be imparted to caregivers. It is important that caregivers should perceive consequences of complications so that the patients are treated quickly and effectively. We have reported the case of asymptomatic SWS presenting with Ewings sarcoma of extraosseous site, (Spine) in this case.

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