Anaesthetic management of a case of Sturge-Webers syndrome

INTRODUCTION

Encephalotrigeminal angiomatosis is commonly referred to as the Sturge-Weber (SWS) after Sturge and Weber, who first described this problem in 1879. The main clinical features of this syndrome are port wine stains, glaucoma and convulsions. The incidence is common in both sexes. These patients can have difficulty with intubation due to angiomas of the mouth and upper airway. Anaesthesia should be planned to avoid trauma to the haemangioma and increase in intraocular and intracranial pressure.

CASE REPORT

A 21-year-old male, a known case of SWS, was posted for open reduction and internal fixation of fracture left ankle. He was 165 cm tall and weighed 78 kg. He had port wine stain on the left side of the face, left eyelid and left half of the palate. He had left hemiparesis. His mental status was normal. On examination, he was found to have grade 5/5 power on the right side and 3/5 on the left side. His mouth opening was adequate and the modified Mallampati classification was 2. All routine tests were unremarkable. He had undergone surgical correction for glaucoma of the left eye, and his intraocular tension was normal. Computed tomography (CT) scan brain showed atrophy and calcification of the left cerebral hemisphere. General anaesthesia was planned. Written informed consent was obtained and adequate duration of fasting was confirmed. The patient was premedicated with midazolam 1 mg intravenously (IV) to allay anxiety. Fentanyl 100 μg IV was given for pain relief. The patient was induced with Propofol 2 mg/kg. The trachea was

intubated with an 8 mm internal diameter portex tube facilitated by 0.1 mg/kg vecuronium bromide. Patient's modified Cormack Lehane score was 3 and no vascular malformations were noted in the airway. Anaesthesia was maintained with N_2O:O_2 ratio of 60:40 and 1–2% sevoflurane, with intermittent doses of vecuronium. Intraoperative course was uneventful. After the surgery, once spontaneous breathing returned, anaesthesia was reversed with neostigmine 0.05 mg/kg and glycopyrrolate 5 mcg/kg. Extubation was performed after confirming return of adequate motor power. He was observed in the post anaesthesia care unit for 2 h before being transferred to the ward. The post-operative course was uneventful.

DISCUSSION

SWS, also called encephalotrigeminal angiomatosis, is a neurocutaneous disorder with angiomas involving 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 port wine stain.[1,2]

SWS is caused by residual embryonal blood vessels and their secondary effects on the surrounding brain tissue. In the sixth week of gestation, a vascular plexus develops around the cephalic portion of the neural tube, under ectoderm destined to become facial skin. Normally, this vascular plexus regresses around the ninth week of gestation. Failure of this normal regression results in residual vascular tissue, which forms the angiomata of the leptomeninges, face and ipsilateral eye. Stasis results in ischemia underlying the leptomeningeal angiomatosis, leading to calcification and cortical necrosis. A "vascular steal phenomenon" may also develop around the angioma, resulting in cortical ischaemia.

The Roach scale is used for classification of SWS, which is as follows[3]:

- Type I – both facial and leptomeningeal angiomas; may have glaucoma
- Type II – facial angioma alone (no central nervous system involvement); may have glaucoma
- Type III – isolated leptomeningeal angioma; usually no glaucoma

The incidence of epilepsy in patients with SWS is 72–93%.[2] Episodes of status epilepticus are dangerous in SWS. In an already compromised vascular system, such seizures are more likely to cause injury. Patients may exhibit intracranial calcification on skull X-ray, which has the characteristic pattern of a parallel serpentine configuration known as the "tram sign" or "railroad sign"[4] [Figure 1]. CT scan may show calcifications in infants and even neonates. Although magnetic resonance imaging does not show calcifications, gadolinium enhancement may show pial angioma.[2]

These patients also suffer from hemiparesis, stroke-like episodes and hemianopsia.

Patients with facial tissue hypertrophy may be at risk for obstructive sleep apnoea, and should be appropriately evaluated.[5] Anne *et al.*[6] reported 2.4% of children with SWS to develop hypothyroidism. These patients are treated with anticonvulsants for seizure control. Seizures refractory to medical treatment are subjected to surgical correction. Surgical procedures include focal cortical resection, hemispherectomy and corpus callosotomy. A recent mode of non-surgical treatment for intractable convulsion is the vagal nerve stimulation.[7] Patients with stroke-like episodes are treated with aspirin. Recurrent thrombotic episodes producing gradual loss of brain function may require use of antiplatelet agents and low-molecular weight heparin. Children with SWS may be submitted to periodic examination of the eyes. The treatment of glaucoma is directed to prevent optic nerve atrophy and resultant blindness. Port wine stains are treated with pulsed dye laser therapy. Physiotherapy should be considered for infants and children with muscle weakness. Educational therapy is often prescribed for those with mental retardation or developmental delays.[8]

The SWS patients may present for facial and cosmetic surgeries, dental procedures, trabeculectomy, and goniotomy, examination under anaesthesia or seizure control surgery.[9] The choice of anaesthesia should be influenced by detailed clinical and radiological

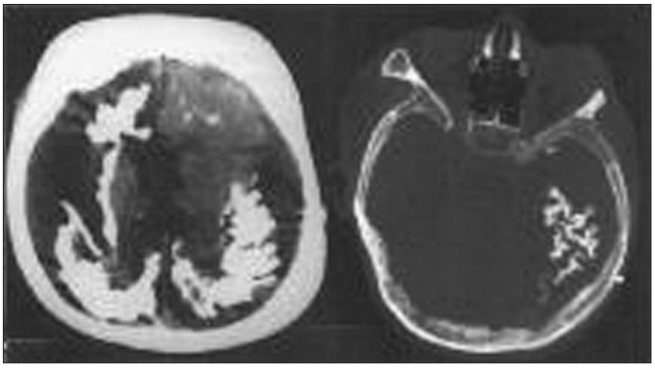

Figure 1: Rail road sign in computed tomography scan

investigation. General anaesthesia may necessitate avoidance of drugs that can cause an increase in the intraocular pressure, such as succinylcholine and ketamine. Anticholinergics should be avoided in patients with narrow angle glaucoma. During anaesthetic induction, measures to avoid the haemodynamic response to laryngoscopy should be adopted. Angiomas may involve the airway, leading to difficult mask ventilation, laryngoscopy and intubation. Extreme delicacy in airway manipulation is warranted as it may result in uncontrolled vascular haemorrhage resulting from ripping of vascular lesions. Pre-operative antiepileptic therapeutic drug level may be performed to guide the therapy. Neuraxial block is associated with the possibility of raised intracranial pressure and precipitation of cerebral coning. Regional anaesthesia should be avoided in patients on antiplatelet drugs and heparin.

Although patients with SWS tolerate anaesthesia well, anaesthetic management includes careful assessment for associated anomalies.

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REFERENCES

1. Gupta RK, Singh A. Sturge Weber Disease in a child. JK Sci 2006;8:109-110.
2. Takeoka M. Pediatric Sturge Weber syndrome. Available from: http://emedicine.medscape.com/article/1177523-overview. Last updated January 5, 2010. [Last accessed on 2011 Dec 2].
3. Roach ES. Neurocutaneous syndromes. Pediatr Clin North Am 1992;39:591-620.
4. Gandhi M, Iyer H, Sehmbi H, Datir K. Anaesthetic Management of A Patient with Sturge-Weber Syndrome Undergoing Oophorectomy. Indian J Anaesth 2009;53:64-7.
5. Irving ND, Lim JH, Cohen B, Ferenc LM, Comi AM. Sturge-Weber syndrome:Ear, Nose and Throat issues and neurological status. Pediatric Neurology 2010;43:241-4.
6. Comi AM, Bellamkonda S, Ferenc LM, Cohen BA, Germain-Lee EL. Central hypothyroidism and Sturge-Weber syndrome. Pediatr Neurol 2008;39:58-62.
7. Saneto RP, Sotero de Menezes MA, Ojemann JG, Bournival BD, Murphy PJ, Cook WB, *et al*. Vagus nerve stimulation for intractable seizures in children. Pediatr Neurol 2006;35:323-6.
8. Takeoka M, Rivello JJ Jr. Sturge Weber Syndrome. 2006. Available from: http://www.emedicine.com/neuro/topic356.htm. [Last accessed on 2011 Dec 2].
9. Ceyhan A, Cakan T, Başar H, Bababalim M, Unal N. Anesthesia for Sturge Weber Syndrome. Euro J Anesth 1999;16;339-41.