

ANESTHESIA FOR ENCEPHALO-TRIGEMINAL ANGIOMATOSIS (STURGE-WEBER SYNDROME)

MOHAMAD BILAL DELVI^{*} AND
MOHAMMAD SAID MAANI TAKROURI^{**}

Abstract

A case report of rare congenital disease of encephalo-trigeminal angiomas (Sturge-Weber Syndrome) (SWS), is presented to alert the anesthesiologist of its potential risk. The case was anesthetized for emergency orthopedic surgery. Anesthesia management is directed towards observing the clinical manifestations of localized superficial skin lesions, extensive systemic involvement, evaluating its associated anomalies and anticipating difficult intubation due to angiomas of the mouth and upper airway and be a ware of concurrent therapy.

The authors reviewed the English-language literature and reported findings regarding the SWS pathophysiologic features, interventions, and outcomes, with emphasis on issues relevant to anesthesiologists. This patient tolerated balanced anesthesia well. Anesthesia should be planned to avoid trauma to the hemangiomas, increases in intraocular and intracranial pressure, and be cognizant of current anticonvulsant therapy.

* MB, Ch.B, FKSUA DABA. Consultant: Assistant Professor Anesthesiologist, King Khalid University Hospital. 11461, P.O. Box: 2925. Riyadh Saudi Arabia. Tel: 009661-4671024.

**MB, Ch.B, FRCA(I), Professor. of Anesthesiology. Department of Anesthesia, King Khalid University Hospital, Riyadh 11461. P.O. Box: 2925, Tel: 009661-4672673, E-mail: takrouri@ksu.edu.sa.

Introduction

Encephalo-trigeminal angiomatosis, known as well as Sturge-Weber Syndrome (SWS), is a rare disorder that occurs with a frequency of approximately 1 per 50,000. The disease is characterized by an intracranial vascular anomaly, leptomeningeal angiomatosis, most often involving the occipital and posterior parietal lobes. Facial cutaneous vascular malformations present as facial port-wine stain in the ophthalmic distribution of the trigeminal nerve, is characteristic. An ipsilateral leptomeningeal angioma, seizures, and glaucoma are among the most common symptoms and signs. Stasis results in ischemia underlying the leptomeningeal angiomatosis, leading to calcification and laminar cortical necrosis. The clinical course is highly variable and some children experience intractable seizures, mental retardation, and recurrent strokelike episodes¹⁻³. Children and adults with this syndrome often develop progressive neurologic problems, including difficult-to-control seizures, migraines, stroke-like episodes, mental retardation, and hemiparesis. Treatment is largely symptomatic and directed at the control of seizures with anticonvulsants or surgery^{2,3}. Only three reports of such cases were found in the anesthetic English literature⁴⁻⁶.

Case report

A 2-year male child was admitted to KKUH with a history of fall and supracondylar fracture. The patient was posted for emergency open reduction and internal fixation of the fracture with K-wire under general anesthesia.

The history elicited from the mother revealed that this patient was admitted to the hospital on few occasions earlier with a history of convulsions and hemiparesis affecting the right side of his body and was treated with anticonvulsants since the last admission. The patient had undergone CT of the brain earlier and the result was normal. He was diagnosed as a case of Sturge Weber Syndrome.

On examination the child was calm and co-operative and had a facial nevus (port wine) on his left eyebrow and forehead (Fig. 1). The other systemic examination was unremarkable.

Fig 1

The child under anesthesia. The facial port wine color of the angiomas is shown on the distribution of ophthalmic branch of the trigeminal nerve



In preparation for surgery, monitoring included ECG, pulse oximeter, capnograph (Datex) NIBP (Dinamap Critikon). Induction consisted of preoxygenation followed by 5 mg/kg thiopentone sodium and 1 mg/kg suxamethonium to achieve a rapid sequential intubation. The trachea was intubated with a 4.5 mm ETT. Anesthesia was maintained with oxygen, nitrous oxide with fentanyl and atracurium. Intraoperative course was smooth and the trachea was extubated after reversing the muscle relaxant with neostigmine and atropine.

The patient was observed overnight and had no complications. Patient was discharged two days later.

Discussion

From the few anesthetic papers reviewed dealing with such case reports, stress is made on airway management in the presence of obstructing angiomas or trauma, situations which may lead to excessive bleeding⁴⁻⁶. A careful approach to the patient induction, the presence of anticonvulsant therapy, and possibility of convulsions in the perioperative

period is also recommended.

The SW syndrome constitute persistence of primordial vascular plexus and primary abnormality in development of ectoderm to form the skin of upper part of face, overlies that part of neural tube destined to form occipital and adjacent parts of the cerebrum. The hemangiomas represent malformations of blood vessels⁷⁻⁹ and the basic lesion involves ipsilateral angiomas of skin, meninges and choroids⁹⁻¹².

The characteristic clinical features are cutaneous manifestations facial nevus (Port Wine Stain). The capillary nevus is flatten, and blanches on pressure. When present, it appears in the trigeminal nerve (ophthalmic branch) distributed to superior eyelid and supraorbital region (Fig. 1) It may also involve maxillary and mandibular branches. It has been reported to cause hypertrophy of the involved areas as well as the nasopharynx. This single finding may contribute to excessive injury in the process of intubation⁴⁻⁶.

With regards to neurological manifestations, the most important are seizures in the form of contralateral partial or secondary seizures appearing before the age of one year. (this was observed in the case reported). Seizures increase in frequency and severity as the child grows and whose type, frequency and severity do not correlate with extent of cutaneous involvement. Recurring paralysis is another important manifestation requiring longer periods of recovery and the possibility of an eventual permanent paresis in one third of the patients.

Hemiparesis can occur and develop gradually contralateral by to the facial nerve. It may also be associated with hemiatrophy of limbs, cortical sensory deficits and hemianopia. Spasticity and pyramidal signs may occur. These manifestation were not observed in the child under discussion.

The reported cases may suffer from mental retardation of progressive type, but it is not clear if this is related to the frequency of seizures or due to the disease process.

The ocular manifestations of the disease consist of: fundal choroidal angiomas, iris heterochromia and congenital (bupthalmos) or acquired

glaucoma (25-50%).

The clinical picture could be confirmed by imaging studies including skull X-ray which may show intracranial calcification in 90% of cases (railroad track pattern). Recent MRI/CT imagings demonstrate leptomenigeal angiomatoses, atrophy of the cortex and calcification. In the case presented, all appeared normal.

In the management of such cases a multidisciplinary approach (pediatric, neurology, ophthalmology, orthopedic, physiotherapy) is indicated. If surgery is entertained, the anesthesiologist should be involved in order to note abnormalities of airway and presence of hemangioma. Also he may assess the degree of mental retardation. The paralysis, convulsions or anticonvulsants are checked. The case herein reported showed a recent episode of convulsions which caused the fracture and necessitated continuing the anticonvulsant therapy.

The consulting team working on the case presented did not find major incident probably due to the extreme care taken in the management and the use of inhalational based anesthesia for rapid recovery. The rationale of reporting this case is to alert the vigilant anesthesiologist to the presence of the SW syndrome and raise his awareness to the potential risks involved.

References

1. THOMAS-SOHL KA, VASLOW DF, MARIA BL: Sturge-Weber syndrome: a review. *Pediatr Neurol*; 30(5):303-10, May 2004.
2. ARZIMANOGLU AA, ANDERMANN F, AICARDI J, SAINTE-ROSE C, BEAULIEU MA, VILLEMURE JG, OLIVIER A, RASMUSSEN TH: Sturge-Weber syndrome: indications and results of surgery in 20 patients. *Neurology*; 55:1472-1479, 2000.
3. SUJANSKY E, CONRADI S: Sturge-Weber syndrome: age of onset of seizures and glaucoma and the prognosis for affected children. *J Child Neurol*; 10:49-58, 1995.
4. BATRA RK, GULAYA V, MADAN R, TRIKHA A: Anaesthesia and the Sturge-Weber syndrome. *Can J Anaesth*; 41(2):133-6, 1994.
5. CEYHAN A, CAKAN T, BASAR H, BABABALIM M, UNAL N: Anaesthesia for Sturge-Weber syndrome. *Eur J Anaesthesiol*; 16(5):339-41, May 1999.
6. DE LEON-CASASOLA OA, LEMA MJ: Anesthesia for patients with Sturge-Weber disease and Klippel-Trenaunay syndrome. *J Clin Anesth*; 3(5):409-13, 1991.
7. COMI AM, HUNT P, VAWTER MP, PARDO CA, BECKER KG, PEVSNER J: Increased fibronectin expression in Sturge-Weber syndrome fibroblasts and brain tissue. *Pediatr Res*; 53:762-769, 2003.
8. GRIFFITHS PD, BLASER S, BOODRAM MB, ARMSTRONG D, HARWOOD-NASH D: Choroid plexus size in young children with Sturge-Weber syndrome. *Am J Neuroradiol*; 17:175-180, 1996.
9. HAPPLE R: Lethal genes surviving by mosaicism: a possible explanation for sporadic birth defects involving the skin. *J Am Acad Derm*; 16:899-906, 1987.
10. HUQ AHMM, GHUGANI DC, HUKKU B, SERAJEE FJ: Evidence of somatic mosaicism in Sturge-Weber syndrome. *Neurology*; 59:780-782, 2002.
11. LEE JS, ASONO E, MUZI O, CHUGANI DC, JUHASZ C, PFUND Z, PHILIP S, BEHEN M, CHUGANI HT: Sturge-Weber syndrome: correlation between clinical course and FDG PET findings. *Neurology*; 57:189-195, 2001.
12. SUJANSKY E, CONRADI S: Outcome of Sturge-Weber syndrome in 52 adults. *Am J Med Genet*; 57:35-45, 1995.