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Anaesthetic Management of a Child with West Syndrome Associated with Tuberous Sclerosis Complex at a Remote Location: A Case Report

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Abstract

West syndrome is a rare syndrome that consists of a triad of infantile spasms, hypsarrhythmia pattern on electroencephalogram and mental retardation. Tuberous sclerosis complex (TSC) is one of the disorders that can cause it. Radiology suites are considered as remote locations for anaesthesiologists, and the delivery of anaesthesia becomes challenging if a patient with such a rare disease having multiple anaesthetic implications arrives. We present anaesthetic management for the radiological procedure of the MRI brain of a year old paediatric patient with the West syndrome having suspected TSC based on presenting signs and symptoms. Anaesthetic consideration and management of this rare syndrome are discussed. Detailed preoperative assessment, pre-emptive preparation for possible difficult intubation and difficult intravenous access, careful positioning and prevention of seizures should be the goal. Thorough knowledge of the disease process, its manifestation and its management is the key to the successful management of such cases.

Keywords: Anaesthesia, child, infantile, isoflurane, sevoflurane, spasms, tuberous sclerosis

Main Points

- West syndrome is a rare syndrome with an incidence of 2-3.5 per 10 000 live births and consists of a triad of infantile spasms, hypsarrhythmia pattern on electroencephalogram and mental retardation.
- Tuberous sclerosis complex (TSC) is one of the rare, autosomal dominant neurocutaneous disorder that can cause the West syndrome.
- Radiology suites are considered as remote locations for anaesthesiologists as they are outside the operating theatres, and the delivery of anaesthesia becomes quite challenging in a patient with West syndrome associated with TSC.
- A thorough multidisciplinary preprocedure evaluation and comprehensive anaesthesia plan are required in such patients to undergo anaesthesia safely.

Introduction

West syndrome is a rare syndrome that consists of a triad of infantile spasms, hypsarrhythmia pattern on electroencephalogram and mental retardation. The incidence of West syndrome is 2-3.5 per 10 000 live births.[1] Any disorder that can cause brain damage in the prenatal, perinatal or postnatal period can cause this syndrome. Tuberous sclerosis complex (TSC) is one of the disorders that can cause the West syndrome. TSC is a rare, autosomal dominant neurocutaneous disorder associated with seizures, eye, heart and kidney hamartoma formations and skin manifestations.

MRI brain is the highest yield tool in identifying the underlying cause of West syndrome, but it is difficult to be carried out on such patients without anaesthesia due to ongoing spasms and noncooperative nature owing to young age and developmental delay.

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Radiology suites are considered as remote locations for anaesthesiologists as they are outside the operating theatres. The delivery of anaesthesia becomes challenging at these places because of cramped dark spaces, unfamiliar environment, insufficient anaesthesia support and variability of monitoring tools.[2]

We present anaesthetic management for the radiological procedure of the MRI brain of a paediatric patient with the West syndrome having suspected TSC based on presenting signs and symptoms. Such patients require a thorough multidisciplinary preprocedure evaluation and comprehensive anaesthesia plan to undergo anaesthesia safely. A written informed consent was obtained from the parent of the child for the case report.

Case Presentation

A year-old boy, weighing 9 kg, diagnosed with West syndrome 15 days back with suspected TSC, presented for having an MRI brain under general anaesthesia. He was already under the care of a paediatric neurologist who started him on vigabatrin for his seizures. He presented with left eye iris coloboma, irregular right pupil and photophobia. He had a couple of ash leaf spots on the body and was developmentally delayed. He was also lactose intolerant and had an egg allergy. His haematological investigations were unremarkable. His electroencephalogram (EEG) showed abnormal generalized seizure disorder and hypsarrhythmia. His ECHO showed an ejection fraction of 85% with normal biventricular function and small echogenic density in the interventricular septum (possible rhabdomyoma). His ultrasound kidney, ureter and bladder (KUB) were normal. His chest was clear on auscultation, and heart sounds were normal. Active infantile spasms were observed preoperatively.

The patient came to us after 6 hours of fasting and was sleeping in his father's arms. He had taken his routine dose of vigabatrin in the morning. The patient was placed on an MRI compatible bed just outside the MRI scanner room, and standard monitoring of heart rate, oxygen saturation, and noninvasive blood pressure were started. Fortunately, the patient kept sleeping during the application of monitoring devices. Before the induction of anaesthesia, a difficult airway intubation trolley was prepared for any unexpected complicated airway issue. Midazolam, phenytoin and levetiracetam injections were kept ready in the drug box. The child was then induced with 8% sevoflurane in 100% oxygen by gently placing a mask on his face to avoid waking him up. After a few minutes, anaesthesia depth was achieved and spasms disappeared. An intravenous cannula was inserted at the right foot as veins were not prominent on the upper extremities. After successful intravenous cannulation, an Ambu AuraOnce laryngeal mask airway (LMA) size 1.5 was inserted successfully. The patient was shifted to bed under an MRI scanner, and careful positioning was done to avoid causing any injury. The LMA was then connected to an MRI-compatible ventilator. The child was covered with blankets to prevent hypothermia. Maintenance of anaesthesia was done by isoflurane MAC 1.5 with 40% oxygen and 60% nitrous oxide. Standard monitoring was continued. The real-time MRI images of the brain showed cortical and subcortical glioneuronal tubers confirming the diagnosis of tuberous sclerosis (Figure 1).

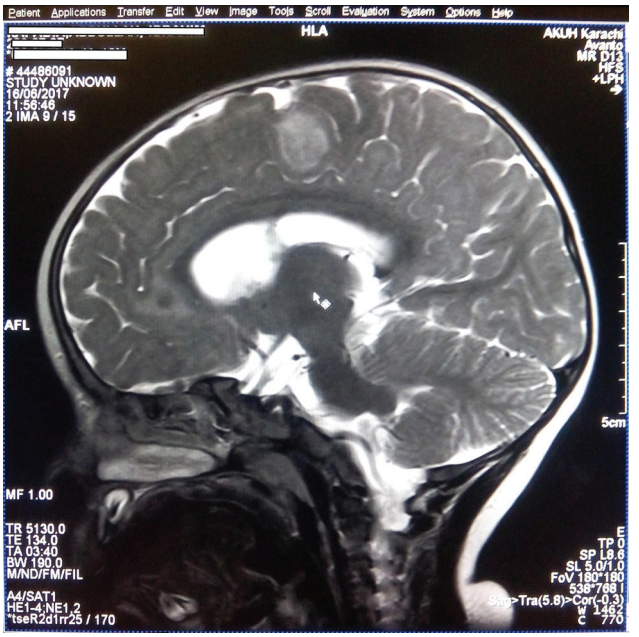

Figure 1. MRI image of patient showing tubers in brain parenchyma.

After 40 minutes of the procedure, isoflurane was discontinued, and the patient was given 100% oxygen. LMA was removed when the child was fully awake. He was then shifted to the recovery room. During the whole procedure, the patient remained haemodynamically stable.

Discussion

West syndrome can occur due to a variety of causes that include genetic, metabolic and malformation syndromes, hypoxic-ischemic or haemorrhagic causes, trauma and infections of the central nervous system. TSC is associated with it and accounts for 10%-30% of prenatal causes.[3]

TSC is a neurocutaneous genetic disorder with multisystem manifestations. The most common neurological manifestation of TSC is mental retardation and seizures.[4] Associated brain lesions include subependymal nodules, cortical tumours and giant cell astrocytomas.[5] Facial angiofibromas are common. Rhabdomyomas of the heart are found in 50% of the patients with TSC. These are benign tumours, but if they are large

enough, they may cause a decrease or obstruction of blood flow through the heart. They may lead to congestive cardiac failure, conduction abnormalities, refractory arrhythmias and severe haemodynamic compromise.[6] Renal manifestations include benign and malignant tumours and renal cysts that may lead to hypertension, renal insufficiency and failure.[5] Lymphangiomyomatosis and multifocal micronodular pneumocyte hyperplasia are pulmonary manifestations associated with TSC and may lead to spontaneous pneumothorax.

Preoperative assessment should be thorough, and one should look out for various abnormalities associated with the disease process. Echocardiogram, ultrasound KUB and chest X-ray are valuable in diagnosing them. Detailed drug history should be elicited as these patients may be on multiple antiepileptic medications that may increase the risk of drug–drug interactions. Haematological and biochemical investigations should be carefully reviewed as well.

The oral examination is crucial as tumours or papillomas may be found on the tongue, palate[7] and posterior pharynx that may cause difficulty in managing the airway. Anatomical malformations may lead to difficult mask ventilation and intubation, and anaesthesiologist should be prepared for such a scenario. It should be made sure that the airway is properly secure after the insertion of a supraglottic device or intubation as the head will become inaccessible after entry into the MRI machine. In our case, a senior anaesthesia resident and consultant anaesthesiologist were present in the scanner room with this patient throughout the procedure.

These patients may also have overactive salivary glands and poor clearance of pharyngeal secretions that require through suctioning during airway manipulation. Postprocedural physiotherapy may be helpful.[8]

Establishing peripheral vascular access may be difficult in patients having active spasms. Moreover, the patient is uncooperative due to age and mental retardation. We induced our patient with sevoflurane and established peripheral vascular access after it. Previous studies conclude that sevoflurane is not associated with seizures during induction and is not contraindicated in patients with epilepsy.[9]

The choice of anaesthetic agents should depend on the condition of the patient and his comorbidities. We used isoflurane for the maintenance of anaesthesia with nitrous oxide and oxygen, and our patient remained hemodynamically stable intraoperatively and postoperatively. Hypoxia, hypercapnia and hypothermia decrease the threshold of seizures and should be avoided. Anaesthetic drugs causing seizures should also be avoided. We prepared anticonvulsive medications beforehand to deal with any intraoperative convulsive event.

Conclusion

Anaesthetic management of a child with West syndrome associated with tuberous sclerosis holds several challenges for anaesthesiologists, especially at the remote location. Thorough knowledge of the disease process, its manifestation and its management is the key to successful management of such cases.

Informed Consent: Written informed consent was obtained from parent of the patient for the publication of this case report.

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