

Anesthetic Management of a Patient with West Syndrome

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Abstract

West syndrome is a rare and severe form of epilepsy that occurs in early infancy. It is characterized by a triad consisting of infantile spasms, mental retardation, and interictal electroencephalogram pattern termed hypsarrhythmia. A thorough pre-operative assessment forms a very important part in the anesthetic management of such patients considering the possibility of difficult intubation because of the coexisting anatomical malformations, establishment of peripheral intravascular access, and careful positioning due to contractures, seizures, and the adverse effects of drugs taken for it. We present anesthesia management of a case of West syndrome with subtrochanteric fracture femur posted for intramedullary nailing.

Keywords: Difficult intubation, infantile spasms, mental retardation, West syndrome.

Introduction

W.J. West was the first to describe West syndrome in 1841 [1]. West syndrome is a rare syndrome characterized by a triad consisting of infantile spasms, mental retardation, and paroxysmal electroencephalogram pattern termed hypsarrhythmia, although diagnosis can be even made if one of the three components is missing [2, 3]. The peak age of presentation is between 3 and 7 months, usually before the age of 2 [4]. Estimated prevalence is 1 in 2000–6000 live births [4]. This syndrome can result from brain dysfunction in the prenatal, perinatal, or postnatal period. Aetiology hypoxic-ischemic or hemorrhagic causes, trauma, infections, genetic syndromes and malformations. Abnormal interplay between the cortex and brainstem has been implicated in its pathophysiology [5]. Such patients should be thoroughly evaluated preoperatively because of difficulties encountered during intubation, intravascular access, positioning, and presence of epileptic seizures with the adverse effects of the antiepileptic medications. We report the successful management of a case of the west syndrome with subtrochanteric fracture femur posted

for intramedullary nailing.

Case Report

A 16-year-old male patient, weighing 18 kg with West syndrome, had sustained a pathological fracture in the subtrochanteric region of the right femur. The patient had an episode of myoclonic convulsion at 8 months of age, was diagnosed to have West syndrome on electroencephalography, and was started with syrup Valparin until 5 years of age and now on tablet valproate sodium 500 mg twice a day. The patient had recurrent seizures despite treatment. Last seizure was 2 months back. The patient had a normal full-term vaginal delivery and cried immediately after birth, and the mother was on anti-Koch's treatment in pregnancy. The patient had global developmental delay and mental retardation with microcephaly and hypertonic movements. The patient was unable to walk but could sit without support. Family history and surgical history were not contributory. Muscle contractures in the extremities and body were present. The respiratory examination revealed decreased air entry on the right side. The cardiac and abdominal exams were normal. Airway examination revealed Mallampati

Class I and no dental caries. The skin was normal. The patient was posted for the fixation of right subtrochanteric femur fracture by intramedullary nailing. We planned for general anesthesia with epidural analgesia. Preoperatively, hematologic, biochemical, and coagulation parameters, Vitamin D3 levels, and arterial blood gas analysis were normal. Chest X-ray revealed scoliosis, and contrast-enhanced computed tomography brain showed hypoplastic corpus callosum with a paucity of sulci. Neurologist opined to continue antiepileptic medications perioperatively. Informed consent was obtained from patient's guardian after explaining the risks involved. NPO status and the administration of the morning dose of antiepileptic medication were confirmed. In the operating room, all standard ASA monitors were attached. Before induction, his pulse was 100 beats/min, and sinus rhythm and blood pressure were 110/70 mmHg, with saturation of 100% on room air. EMLA cream was applied before intravenous (IV) line establishment, and Ringer's lactate was started. Injection thiopentone and injection phenytoin were kept ready in the drug cart. The patient was preoxygenated with 100% oxygen and sedated with injection midazolam 0.03 mg/kg and injection fentanyl 1 µg/kg IV. Induction was done with injection propofol 1 mg/kg IV and injection atracurium 0.8 mg/kg IV. Doses were titrated as per the patient's response. The airway was secured

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with a 6.0 mm endotracheal tube, and after confirmation, the patient was maintained on oxygen:nitrous oxide (1:1) and sevoflurane MAC 0.8. Minute ventilation was adjusted to maintain normocapnia. Epidural catheter was inserted in L2-3 interspace for analgesia in the left lateral position. 0.125% bupivacaine at 5 ml/h infusion was used for intraoperative analgesia. Dependent parts were adequately padded. The patient was positioned carefully in the lateral position to avoid reoccurrence of fracture and supported with silicone pads. Temperature was monitored, and forced air warmers, warm IV, and irrigation fluids were used to prevent hypothermia. The intraoperative course was uneventful with stable hemodynamics and adequate urine output. At the end of surgery, the patient was turned to the supine position. After thorough suctioning, return of airway reflexes and with the patient being fully awake, extubation was done. He was shifted to the recovery room for observation. 0.125% bupivacaine boluses 4 ml were given at 8 h intervals postoperatively for analgesia. There were no seizures in the post-operative period.

Discussion

Dr. W. J. West first described infantile spasms in 1841 in his son. He had written a letter to the editor of the journal "Lancet" describing the events as heaving of the head forward to the knees followed by relaxation into upright position [1, 3, 6]. The eponym West syndrome was created in the early 1960's by Millichap [7]. For years, there was no treatment success achieved for this syndrome. Sorel and Dusaucy reported in 1958 the use of ACTH for the treatment of infantile spasm [7, 8]. West syndrome has a poor prognosis and resistant to standard antiepileptic medications [3]. West syndrome has a heterogeneous etiology, with hypoxic-ischemic encephalopathy, and

infections being important causes. Etiologically, it is divided into symptomatic with known etiology (60–80% of cases) and cryptogenic etiology [5]. Management of a case of West syndrome poses several challenges. It can be associated with tuberous sclerosis which accounts for 10–30% of the causes in the prenatal period, involving cardiac and kidney tumors [9]. Skin examination was done to rule out the characteristic "ash leaf" spots seen in patients with tuberous sclerosis. Therefore, the underlying etiology should be carefully evaluated in the pre-operative anesthetic assessment, and the appropriate mode of anesthesia and drugs should be preferred. The drug history and the laboratory findings should be carefully evaluated. Periodontal problems are common in patients taking antiepileptic medications. Dental caries are common in these patients [10]. Gingival hyperplasia with bleeding, petechiae, and decreased platelet aggregate are noticed on long-term administration of valproate [11]. Prolonged high dose of vigabatrin leads to visual constriction in these patients [12]. ACTH is effective for seizure control in West syndrome but is associated with serious adverse effects such as osteoporosis, cardiac hypertrophy, hypertension, tendency to develop infections, electrolyte imbalance, behavioral changes, and weight gain [13]. Chronic use of antiepileptic medications can have effects on liver enzyme activity and metabolism of anesthetic drugs, thus needing dose adjustment. Patients with West syndrome have global developmental delay with mental retardation. They have poor clearance of pharyngeal secretions. Overactive salivary glands and cranial nerve dysfunction increase salivation requiring thorough suctioning during intubation and extubation. Post-operative chest physiotherapy should be instituted. Malnourishment and hypothalamic

dysfunction predispose to hypothermia. The minimum alveolar concentration of inhalational anesthetic agent may be decreased which makes them more prone to delayed emergence. It should be ensured that the patients are fully awake before extubation [14]. They require longer observation periods and longer hospital stay. Muscle spasm in the form of head and body flexion and leg extension are frequent, and hence, IV line establishment can be challenging. Anatomical malformations can predispose to difficult intubation. In our case, the patient had muscle contractures in the extremities and body. To avoid fractures, the patient was positioned carefully and the joints were padded. Measures were taken to prevent seizures by avoiding hypoxia, hypercapnia, hypothermia/hypothermia, and medications that decrease the seizure threshold. There was preparedness for the management of convulsions if it occurred in the intraoperative course.

Conclusion

A thorough pre-operative evaluation with a good anesthetic plan for the course and measures to avoid decrease in seizure threshold will help in managing this difficult and rare case of West syndrome.

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