Anaesthesia Management of a Child with West Syndrome

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West syndrome (WS) is an epileptic encephalopathy usually occurring during the first year of life and is characterized by severe electroencephalography (EEG) derangement. Most of these patients may develop cerebral palsy, facial malformations, and skeletal deformities. The anaesthesiologist should make the preoperative assessment carefully due to epileptic seizures and should consider the possibility of difficult intubation because of coexisting anatomic malformations during the anaesthesia management of patients with WS. This report presents a case of general anaesthesia management in a left femoral fixation operation in an 11-year-old, 18 kg male patient.

Key Words: West syndrome, general anaesthesia, facial malformations, skeletal deformities

Introduction

West syndrome is one of the malignant epilepsies rarely seen in the infantile period, which consists of the hypsarrhythmia triad, characterised by infantile spasm, mental disorder and electroencephalography (EEG) abnormality (1). This syndrome can result from brain dysfunction in the prenatal, perinatal (during delivery) or postnatal period. Its aetiology includes infections, trauma, hypoxic-ischemic or haemorrhagic causes, malformation syndromes and genetic syndromes. Its pathophysiology contains an abnormal interaction between the brain stem and cortex (2). Females are affected more than males at a proportion of 1.3:1. Its incidence rate is 1 per 3200-3500 live births.

Prednisone, adrenocorticotrophic hormone (ACTH), antiepileptic drugs and benzodiazepine are used for the treatment of West syndrome. At any time and for any reason, surgery can be needed for a child diagnosed with WS. A preoperative evaluation should be carefully performed for these patients, considering possible difficulties in intubation, establishment of peripheral vascular access and positioning due to anatomic malformations, side effects of the drugs and epileptic seizures. Also, a proper anaesthesia plan should be prepared. This case report aims to present anaesthesia management in a patient with WS who was planned to undergo open reduction and internal fixation under general anaesthesia.

Case Presentation

The patient, born at 40 weeks, had not had any problem during delivery. When he had been taken to a paediatrician for routine control at the age of 3.5 months by his parents, his convulsions had been noticed. He had been diagnosed with WS as a result of the examinations performed, and his treatment had been initiated. Open reduction and internal fixation were planned for 11-year-old and 18 kg patient due to a left femur fracture. In the preoperative evaluation, the patient was found to have mental retardation. One year ago, he had convulsions 2-3 times in a week. However, he had not been having convulsions for the last year, and he was taking vigabatrin 1000 mg day⁻¹ as anti-epileptic treatment. His haemogram, serum electrolytes, fasting blood glucose and liver function tests were within the reference intervals. The physical examination of his appearance revealed a prominent ear, chest deformity, contraction deformities in all extremities and micrognathia. His respiratory sounds were wheezing, and there were crackles in both lower zones. The Mallampati score was III. For the patient, from whose parents written consent was obtained, all alternative airway tools were kept available in the operating room for the possibility of a difficult intubation due to micrognathia. After preoperative 8-hour fasting, peripheral vascular access was hardly established because of muscle contractures. Then, 0.05 mg kg⁻¹ midazolam i.v. was administered. and the patient was taken to the operating room. For the patient. who underwent standard monitoring, the blood pressure was 110/70

mm Hg, heart rate was 98 beats min^{-1} , SpO_2 was 98% and body temperature was 36.7°C. Muscle contractures in the extremities, body and neck were apparent, and re-occurrence of fractures was avoided during positioning. In parallel with the physiological state of the patient, he was placed into the lateral position and supported with silicone pads (Figure 1). It was seen that ventilation was easy after 2.5 mg kg^{-1} propofol and 0.5 $\mu\text{g kg}^{-1}$ fentanyl i.v. were given in induction, and preoxygenation was applied with 100% O_2 for 3 minutes. Then, 0.1 mg kg^{-1} vecuronium bromide was given. After that, endotracheal intubation was performed using a number 5 endotracheal tube without any problem. The maintenance of anaesthesia was provided with a mixture of nitrous oxide and oxygen at a ratio of 2:1 and 1MAC sevoflurane. Ventilation was administered in such a way that end-tidal CO_2 was 30-40 mmHg. No intraoperative problem was observed, and the patient was taken into the paediatric intensive care unit following the surgery. No seizure-like activity developed clinically during the 48-hour postoperative period. The patient was taken into the service.

Discussion

West syndrome was defined as “infantile spasms” by Dr. West in 1841; 110 years later, it was reported that the syndrome consisted of the triad of “infantile spasm, physical and mental disorder and characteristic EEG findings”. One year later, the characteristic EEG finding was named “hypsarrhythmia”. For years, no treatment success was achieved for the syndrome. Eventually, in 1958, Sorel and Dusaucy-Bauloye reported that infantile spasm disappeared with ACTH, and hypsarrhythmia in the EEG was treated.

There may be many reasons for the occurrence of West syndrome. Among these, especially genetic and malformation syndromes, hypoxic-ischemic or haemorrhagic causes, central nervous system infections and trauma have an important place. Tuberous sclerosis accounts for 10%-30% of the causes in the prenatal period. Since tuberous sclerosis is a syndrome involving cardiac and kidney tumours, it is important for anaesthesia (2). Therefore, the underlying causes should be carefully investigated in the preoperative evaluation of cases, and the appropriate anaesthetic method and agents should be preferred.

Infantile spasms are generally resistant to antiepileptic drugs. Although the mechanism of steroid therapy is not known (ACTH or prednisone), it can yield better results. Recently, positive results have been reported, particularly with vigabatrin treatment, among antiepileptic drugs. However, long-term use of high-dose vigabatrin leads to visual field constriction in nearly half of the patients (3). Despite the fact that ACTH is effective in seizure control for most WS patients, it has serious side effects, including osteoporosis, cardiac hypertrophy, hypertension, tendency to infection, electrolyte imbalance, behavioural changes and weight gain (4). While evaluating patients in the preoperative period, the drugs that

Figure 1. Physical appearance features of the child: prominent ear, chest deformity, contraction deformities in all extremities and micrognathia

they use should be investigated in detail, and laboratory findings should be assessed.

In these patients, head, body and hip flexion or extension or often head and body flexion and leg extension can be seen secondary to muscle spasm (4). All of these are factors that cause difficulty in the establishment of peripheral vascular access, as well as being anatomic malformation reminding difficult intubation. Nonetheless, this physiological posture of the patient requires attention to the intraoperative position. Our case had contractures in the head, neck, body and extremity, micrognathia and chest deformity.

Although we had difficulty in the establishment of peripheral vascular access, no problem was encountered during intubation carried out in the lateral position. In our case, muscle contractures were apparent in the extremities, body and neck. Therefore, positioning was performed carefully in order to avoid re-occurrence of the fracture. The joints of the patients were supported with silicone pads, and he was placed into the lateral position in accordance with his physiological state.

Co-morbid diseases and convulsions should be taken into consideration while administering anaesthetic agents in patients with West syndrome. Although the aetiology of convulsions with propofol is controversial in studies, it was reported that intraoperative convulsion might have resulted from propofol in a 75-year-old patient who underwent laryngeal microsurgery under total intravenous anaesthesia with propofol (5). Despite this, Yamaguchi et al. (6) used propofol for anaesthesia induction and maintenance of a patient with WS. As a result, they stated that propofol was an appropriate agent for this patient, but it had to be used carefully, since it did not decrease the seizure threshold. In another case with WS, who was to be operated on for gingival hypertrophy and was expected to have intubation difficulties, nasotracheal fiberoptic intubation was performed under sedation with remifentanyl (7).

In some studies, it was demonstrated that the use of inhalation agents in high concentrations in anaesthesia induction caused seizure-like electrical activity. However, it was revealed that the agitation occurring during sevoflurane anaesthesia was not associated with seizures and that sevoflurane was not

contraindicated in patients with epilepsy (8-10). Nieminen et al. (11) stated that an epileptiform pattern in the EEG was not observed during anaesthesia performed using i.v. midazolam for premedication in children and 2% sevoflurane for maintenance.

In our case, midazolam was used for premedication due to its anxiolytic, sedative and anticonvulsive effects. Despite the inconsistency with the literature, propofol was employed in induction in order to avoid the side effects of high-dose inhalation agents. Sevoflurane, which is an anaesthetic commonly used in children and has become clear for convulsion, even a little, was preferred for maintenance.

Conclusion

In West syndrome, during preoperative preparation, the drugs used and associated complications should be investigated, and the patient should be evaluated with regard to difficult intubation and peripheral vascular access. Use of midazolam for premedication, propofol for anaesthesia induction and sevoflurane for maintenance can be adequate and safe. We suggest that undesirable events can be prevented by providing good anaesthetic depth and analgesia during surgery and by avoiding conditions, such as hypoxia and hypocarbia, that will decrease the convulsion threshold.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - S.H.Ş.; Design - S.H.Ş., E.Ç.; Supervision - S.H.Ş., E.Ç.; Funding - S.H.Ş., G.S.; Materials - S.H.Ş.; Data Collection and/or Processing - S.H.Ş.; Analysis and/or Interpretation - S.H.Ş., H.U., A.Ç., G.S., E.Ç.; Literature Review - S.H.Ş., H.U., G.S.; Writer - S.H.Ş., Critical Review - S.H.Ş., E.Ç., H.U., A.Ç., G.S.; Other - S.H.Ş., E.Ç., H.U., A.Ç., G.S.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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