CASE REPORT

Anesthesia Management in a Child with Letterer Siwe Disease

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letterer siwe disease is a rare disease and anesthesia management of patient with this disease is very challenging because of multiorgans involvement and poor information about anesthesia. We manage a child with this disease was refered to operating room for humerous bone fracture surgery.
Keywords: letterer siwe; anesthesia

Letterer–Siwe disease is one of the four recognized clinical syndromes of Langerhans cell histiocytosis (LCH). It causes approximately 10% of LCH disease and is the most severe form [1]. Prevalence is estimated at 1:500,000 and the disease almost exclusively occurs in children less than three years old [2]. The name is derived from the names of Erich Letterer and Sture Siwe. Letterer-Siwe is characterized by skin lesions, ear drainage, lymphadenopathy, osteolytic lesions, and hepatosplenomegaly. The skin lesions are scaly and may involve the scalp, ear canals, and abdomen [3]. The disease is often rapidly fatal, with a five year survival rate of 50%. The development of thrombocytopenia is a poor prognostic sign [1].

We represent a 4 yrs old child with Letterer Siwe Disease that was referred to operating room because of humerous bone fracture.

Case Description

A 4-year-old female child, weighing 13 kg, diagnosed as a case of Letterer Siwe disease was consulted for humerous bone fracture surgery under general anesthesia. In preoperative evaluation vital sign were in normal range, temperature was 37.1°c, hemoglobin was 8.5 mg/dl and platelet were 105000 and we reserved one unit packed red blood cell before surgery. We found some seborrheic lesions in the scalp. Mouth opening was in normal range but dentation was irregular. Lung auscultation was clear and CXR according to a radiologist report was acceptable. We found painless hepatosplenomegaly in our patient. Hepatic and coagulation system labratoary tests were in normal range. Because of probability of diabetes insipidus in this patient we requested electrolyte and urine osmolarity tests and they were normal. The patient was under glucocorticoid treatment and we infused stress dose corticosteroid before surgery. Because of bone involvement in this patient we were very sensitive about patient transferring and positioning. We prescribed 0.25 mg/kg midazolam 0.5 hour before surgery and in operating bed we used fentanyl 2 µ/kg and 1mg midazolam as a premedication and then we used thiopental 4mg/kg and 200 micro cisatracuriom for induction. After 3 minutes patient was intubated with 5 uncuffed tracheal tube with video laryngoscope. We used pressure control mode (PCV) for ventilation during surgery. Our monitoring was pulse oximetry, capnography, 3 leads electrocardiography, invasive blood pressure monitoring and temperature monitoring. Our maintenance drugs were sevoflurane 4-5 % and remifentanil 0.2 µ/kg/min. Surgery lasted for about 2:30 hours and we infused 80 cc packed RBC isogroup and iso RH because of bleeding according to MABL (maximum allowable blood loss). We infused 0.1 mg/kg morphine sulfate for pain management near the end of surgery. At the end of surgery when we found spontaneous breathing we reversed neuromuscular block with neostigmine 40 µ/kg and atropine 20 µ/kg and when airway reflex was normal the patient was extubated. We monitored patient in PACU (post anesthetic care unit) for one hour and then patient was transferred to PICU (pediatric intensive care unit). The patient was discharged 2 days later without any problem related to surgery and anesthesia.

Discussion

Letterer-Siwe disease represents a disseminated form of LCD, which usually afflicts children in the first year of life, but can be seen in any age, including adults [4]. There are pinkish-yellow to red-brown coalescent scaly papules, plaques, and patches over seborrheic areas, including the scalp, groin, nasolabial folds, perioral area, and upper trunk. Less commonly, there are pustules, petechiae, erosions, or ulcerated nodules [5]. The skin lesions are accompanied by systemic symptoms of fever, lymphadenopathy, weight loss, pancytopenia, and hepatosplenomegaly [6]. The main

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features of Letterer–Siwe disease are lymphadenopathy, hepatosplenomegaly and bone and skin lesions. Fever, infections and bleeding tendencies are secondary to pancytopenia, which results from marrow displacement by the histiocyte-like cells [7].

It sometimes follows a rapidly fatal course. Rarely, death follows within a week of diagnosis. Diagnosis is by biopsy, showing infiltration of the tissues by proliferating histiocyte-like cells. Treatment is with radiotherapy, corticosteroids and cytotoxic drugs, and may be successful. Occasionally, the course of the disease is less acute and recoveries are possible [8]. This disease is a rare disease and information about anesthetic management is poor. We should consider many organ involvement and request laboratory tests related to these involvements. We managed a 4 yrs old child in this case presentation successfully.

Conclusion

We have to consider multiorgan involvement and request appropriate laboratory test during management of letterer-siwe disease. It's better to manage this patient in PICU after surgery.

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