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LETTER TO THE EDITOR

Anesthesia for intestinal obstruction in a 6 year old child with COFS syndrome

Anestesia para obstrução intestinal em uma criança de 6 anos de idade com síndrome COFS

Dear Editor:

We report our anesthesia management of a 6 years old girl with Cerebro-Oculo-Facio-Skeletal (COFS) Syndrome who was consulted as intestinal obstruction in emergency department of our hospital. She was already a follow-up patient of pediatric department and diagnosed as COFS syndrome by them previously.

She had characteristic signs of the syndrome including growth failure, weighning 6 kg, cachexia, micrognathism, microcephaly. She had small mouth opening and dental abnormalities, flexure contractures at her elbow and knees. Her preoperative blood tests were within normal limits except C-reactive protein (40 mg/dL) and White Blood Cell (18×10^3). Plevral effusion and abdominal volvulus was found in her radiological images. Her initial vital signs were in normal range. Intravenous access was established by 24 gauge catheters. Anesthesia was induced with sevoflurane 8% in air–oxygen mixture and 12 mcg fentanyl intravenously. Intubation was done with a cuffed 4.0 mm endotracheal tube succesfully. Anesthesia maintenance was provided with sevoflurane 2% in air–oxygen mixture, and fentanyl. She was ventilated in pressure control ventilation. There was no anesthetic problems during the operation that continued for 135 min. After adequate breathing spontaneously, she was extubated smoothly.

COFS syndrome is initially described by Pena and Shokeir in 1974 and has been recognized as a rare, autosomal recessive disorder characterized by DNA repair defect.[1] Cockayne syndrome Type II is known as COFS syndrome. Degenerative problem of the brain and spinal cord is usually seen before birth. Clinical findings are psychomotor development delay, neurological dysfunction, peripheral neuropathy, microcephaly, micrognathism, hypotonia, hyporeflexia, convulsions, and congenital cataract. And is associated with feeding difficulties, sensorineural hearing loss, coxa valga, knee flexion contracture.[1] Progressive neurological degeneration can lead to a risk of hyperkalemia by using succinylcholine in these patients. Reports can be seen describing the anesthesia management of Cockayne syndrome in the literature but there was no about the anesthesia management of COFS syndrome. Difficult airway management was one of the most important problem in anesthesia procedure. Also, due to the contractions and osteoporosis, adequate care should be considered while giving position during the surgery.

That is difficult to predict the effects of muscle relaxants' in these patients for they are cachectic, almost lack of muscle with insufficient development of the neuromuscular junction for this, they are need to be avoided. We saw that only one case that muscle relexant was not used during anesthesia in the literature.[2] Endotracheal intubation was done via sevoflurane combined with nitrous oxide in this patient who was scheduled for liver biopsy, and the procedure lasted for 13 min. We did not use muscle relexants in induction and maintenance of anesthesia, too. Our procedure continued 135 min. We used weight-appropiate endotracheal tube than age-appropiate one but we prefferred cuffed – tube to avoid the risk of aspiration.

In conclusion; all of these associated anomalies and dysfunctions requiring attention and experience. Using smaller endotracheal tubes and avoiding muscle relexants during intubation can be kept in mind during anesthesia management of COFS syndrome.

Conflicts interest

The authors declare no conflicts of interest.

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