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Letter to Editor

Anesthesia Management of a Child with Mosaic Trisomy 22 for Adenotonsillectomy Surgery

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Mosaic trisomy 22 syndrome is an infrequent autosomal abnormality that is compatible with life. In contrast, complete mosaic trisomy 22 is incompatible with life. Occurrence of trisomy 22 is 1/30000-50000 of living births. The syndrome is characterized with intellectual and developmental disability, cleft palate, webbed neck, facial dysmorphism and organ malformations such as cranial abnormalities, renal malformations and congenital heart disorders. Therefore, this syndrome is a challenge to physicians [1-3]. In the review of articles, a few articles have been found about anesthesia management in this syndrome. So, herein we report an 8-year-old boy (19 kg) who diagnosed with mosaic trisomy 22 in the neonatal time, was planned for adenotonsillectomy surgery. He was second child of the family and had an elder healthy brother at age of 16. Both his mother and father were healthy and third- degree consanguineous. Previously, he had surgery for his atrial septal defect (ASD), cleft palate, cryptorchidism and congenital diaphragmatic hernia repair surgeries, and a history of seizure. He also had a history of gastroesophageal reflux disease (GERD), congenital hypothyroidism and congenital bilateral hearing loss. He took levothyroxine and pantoprazole and used hearing aid. He had normal intelligence and asymmetry in face and ears, esotropia and blepharoptosis in the left eye and flattened nasal bridge (Figure 1).

Figure 1- Known case of mosaic trisomy 22 syndrome at age 8 years with ear asymmetry, left eye blepharoptosis and flattened nasal bridge

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All routine laboratory investigations and thyroid function tests were in normal range. EF (Ejection fraction) and PAP (pulmonary artery pressure) were normal. In physical examination, mallampati score was 2. The electrocardiogram revealed sinus regular rhythm. After counseling with a pediatric cardiologist and pediatrician, he was transferred to operating room for surgery. Before surgery, standard monitoring was attached and vital signs were: BP 101/61 mm Hg, HR 127 beats/min, RR 16 breaths/min and SPO2 was 96. After preoxygenation with oxygen, midazolam (1mg) and fentanyl (50 µg) were administered intravenously. Anesthesia was induced with sodium thiopental (5 mg/kg) and atracurium (0.5 mg/kg). Then he successfully intubated with endotracheal tube size 5 (cuffed). Anesthesia was continued uneventfully with isoflurane (0.5-1.2%) and O2:N20 (50:50). During surgery, ETCO2 (end-tidal CO2) maintained between 35 to 40 mmHg. Neostigmine (1 mg) and atropine (0.5 mg) were injected intravenously to reverse the neuromuscular block in the end of surgery. He was extubated successfully and transported to the recovery room. Mosaic trisomy 22 syndrome is a chromosome derangement in which chromosome 22 is exhibit three times in some cells of the body, instead of the normal two times. The range and severity of this rare abnormality can vary widely. Although a number of these cases have normal growth and development, others have growth delays and hemidystrophy (unequal development of both sides of the body). In the literature, there is a fewer than 20 reports of live born children and this patient is one of them. Characteristics and associated clinical findings are prenatal and postnatal growth delay, mental retardation, cranial abnormalities, hemiatrophy, facial dysmorphism, webbed neck, ptosis, ocular, renal and genital anomalies, flat nasal bridge, hearing loss, ear and limb malformations, cleft palate and cardiac abnormalities [1-4]. In review in literature, we could not find any anesthesia management for these patients. In our patient, because of history of seizure, sodium thiopental was used. Eventually, this complicated child successfully managed with midazolam, fentanyl, sodium thiopental, atracurium and isoflurane 0.5-1.2% in O2:N2o (50:50). The tracheal intubated with a smaller cuffed endotracheal tube, and ETCO2 maintained in normal range. Patients with this condition are at increased risk for concurrent disease such as cardiac dysfunction, congenital anomalies and disorders (such as cleft palate or hypothyroidism) and aspiration risk due to GERD (like our patient) [1,2,4]. In clinical practice, sometimes we cannot find any anesthesia management of a rare clinical case in the published literature, therefore, in the absence of the same, practice of safe anesthesia should guide the management for reducing morbidity and mortality. Ultimately, surgery in childhood for mosaic trisomy 22 syndrome with congenital abnormalities that affect multiple systems require understanding of the problems that may be encountered. This case showed that in some situation, isoflurane, nitrous oxide and atracurium could be safely used for anesthesia management in this syndrome.

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