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Case Report

Anesthetic Management of an Infant with Bilateral Radial Dysplasia and Isolated Patent Ductus Arteriosus: A Case Report and Brief Review of Literature

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ABSTRACT

Bilateral radial dysplasia is a rare condition and usually associated with familial malformation syndromes and spectrum of cardiac anomalies. These patients often require corrective orthopedic surgeries in early age. Radial dysplasia associated with cardiac abnormalities is particularly seen in certain syndromes such as Holt-Oram (HOS) syndrome and TAR (thrombocytopenia, anemia, and radial aplasia) syndrome. We report a successful management of twelve months old male child with rare combination of bilateral radial dysplasia with isolated patent ductus arteriosus (PDA), for corrective orthopedic surgery. A comprehensive approach, which includes an extensive preoperative evaluation and pre-defined perioperative hemodynamic goals for PDA, plays a pivotal role in its perioperative management.

Radial dysplasia is a longitudinal deficiency of radius resulting in radial deviation of wrist and shortening of forearm. It is an uncommon condition with estimated incidence of 1:30,000 to 1:100,000 births and is more common in boys [1-2]. The anesthetic management in these patient is complex owing to its association with various cardiovascular anomalies including atrioseptal defect (ASD), ventriculoseptal defect (VSD), coarctation of aorta, dextrocardia and pulmonary stenosis [1-2]. It may be associated with various syndromes such as Holt-Oram (HOS) syndrome, VACTERAL association (vertebral abnormalities, anal atresia, cardiac anomalies, tracheoesophageal fistula, and renal defects), Fanconi anemia and TAR (thrombocytopenia, anemia, radial aplasia) syndrome [1-3]. We report a successful management of an infant with rare combination of bilateral radial dysplasia with isolated patent ductus arteriosus (PDA), for corrective orthopedic surgery.

Case Report

A twelve months old male child, weighing 6 kg, was scheduled for surgical correction of bilateral radial dysplasia with abnormally short forearms and absent thumbs. Child had an uneventful full term delivery with no developmental delay. There was no history of similar congenital abnormality in the family. General physical and respiratory system examination were insignificant. Cardiovascular examination revealed bounding peripheral pulses, grade III continuous murmur in the left infraclavicular area. Routine blood investigations were normal. Two-dimensional echocardiography (2D-Echo) findings were suggestive of a patent ductus arteriosus (PDA) with left to right shunt with Qp/Qs ratio of 1:2. There was no history of concurrent chest infection and cyanosis.

On the day of surgery, child was kept fasting for breast milk, 4 hours prior to surgery and was premedicated with

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oral midazolam (3 mg) and EMLA™ (Eutectic Mixture of Local Anesthetic) cream was applied on dorsum of left hand in preoperative room, half an hour before taking in the operating room (OR). Subsequently, after 30 minutes of EMLA application, 22 gauge intravenous (IV) cannula was secured. In the OR, standard monitors [continuous electrocardiography (ECG), pulse oximeter (SPO2), noninvasive blood pressure (NIBP)] were applied. Child was induced with sodium thiopentone (30 mg IV) and fentanyl (15µgm IV) was administered. Child trachea was intubated with uncuffed endotracheal tube (4.0 mm internal diameter), two minutes after administration of cis-atracurium (1mg IV). General anesthesia was maintained with Air: O2 (50:50) and sevoflurane (2-6%) to achieve a minimal anesthetic concentration (MAC) of 1.7-2.0. Child was placed on pressure controlled volume-guaranteed (PCV-VG) mode of ventilation (Tidal volume: 35 ml, respiratory Rate: 20/minute, inspiratory-expiratory (I: E) ratio of 1:2). Isotonic IV fluid (0.9% Normal Saline) was administered for fasting, maintenance and surgical loss.

Multimodal analgesia was initiated with paracetamol (45mg IV) and titrated aliquots of fentanyl (5-10ug IV) during intraoperative period. Surgical procedure took 2 hours which was followed by reversal of neuromuscular blockade (Neostigmine: 360 ug IV and Glycopyrolate 60ug IV) and child trachea was extubated successfully. Intraoperative and postoperative period remain uneventful.

Discussion

Bilateral radial dysplasia is usually associated with familial malformation syndromes [1-2]. Radial dysplasia associated with cardiac abnormalities are particularly seen in certain syndrome such as HOS and TAR syndrome [1-3].

TAR syndrome describes thrombocytopenia in infancy with bilateral radial aplasia and congenital heart disease, however, thumbs are invariably present [4]. We ruled out the possibility of the TAR syndrome, owing to the absence of thumb, anemia or any coagulation abnormalities in the child.

HOS syndrome is an autosomal dominant disorder with genetic defect on long arm of chromosome 12 (12q2). Mutation on T-box genes on chromosome 12q2 leads to certain skeletal and spectrum of cardiac malformations [3]. The cardiac anomalies mainly constitute atrial septal defect (ASD), ventricular septal defects (VSD) and other complex conduction defects [3]. The isolated PDA with bilateral radial dysplasia as seen in this child, may present as an atypical presentation of HOS syndrome.

Children with congenital heart disease (CHD) for non-cardiac surgeries are associated with two-fold increase in mortality [5]. Ramamoorthy et al reported that anaesthesia-related paediatric cardiac arrest occurred in around 75% of patients under 2 years of age with CHD during non-cardiac surgery [6]. Hence, comprehensive preoperative evaluation and testing is essential for delineating the type, nature, severity of lesion and subsequent anesthetic plan. Preoperative evaluation includes birth history, history of cyanosis, frequent respiratory infections, previous cardiac surgery and familial association. Medication history must be thoroughly elicited as these children with cardiac anomalies may be on various drugs such as aspirin, warfarin, diuretics, angiotensin converting enzyme (ACE) inhibitors and antiarrhythmic drugs and has its perioperative implications. Cardiovascular examination may reveal characteristic murmur and signs and symptoms suggestive of congestive heart failure such as respiratory infections, feeding intolerance, fatigue, dyspnoea, irritability and failure to thrive. Signs of pulmonary congestion, increased pulmonary vascular resistance (PVR) and reversal of left to right shunt (Eisenmenger's syndrome) should be ruled out before planning for elective non-cardiac surgeries [7].

Preoperative investigations must include complete hemogram and coagulation profile to rule out particular congenital malformations such as Fanconi anemia and TAR syndrome, specifically seen in association with radial dysplasia. In addition, chest x-ray imaging maybe required to rule out cardiomegaly and signs of pulmonary congestion. These subset of patients also require 12-lead ECG to rule out dysrhythmias and conduction defects. It is pertinent to obtain hand X-ray to determine presence or absence of thumb to differentiate between TAR and HOS syndrome. Baseline 2D-Echo is required to determine nature, severity of cardiac lesions, pulmonary hypertension and pulmonary blood flow to systemic blood flow ratio (Qp/Qs). Genetic karyotyping is also advised to rule out the genetic abnormalities. In the present child we suspect atypical presentation of HOS syndrome, as isolated PDA with bilateral radial dysplasia is rarely mentioned in the literature [3]. However, we have not done gene karyotyping due to limited access of resources.

Patients with isolated PDA has left to right shunt, hence balanced anesthetic technique must be planned to prevent any increase in PVR and prevention of pulmonary congestion and reversal of shunt. Maintaining a fine balance between the systemic and pulmonary vascular resistance is crucial during perioperative period. Pre-operative fasting should not be prolonged due to the risk of hypovolemia and subsequent cardiac compromise. Certain drugs such as ACE inhibitors should be avoided preoperatively and volume status should be restored if child is on diuretics. Sympathetic stimulation due to crying should be avoided by adequately premedicating the child with drugs such as midazolam preferably by oral or nasal route [8].

Hyperventilation and 100% oxygen administration should be avoided as it may lead to pulmonary vasodilation and subsequent pulmonary congestion.

Inhalational induction is well tolerated in this subset of patient with minimal decrease in cardiac output. Nitrous oxide (N2O) should be avoided as it increases PVR. Incremental doses of sodium thiopentone (4-6mg/kg IV) is well tolerated in normovolemic child with compensated CHD. Alternatively, ketamine (1-2mg/kg IV) maintains contractility and SVR without increasing in PVR and maybe used as an induction agent. SVR should be maintained as precipitous fall in SVR may lead to reversal of shunt. Propofol is generally avoided as it may cause precipitous fall in SVR which can be detrimental for a patient with large PDA, by causing pulmonary run off during diastolic phase leading to coronary ischemia especially in a hypovolemic child [9]. We chose thiopentone as an induction agent, as the child was very apprehensive and non-cooperative for inhalational induction. Moreover, we have secured IV line in the preoperative room after application of EMLA patch and hence IV induction was administered with ease.

Children with PDA are also prone to fatal myocardial ischemia due to pulmonary runoff during diastolic phase leading to low diastolic blood pressure [10]. Close monitoring of hemodynamic and ECG changes indicating myocardial ischemia is hence essential. Factors leading to increased PVR lsuch as hypothermia, hypercarbia, hypoxia, acidosis, catecholamine release should be addressed. Adequate intra- and post-operative pain management is essential to prevent any further rise in PVR, hence we initiated multimodal analgesia during perioperative period.

Conclusion

Infant with bilateral radial dysplasia along with isolated PDA, which maybe an atypical presentation of HOS syndrome, was reported as a rare clinical entity in literature. These subset of patients are commonly posted for corrective orthopedic surgery. A comprehensive approach, which includes an extensive preoperative evaluation and pre-defined perioperative hemodynamic goals for PDA, remains a crucial factor for successful anesthetic management.

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