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Case Presentation

Anaesthesia Challenge: Kasai's Portoenterostomy in a CMV Positive Infant with Tetralogy of Fallot and Biliary Atresia

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Biliary atresia (BA) is a rare progressive neonatal disorder of the biliary tree leading to inflammatory obstruction and destruction of intra hepatic and extra hepatic bile ducts along with liver cirrhosis [1].

Even though the exact causative factor of biliary atresia is unknown, several factors such as immune dysregulation, viral infections, toxins are potential agents [2].
Multiple studies on Biliary atresia in many countries has shown CMV to be a potential cause of biliary disease in infants leading to biliary tree sclerosis [3-5].

Tetralogy of Fallot (TOF) is the most common congenital cyanotic heart disease in paediatric age, characterised by four defects; Right ventricular enlargement, Ventricular septal defect, overriding of aorta and right ventricular outflow obstruction [6]. No association of TOF has been found with BA, and only one case of CMV with TOF has been reported in literature. Biliary atresia, which shows a greater association with congenital cardiac anomalies than the general hospitalised population, but potentially similar to the association between cardiac anomalies and choledochal cysts, has also been previously linked to cardiac disease [7].

Case Report

A 3-month-old female infant weighing 4.5kg was admitted with complaints of reduced feeding, fever and abdominal distension. On examination the child was found to have a heart rate of 141 per minute, temperature of 100 Degrees Fahrenheit and an oxygen Saturation of 78% on right arm and 91% on left arm suggesting a Patent Ductus Arteriosis. The child was also found to be icteric. Chest examination revealed an ejection systolic murmur and bilateral coarse crepitations.

A chest X Ray showed a boat shaped heart and prominent broncho vascular markings. Ultrasound of the abdomen showed situs inversus abdominis.
A 2D echocardiography showed a 6.2mm peri membranous Ventricular septal defect with left to right shunt; valvular Pulmonary Stenosis with a 70mm Hg of PSG; Enlarged Right ventricle, Right atrium with a flat Inter-ventricular septal motion, Aorta arising from the Right ventricle; DORV and 2-4 mm PDA with left to right shunting.

Blood investigations showed Hb of 14.5gm/dL, Total leukocyte count of 12,000 cells/ cubic mm, INR of 1.0, Serum urea 11mg/dl, Serum creatinine of 0.4, Alpha fetoprotein 123.2, Albumin of 3.1gm/dL, Total bilirubin of 8.87, Direct Bilirubin of 4.98, SGOT 73, SGPT of 60.

The child was started on Oral Valgancyclovir 450mg BD. Respiratory tract infection was managed with IV antibiotics, antitussives and steam inhalation.

The child was shifted to a pre-warmed operating theatre, routine monitors (ECG, Pulse oximeter, Non-invasive blood pressure monitor, temperature probe) were attached. Premedication of Injection Glycopyrrolate 0.032mg, Injection Fentanyl 8mcg IV was given. Induction was done with Injection Ketamine 8mg IV and Injection Succinylcholine 9 mg IV after pre oxygenation with 100% for 5 minutes. The child was intubated with 3.5mm Uncuffed endotracheal tube with direct laryngoscopy. Maintenance of anesthesia was done with mixture of Oxygen and Sevoflurane (MAC - 1) using JR circuit, Injection Atracurium 2mg IV loading dose and a maintenance dose of 0.4mg Injection Fentanyl 1 mcg/kg IV was given as per need.
The child remained hemodynamically stable intraoperatively. IV fluids of 40 mL vol Lactated ringers was administered during the peri operative period. Injection Dexamethasone 0.8mg IV and Inj. Calcium gluconate 40mg IV was given at the end of the procedure. Neuromuscular blockade was reversed with Injection Neostigmine 0.8mg IV and Inj. glycopyrrolate 0.06mg IV. The child was extubated with good motor power and spontaneous respiration. Injection Paracetamol 60 mg IV was given for Post operative analgesia and the child was shifted to surgical ICU.

Discussion

A rare triad of Biliary atresia, Tetralogy of Fallot and Cytomegalovirus Infection is a unique aspect of the case.

Biliary atresia can either present in isolation, associated with laterality malformations – poly splenia, situs inversus, cardiac anomalies like PDA, ASD, VSD or be associated with other congenital anomalies like intestinal atresia, renal malformations etc [8].

It is a common cause of surgical cholestatic jaundice in neonates and Kasai's hepato- portoenterostomy remains the only treatment done in the first three months of life, especially in developing countries like ours thereby decreasing the need of eventual liver transplant.

CMV infection further reduces the prognosis in these patients due to impaired immunity [9].

Infants with unrepaired cyanotic congenital heart disease posted for major non cardiac surgery are always considered high risk and should be assessed pre operatively. Dehydration should be avoided. Increased myocardial oxygen consumption should be minimized by reducing sympathetic stimulation and premedication with Injection Midazolam 0.008mg/kg -0.01mg/kg doses.

Ketamine is the preferred induction agent. Systemic Vascular resistance should be maintained and increase in pulmonary vascular resistance should be avoided.

Pulmonary hypertension should be prevented by avoiding hypothermia, stress responses to pain, acidosis, hypercarbia, hypoxia, increase in intra thoracic pressure.

ECG monitoring is essential and pain management is critical.

In our scenario IV opioid was the mainstay analgesic agent. Regional anesthesia was avoided due to the altered hepatic physiology. Bradycardia induced by traction on the IVC and liver and peritoneum were managed with Injection Atropine IV. Warm ambient temperature of the operating room was maintained and complete draping the child was done to prevent hypothermia.

Hence, we infer that infant with congenital anomalies heart defects and viral infections should be examined meticulously and managed appropriately on a case-to-case basis.

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