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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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Abstract

Biliary atresia is a multifactorial neonatal disorder affecting the extrahepatic bile ducts leading to cholestatic jaundice and eventual liver transplantation. The most frequent viral infection linked to biliary atresia is CMV. It has not yet been documented that Tetralogy of Fallot, a common cyanotic congenital cardiac defect, is connected to BA. An unusual feature of the case is the rare combination of cytomegalovirus infection, tetralogy of Fallot, and biliary atresia.

Abbreviations

ASD: Atrial Septal Defect; BA: Biliary Atresia; CMV: Cytomegalovirus; IV: Intravenous; IVC: Inferior Vena Cava; JR: Jackson Reese; MAC: Minimal Anaesthetic Concentration; PDA: Patent Ductus Arteriosus; RVOT: Right Ventricular Outflow Tract Obstruction; SVR: Systemic Vascular Resistance; TOF: Tetralogy of Fallot; VSD: Ventricular Septal Defect

Introduction

An uncommon progressive newborn condition of the biliary tree, biliary atresia (BA) causes liver cirrhosis in addition to inflammatory blockage and destruction of the intra- and extrahepatic bile ducts [1].

Even though the exact causative factor of biliary atresia is unknown, several factors such as immune dysregulation, viral infections, toxins have been found to be incriminating 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].

As the most prevalent congenital cyanotic heart disease in children, tetralogy of Fallot (TOF) is defined by four defects: Aortic overriding, ventricular septal defect, right ventricular hypertrophy, and right ventricular outflow blockage. There is only one documented instance of CMV with TOF in the literature, and no correlation between BA and TOF has been discovered as yet [6]. Congenital heart defects in general have been previously connected to biliary atresia, which exhibits a higher connection with congenital cardiac malformations than the general hospitalized population, but may be similar to the association between cardiac defects and choledochal cysts [7].

Case Report

A 4.5 kg, 3-month-old female infant was brought in with complaints of decreased feeding, fever, and distended abdomen. Upon inspection, the child's temperature was recorded at 100 degrees Fahrenheit, heart rate of 141 beats per minute, and oxygen saturation levels of 78% on the right arm and 91% on the left, indicating the possibility of a patent ductus

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arteriosus. 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 6.2 mm perimembranous Ventricular septal defect with left to right shunt; valvular Pulmonary Stenosis with a 70 mmHg 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.

The laboratory investigations were as follows. Hb was 14.5 gm/dL, total leukocyte count was 12,000 cells/cubic mm, International Normalised Ration (INR) was 1.0, serum urea was 11 mg/dl, serum creatinine was 0.4, alpha fetoprotein was 123.2, albumin was 3.1 gm/dL, total bilirubin was 8.87, direct bilirubin was 4.98, SGOT was 73, and SGPT was 60.

The child was started on oral Valgancyclovir 450 mg twice a day. Steam inhalation, antitussives, and intravenous antibiotics were used to treat the respiratory tract infection.

On the day of the surgery, the child was shifted to a pre-warmed operating theatre and routine monitoring devices (Electrocardiogram (ECG), Pulse oximeter, Non-invasive blood pressure, temperature probe) were attached. Premedication of Injection Glycopyrrolate 0.032 mg, Injection Fentanyl 8 mcg IV was given. After preoxygenation with 100% oxygen for 5 minutes, induction was done with Ketamine 8 mg IV and Succinylcholine 9 mg IV. The child was intubated with 3.5 mm Uncuffed endotracheal tube under direct laryngoscopy. Maintenance of anesthesia was done with mixture of Oxygen and Sevoflurane (MAC-1) using Jackson-Reese (JR) circuit, Atracurium 2 mg IV loading dose and a maintenance dose of 0.4 mg IV after intervals of 15-20 minutes. For analgesia, Fentanyl 1 mcg/kg IV was given as per need.

The child remained hemodynamically stable intraoperatively. IV fluids of 40 mL vol Lactated ringers were administered during the peri operative period. Dexamethasone 0.8 mg IV and Calcium gluconate 40 mg IV was given at the end of the procedure. Neuromuscular blockade was reversed with Neostigmine 0.8 mg IV and Glycopyrrolate 0.06 mg IV as the child started showing signs of spontaneous respiration and weaning off muscle relaxation. Subsequently, infant was extubated with good motor power and spontaneous respiration. Paracetamol 60 mg IV was given for postoperative analgesia and the child was shifted to the pediatric intensive care unit (PICU).

Discussion

The rare combination of tetralogy of Fallot, biliary atresia, and CMV infection is an uncommon aspect of this case.

Based on anatomical bases, BA is categorized according to the degree and kind of obstruction. The majority of the time, BA is either complete (Japanese/Anglo-Saxon type III, 73%) or subcomplete (type IIb, 18%), with types IIa and I (6% and 3%, respectively) and "distal" BA being rare. In addition to being isolated, biliary atresia can also be linked to other congenital abnormalities such as intestinal atresia, absence of retrohepatic vena cava, renal malformations, polysplenia, situs inversus, and cardiac defects such PDA, ASD, and VSD [2,8].

It is a prevalent cause of surgical cholestatic jaundice in newborns, and in underdeveloped nations like ours, Kasai's hepatoportoenterostomy is still the only treatment performed during the first three months of life, reducing the need for eventual liver transplant in the future. The addition of CMV infection further reduces the prognosis in these infants due to impaired immunity [9].

Preoperative evaluation is recommended for infants with untreated cyanotic congenital heart disease who are scheduled for significant non-cardiac surgery, as they are always at high risk. Perioperative management of infants with uncorrected TOF posted for noncardiac surgery is difficult because of the long-term effects of hypoxia and decreased pulmonary blood flow, which lead to significant physiologic alteration and neurological sequelae. Cyanotic episodes are typically caused by either a reduction in Systemic vascular resistance (SVR) or an infundibular spasm, or spasm of the cardiac muscle in the Right Ventricular Outflow Tract (RVOT) region as a result of sympathetic activation. Beta blockers like propranolol or esmolol are used to treat infundibular spasm, whereas α-adrenergic agonists like phenylephrine or norepinephrine and intravenous fluids are used to address any drop in SVR. Because it increases SVR, which reduces right-to-left shunting, ketamine is the recommended induction drug in TOF.

Mitigating increased myocardial oxygen consumption requires lowering sympathetic activation and using dosages of midazolam between 0.008 and 0.01 mg/kg. Avoiding hypothermia, stress reactions to pain, acidosis, hypercarbia, hypoxia, and increases in intrathoracic pressure can help prevent pulmonary hypertension. In order to diagnose arrhythmias and tachycardia, intraoperative ECG monitoring is crucial. Avoiding dehydration is advised.

Pain management is crucial because elevated sympathetic activity brought on by pain can precipitate a cyanotic episode during the perioperative phase. For pain management, we administered fentanyl boluses of 1 mcg/kg during surgery and a paracetamol infusion afterward.

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 Inferior vena cava (IVC) and liver and peritoneum, were managed with Atropine IV. In order to prevent hypothermia, the operating room was kept at a comfortable temperature, and the infant was completely draped. Care was taken to ensure that the IV line was clear of air bubbles as they have the potential to cause fatal paradoxical embolism due to right-to-left shunting.

Thus, it follows that babies with congenital cardiac abnormalities and viral infections need to be carefully inspected and treated according to their individual needs.

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Conflicts of Interest

None.

References

1. Hartley JL, Davenport M, Kelly DA (2009) Biliary atresia. Lancet 374: 1704-1713.
2. Wildhaber BE (2012) Biliary atresia: 50 years after the first Kasai. ISRN Surg 2012: 132089.
3. Zani A, Quaglia A, Hadzić N, Zuckerman M, Davenport M (2015) Cytomegalovirus-associated biliary atresia: An aetiological and prognostic subgroup. J Pediatr Surg 50: 1739-1745.
4. Lakshminarayanan B, Davenport M (2016) Biliary atresia: A comprehensive review. J Autoimmun 73: 1-9.
5. Xu Y, Yu J, Zhang R, Yin Y, Ye J, et al. (2012) The perinatal infection of cytomegalovirus is an important etiology for biliary atresia in China. Clin Pediatr (Phila) 51: 109-113.
6. Dwivedi P, Kumar S, Ahmad S, Sharma S (2020) Uncorrected tetralogy of fallot's: Anesthetic challenges. Anesth Essays Res 14: 349-351.
7. Murphy AJ, Axt JR, Lovvorn HN 3rd (2012) Associations between pediatric choledochal cysts, biliary atresia, and congenital cardiac anomalies. J Surg Res 177: e59-e63.
8. Schwarz KB, Haber BH, Rosenthal P, Mack CL, Moore J, et al. (2013) Extrahepatic anomalies in infants with biliary atresia: Results of a large prospective north american multicenter study. Hepatology 58: 1724-1731.
9. Kodo K, Sakamotoa K, Imaib T, Otac T, Miyachic M, et al. (2018) Cytomegalovirus-associated biliary atresia. J Pediatr Surg Case Rep 35: 17-20.