Case Report

Perioperative anesthetic management of a patient with biliary atresia, situs inversus totalis, and kartegener syndrome for hepatobiliary surgery

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Abstract

Patients with genetic disorders associated with multiple congenital anomalies present unique challenges to the anesthesiologist. We report the successful perioperative management of a child with biliary atresia, situs inversus totalis, and Kartegener syndrome scheduled for corrective biliary surgery. We recommend that patients with multiple congenital anomalies need to be thoroughly and cautiously evaluated. The perioperative management should be individualized based on associated anomalies along with appropriate monitoring.

Key words: Anesthesia, biliary atresia, hepatobiliary surgery, Kartegener syndrome, situs inversus totalis

Introduction

Patients with disorders associated with multiple congenital anomalies present unique challenges to the anesthesiologist during surgical or technical procedures. Biliary atresia (BA) is a rare disease and the end result of a destructive inflammatory process in bile ducts, leading to fibrosis and liver cirrhosis.[1] Situs inversus (SI) is a condition characterized by a mirror image orientation of the abdominal and thoracic viscera relative to the midline. Interestingly, up to 28% of children with SI have biliary atresia (BA) and SI is one of the components of Kartegener syndrome.[2,3] The perioperative surgical concerns in such a child are mentioned in literature but literature on perioperative anesthetic concerns is lacking. We report a child with syndromic biliary atresia, situs inversus totalis, and Kartegener syndrome scheduled for corrective biliary surgery.

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Case Report

A 2-month-old 4.3 kg male child was diagnosed as extrahepatic biliary atresia and was scheduled for laparotomy. The child was brought to the pediatrics clinics 2 weeks earlier with complaints of rapid breathing since 12 days, vomiting (non bilious) since 8 days, and decreased oral intake since 1 day. The child was passing clay colored stools and dark colored urine. There was yellow discoloration of skin since 2 weeks, which was progressively increasing. The mother revealed the presence of recurrent rhinorrhoea from the first day of life.

Examination revealed icterus. Abdomen examination showed hepatospenomegaly (liver on left side and spleen on right side) and umbilical hernia. Electrocardiogram (ECG) displayed inverted P waves in I, aVL and aVR, which were suggestive of situs inversus. Chest X-ray revealed evidence of bronchopneumonia and the cardiac shadow was on the right side. Ultrasonography abdomen also indicated situs inversus. Echocardiography was suggestive of situs inversus with dextrocardia with normal cardiac anatomy. Hepatobiliary iminodiacetic acid (HIDA) scan was indicative of extra hepatic biliary atresia (EHBA) with the liver in left upper abdomen. Child was diagnosed as situs inversus with dextrocardia, bronchopneumonia, and extrahepatic biliary atresia.

Bronchopneumonia was managed with intravenous antibiotics, paracetamol, and steam inhalation. The child was placed

on therapy with oral phenoparbitone 10 mg twice a day and intravenous Vitamin K 5 mg once a day for 10 days. Investigations revealed hemoglobin 9.5 g/dL, total leucocyte count (TLC) 17400/mm^3, platelet count 4.5 lac/mm^3, serum sodium 131 meq/L, serum potassium 4.4 meq/L, random blood sugar 70 mg/dL, serum bilirubin 19.2 mg/dL (normal 0.2-1.2) (direct-12, indirect 7.2), SGOT 148IU/L (normal 15-50) , SGPT 93 IU/L (normal 15-50), alkaline phosphatase 1232 IU/L (normal 50-300), blood urea 18 mg/dL (15-45), serum creatinine 0.6 mg/dL (0.6-1.2), prothrombin time (PT) 11.3 sec (test), 12 sec (control), and aPTT test 38.7 sec Control 29 sec (normal 20-40 sec). The child was listed for corrective biliary surgery.

The operating room was kept warm prior to shifting of the child. In the operating room, routine monitors (electrocardiogram, pulse oximeter, non invasive blood pressure) were attached. Left-sided ECG leads were attached on the right side and vice versa. Anesthesia was induced with intravenous fentanyl (9 μg) and thiopentone (25 mg), and lungs were ventilated with isoflurane in oxygen and nitrous oxide (50:50) (MAC 1.2) using Jackson Rees modification of Ayre's T piece. After achieving neuromuscular blockade with atracurium (2.5 mg), trachea was intubated with endotracheal tube size 3.5 mm ID. Capnography and temperature monitoring was also initiated. Urethra was catheterized and urinary output was monitored. Anesthesia was maintained with desflurane in oxygen and nitrous oxide (50:50) (MAC 1) using paediatric close circuit along with boluses of atracurium (0.5 mg) and fentanyl (1 μg) as required. The child was kept warm using warm fluids, warming mattress and by adequately covering the child with cotton.

The surgeon found the liver on left side with right lobe on the left side, malrotated gut (caecum, appendix on the left side), two atretic gall bladders, annular pancreas with preduodenal portal vein, cirrhotic enlarged liver, and a multi-lobulated spleen. Hepatic roux-en-y portoenterostomy with Ladd's procedure was done. The surgical incision site was infiltrated with 6 mL of 0.125% bupivacaine. The hydration was maintained using normal saline. The surgery lasted 5 hours, and blood loss of 100 mL was replaced with packed red blood cells.

At the end of surgery, the airway resistance increased. The airway pressures increased from 18 cm H_2O to 28 cm H_2O. Endotracheal tube suctioning was done. On auscultation, breath sound was reduced on the right lung fields. Repeat suction was done. We suspected a mucus plug to be obstructing the endotracheal tube and changed the tube after repeat laryngoscopy. A mucus plug was seen at the tip of the tube. The ventilation became better. Residual neuromuscular blockade was reversed and trachea was extubated. The child had good respiratory efforts with bilateral equal air entry. The child was shifted to the pediatric intensive care unit for further management. The child was nursed in the incubator and ECG and pulse oximeter monitoring were continued. The vitals and urinary output were within normal limits. The humidified oxygen was supplemented using an oxygen hood. The postoperative hemoglobin was 9.9 g/dL. The serum electrolytes were normal. The analgesia was provided with rectal paracetamol suppository. The child had an uneventful recovery and was discharged on the 18th postoperative day.

Discussion

The case reported highlights an extremely rare entity of syndromic biliary atresia associated with Kartagener syndrome, which has not been reported in the literature. The anesthetic concerns in our patient included managing the abnormalities related to liver pathology (deranged metabolism, coagulation problems), major surgery and its associated concerns (analgesia, blood loss, hypothermia), respiratory concerns (Kartegener syndrome, repeated infections, inspissations of secretions), monitoring (electrocardiogram interpretation, invasive monitoring).

BA has two forms: 1) syndromic or fetal or embryonic (10-35%) with various congenital anomalies such as splenic anomalies (polysplenia, double spleen, asplenia), portal vein anomalies (preduodenal, absence, cavernomatous transformation), situs inversus, malrotation, cardiac anomalies, annular pancreas, Kartagener's syndrome, duodenal atresia, esophageal atresia, polycystic kidney, cleft palate, and jejunal atresia 2) non syndromic (70-90%), in which BA is an isolated anomaly.[4-6]

BA is said to occur in 28% of infants born with SI as compared to the 0.01% of the general population.[7] SI is a rare anomaly, with a frequency reported to be between 0.002% and 0.1%.[3,8] Complete heart block is found in over 20% of patients in patients with situs inversus.[3]

Kartagener syndrome (primary ciliary dyskinesia), a combination of situs inversus, bronchiectasis, and male infertility is attributed to abnormal ciliary motility.[2,3] Diagnosis is frequently made late, in part because it presents with symptoms which are common in children (rhinitis, secretory otitis media, cough). It was suspected in our case because of the presence of recurrent rhinorrhea, situs inversus totalis, and biliary atresia. Symptomatology was indicative of the presence of Kartergener syndrome in our patient though we could not get confirmatory evaluation done by ciliary motility studies. This mandated the need of optimization of respiratory

status prior to surgery and care for prevention of inspissations of secretions but it still happened in our patient. In view of the possibility of secretions blocking the endotracheal tube one has to have a low threshold for endotracheal tube suctioning, and keep a close watch on changes in lung compliance and peak inspiratory pressures.

The ECG must be cautiously monitored as these patients may have associated heart blocks. We placed the ECG leads in reverse orientation for monitoring intraoperatively to correctly interpret the findings. Central venous catheters must be placed cautiously because of the likelihood of associated vascular anomalies in relation to situs inversus and dextrocardia. The chest X-ray needs to be cautiously interpreted while checking for the correct placement of invasive catheters.

The postoperative oxygen supplementation should always be humidified. The patient's hydration should be well maintained to prevent any inspissation of secretion in the airway and potential chance of airway compromise. Analgesia was a major concern in this patient in view of major surgery. Inadequate analgesia may lead to respiratory insufficiency in milleu of compromised respiratory mechanics due to presence of Kartegener syndrome. A caudal block was avoided because of hepatic pathology. Intraoperative analgesia was provided with intravenous fentanyl. Postoperative analgesia was taken care by incision site infiltration with bupivacaine and rectal paracetamol. There are currently no biologic markers available for monitoring possible hepatotoxicity in such patients with liver disease, but a single-dose suppository has been mentioned to be satisfactory analgesic alternative.[9] Paracetamol should be used as a part of multimodal analgesia but care should be taken to limit its use in patients with liver pathology.

The altered anatomy during the surgical procedures in such patient was a concern for the surgeons. They required relatively more duration for the surgery as tissue handling was difficult and the orientation was difficult to interpret. Placement methodology of surgical instruments for surgical procedure also required modification for suitable exposure of the tissue. Handling of the liver tissue lead to hemodynamic changes probably because of vessels kinking (inferior vena cava) and thus lesser venous return, but this was conservatively managed. In view of prolonged surgery, temperature should be monitored and temperature actively maintained using warm fluid, warming mattresses, blankets, and covering the child.

To conclude, patients with syndromic anomalies with multiple congenital defects need to be thoroughly and cautiously evaluated. The perioperative management should be individualized based on associated anomalies along with appropriate monitoring.

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