Case Reports
ISSN (Print): 2394-6954
ISSN (Online) : 2394-9775
Karnataka Anaesthesia Journal Vol 18(1-2), January-June 2020, p. 37-40 DOI: 10.4103/kaj/2020/v18i1-2/159365

Anaesthetic Management of Kartageners Syndrome Posted for Kasai Surgery

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Abstract

Kartagener's syndrome is a rare autosomal recessive disorder presenting with a triad of sinusitis, bronchicetasis and situs inversus with dextrocardia. It occurs in 50% of patients with situs inversus .Patients with genetic disorders associated with multiple congenital anomalies present unique challenges to the anesthesiologist. We report the successful intraoperative 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.

Keywords: Anesthesia, Biliary Atresia, Hepatobiliary Surgery, Situs Inversus Totalis

1. Introduction

Kartagener Syndrome (KS) is a rare, autosomal recessive disorder with variable penetrance. This entity, first described by Kartagener in 1933, is now known to be a form of primary ciliary dyskinesia, better known as immotile cilia syndrome[1]. Its clinical presentation is varied with non-specific and heterogeneous clinical manifestations. There is not a single, gold standard, diagnostic test which delays the diagnosis. It's important to have high index of suspicion and prepare the patient accordingly to ensure smooth conduct of anaesthesia and perioperative care.

2. Case Report

A 7-month-old 5.35 kg female child diagnosed as extrahepatic biliary atresia was scheduled for kasai procedure. Child presented with a 2 month history of gradually progressing yellowish discolouration of eyes and whole body, and passage of pale coloured stools Normal birth history and immunisation with an insignificant past history were noted. On physiological examination baby was irritable with icterus. Per abdominal examination revealed abdominal distension with hepatomegaly. Cardiovascular examination showed dextrocardia, with normal heart sounds and no murmurs. Blood biochemistry showed haemoglobin of 8.5mg with altered coagulation profile and increased liver parameters. Ultrasound abdomen showed hepatosplenomegaly and situs inversus. Situs solitus and dextrocardia were seen on 2d-Echocardiography. Tc99m mebrofenin hepatobiliary scan showed enlarged liver with impaired hepatocyte function and non-visualisation of gut activity up to 24hr, consistent with extrahepatic biliary obstruction (Figure 1).

Baby was premedicated with inj.midazolam 0.5mg IV and nebulised with saline, the operating room was kept warm prior to shifting of the child. Routine monitors like ECG, non-invasive blood pressure cuff, pulse-oximeter were attached. Left-sided ECG leads were attached on the right side and vice versa. Precordial stethoscope attached on the left side of the chest. Baseline Heart rate was 110bpm, blood pressure being 66/40mmHg and room

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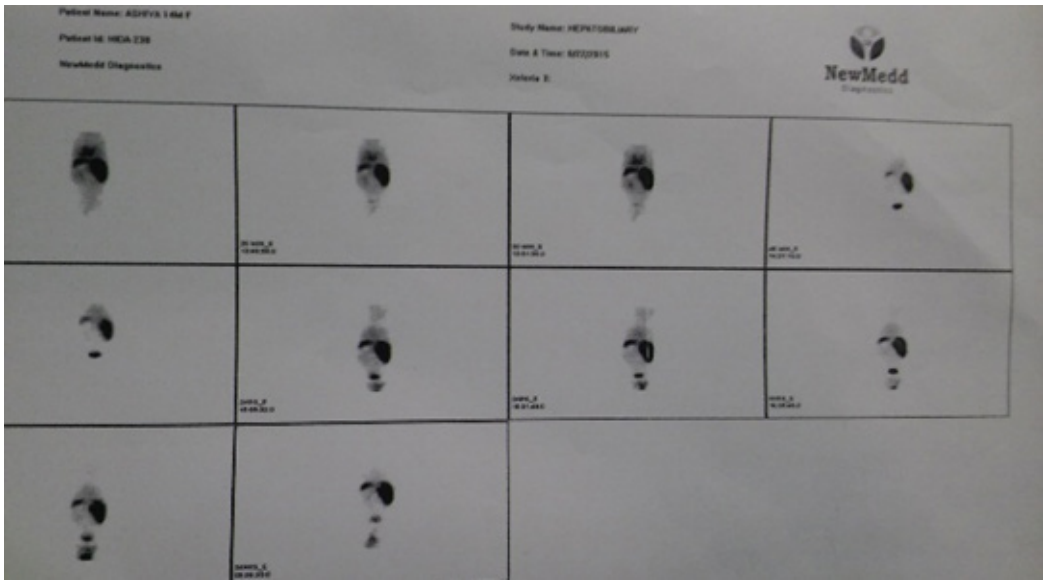

Figure 1. Radionuclide imaging showing biliary atresia.

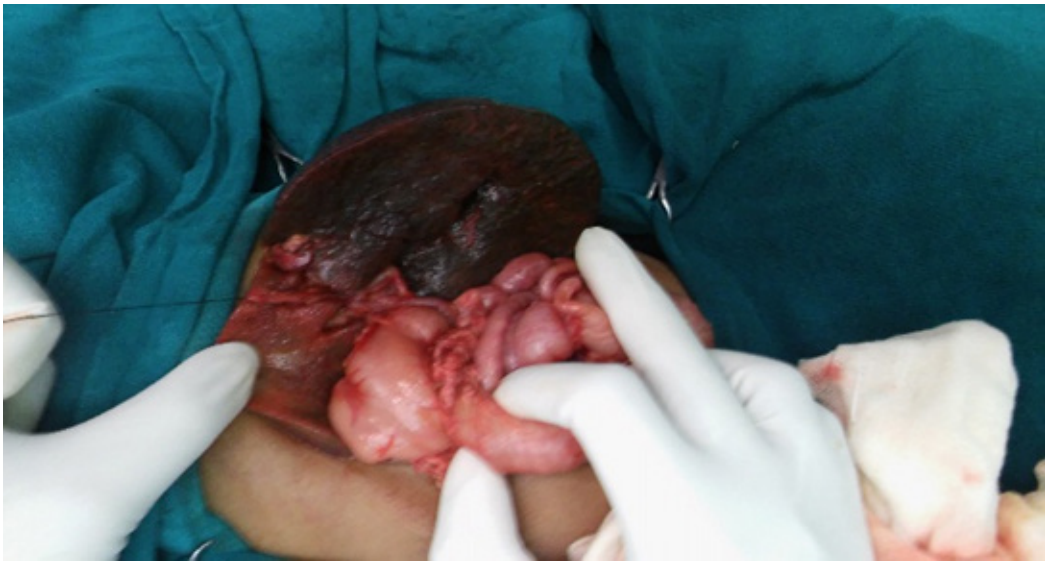

Figure 2. Multiple spleen.

air saturation of 98%. Intravenous Ringer lactacte 30ml/hr infusion was started. General anesthesia was induced with intravenous fentanyl 10ug, thiopentone 30mg. After achieving neuromuscular blockade with atracurium (3mg), trachea was intubated with endotracheal tube size 4.5 mm ID uncuffed tube. Capnography and temperature monitoring was also initiated. Urethra was catheterized and urinary output was monitored. Anesthesia was maintained with isoflurane in oxygen and air (50:50) (MAC 1) using paediatric close circuit along with boluses of atracurium (0.5 mg) and fentanyl (1 μ g) as required. The child was kept warm using warm fluids, humidified air, warming mattress and by adequately covering the child with cotton pads. Operative findings were of a cirrhotic liver, ascitic gall bladder with annular pancreas, preduodenal portal vein with malrotation and polysplenia (3 in no) (Figure 2). 60ml blood trasfusion was done

using blood warmer over 40 minutes. After biliary atresia correction and closure of abdomen, local infiltration with Bupivacaine 0.125 5ml and Inj paracetamol 200mg IV given. Baby was electively ventilated, shifted to ICU for observation. Day 2 baby was extubated and day 10 discharged from hospital.

3. Discussion

The incidence of the genetic disorder is 1 in 32,000 births[2], however, higher incidences have been found in consanguineous marriages[3]. Kartagener's syndrome is a variant of the immotile cilia syndrome and occurs in 50% of patients with situs inversus[4]. ICS is rarely recognised in the neonatal period and the diagnosis is commonly made in childhood, adolesence or adulthood. It is characterized by triad of sinusitis, bronchiectasis and situs inversus with or without dextrocardia. Sinusitis and chronic ear and respiratory infections as well as male infertility are the most common presentations, but congenital heart diseases could also be observed[4]. Patients with disorders associated with multiple congenital anomalies present unique challenges to the anesthesiologist during surgical or technical procedures.The main anaesthetic considerations among patients with Kartagener's syndrome are related to the pulmonary function which include preoperative respiratory infections due to bronchiectasis[4]. So in this study we took special care to prepare the child that included preoperative intravenous antibiotics, saline nebulisation, chest physiotherapy, good hydration. We should also monitor potentially occluded congenital heart diseases. 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[5]. During the embryological development, a 270 degree clockwise rotation instead of normal 270 degree anti-clockwise of the developing thoraco-abdominal organs results in mirror image positioning of the abdominal and thoracic viscera[6]. The association of situs inversus totalis with syndromes such as Kartagener's syndrome, cardiac anomalies, spleen malformations and other such clinical entities makes the clinical scenario extremely challenging for the concerned anaesthesiologist[6].

Primary (genetic) defects in the structure and function of sensory and motile cilia result in multiple ciliopathies. A reduction in the number of arms which propel mucus (dynein arms) is a common abnormality but many other structural defects of the cilia have been found[7].

Structures that make up the cilia including inner and/or outer dynein arms, central apparatus, radial spokes, etc. are missing or dysfunctional and thus the axoneme structure lacks the ability to move. Axonemes are the elongated structures that make up cilia and flagella. Additionally, there may be chemical defects that interfere with ciliary function in the presence of adequate structure. Whatever the underlying cause, dysfunction of the cilia begins during and impacts the embryologic phase of development.

The Kasai aims at restoring bile flow between the liver and the intestine, using a jejunal Roux-en-Y limb, which is anastomosed to the porta hepatis after resection of the biliary remnant. This hepato-porto-enterostomy is named after the Japanese surgeon Morio Kasai, who first performed this procedure in 1959.

Synonyms: Afzelius' syndrome, KS, Kartagener's triad, Siewert's syndrome, dextrocardia-bronchiectasis-sinusitis syndrome, primary ciliary dyskinesia.

4. Conclusion

General anaesthesia would be safe after complete preanaesthetic examination of the patient. ECG, chest CT scans, spirometry and echocardiography are mandatory before planning for surgery. The precise diagnosis of situs inversus and a thorough pre-operative evaluation can minimize, to a large extent, the difficulties and the various potential challenges associated with its anaesthetic management. Keeping all this in mind, it becomes easier and safer to successfully manage the patients of Kartagener's syndromein the operation theatre.

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