Case Report

Anesthetic management of a pediatric patient with situs solitus and dextrocardia

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ABSTRACT

Dextrocardia with situs inversus is a rare heart condition with a genetic predisposition. Although most individuals lead a normal healthy life and usually, it is an incidental finding. Due to their unique anatomical variations and associated congenital variations, they may pose challenges to attending clinicians. We are hereby reporting a successful anesthetic management of the case of a 2.5-year-old child who presented for emergency laparotomy and on investigations, was found to have dextrocardia along with situs solitus.

Key words: Anesthesia, Dextrocardia, Emergency, Situs solitus

Dextrocardia is a rare heart condition characterized by abnormal positioning of the heart in the right hemothorax. In this condition, the tip of the heart (apex) is positioned on the right side of the chest [1,2]. Its incidence is about 1/10,000 live births [1]. The exact cause of dextrocardia is also unknown. However, it has been linked with several factors such as maternal diabetics, cocaine use, autosomal recessive gene inheritance, and conjoined twinning with no gender predisposition [3-6]. Usually, the individuals lead a normal life, and anomaly is usually an incidental finding when the individual reports to the hospital for some other medical condition. Such patients can pose special challenges during perioperative periods, especially in emergency surgery that too in a pediatric subgroup.

We are hereby reporting the successful management of a 2.5-year-old female child with dextrocardia and situs solitus, who reported to our hospital with the complaint of intestinal obstruction and undergo emergency laparotomy.

CASE REPORT

A 2.5-year-old female presented with a history of constipation and associated abdominal distension. She was born by a cesarean section after a full-term pregnancy. She was having no history of cyanosis, jaundice, respiratory distress, or seizures since birth. Her developmental history was normal as per age.

On examination, the child was lethargic, with a weight of 10 kg. The heart rate was 106 beats/min and arterial saturation (SPO_2) was 96% on room air. On chest auscultation, air entry was present with no added sound. There was the presence of hand anomalies on both hands (Fig. 1).

The airway examination was within normal limits. However, on computer tomography (CT) abdomen, the internal organs were found to be in a mirror position with her 2-D echo showing dextrocardia. Pulmonary venous drainage was not well seen, but there was no evidence of intracardiac structural disease. Chest X-ray showed dextrocardia (Fig. 2). On blood investigations, her hemoglobin was 8.6 g/dL, and on electrocardiogram (ECG), q waves were present.

After consultation with a cardiologist, the patient was planned for diversion colostomy for surgery under moderate risk. The patient was taken to the prewarmed operation theater. The Standard American Society of Anesthesiologists monitors were attached including ECG and SpO_2. ECG was showing a reverse pattern in the normal position, so reverse lead placement was done as the patient was having dextrocardia. An intravenous line was secured on the left foot with a 24G cannula. The patient was premedicated with injection glycopyrrolate 5 ug/kg iv and fentanyl 1 ug/kg. The patient was induced by injection ketamine 2 mg/kg and intubated with injection atracurium 5 mg. She was put on the pressure control mode of ventilation. Anesthesia was maintained with N_2O: O_2 50:50 sevoflurane. After the procedure, the patient was reversed with injection neostigmine 0.03 mg/kg iv and injection glycopyrrolate 0.05 mg iv. The patient was extubated after the condition of the patient got satisfactory and shifted to the recovery room. The patient has an uneventful stay at the hospital and is discharged on the seventh postoperative day in satisfactory condition.

Access this article online

Received - 19 March 2023
Initial Review - 31 March 2023
Accepted - 26 May 2023

Quick Response code

DOI: 10.32677/ijcr.v9i5.3933

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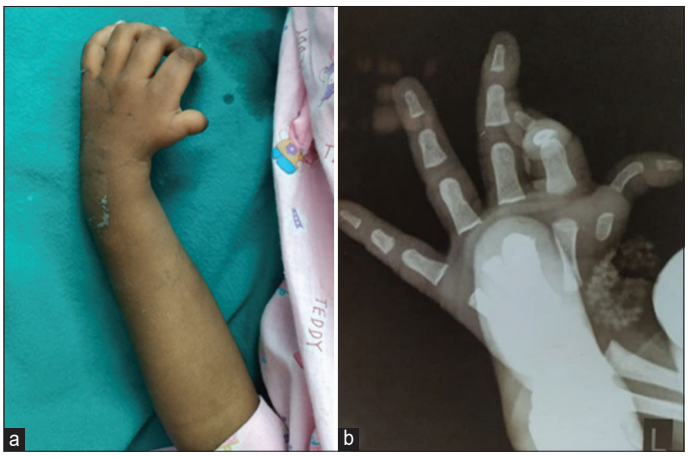

Figure 1: The presence of associated hand anomaly of both hands upon (a) clinical and (b) radiological examinations

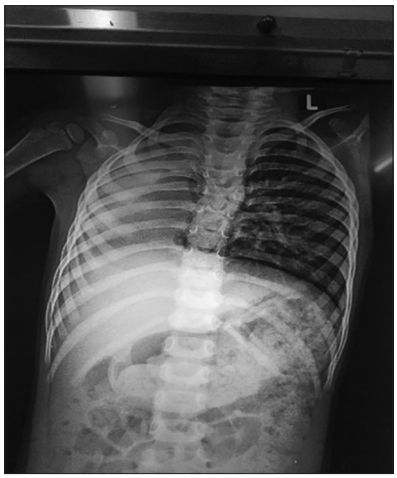

Figure 2: Chest X-ray of the patient showing dextrocardia

DISCUSSION

Situs solitus with dextrocardia is a rare congenital disorder. In situs inversus, the position of the heart chambers, as well as, the visceral organs such as the liver and spleen are reversed, but in situs solitus, only the position of the heart is reversed; however, the abdominal organs are at their respective anatomical position. Although the individual may lead a normal healthy life and its finding is usually incidental. From an anesthetic point of view, it is important to know that situs solitus is usually associated with cardiac abnormalities in almost 90% of patients in contrast to situs inversus which is associated with cardiac abnormalities in only 5–10% of patients [7].

The patient with dextrocardia with situs solitus who may be asymptomatic for the whole of their life, still needs cardiac evaluation preoperatively, and may decompensate intraoperative under the stress of surgery and anesthesia. Hence, careful history regarding the cardiac symptoms is a must and echocardiography and cardiac consultation are required in preoperative time.

Recent genetic studies have suggested that the mutation in the ACVR2B, Pitxz genes node gene, ZIC 3, and the genes existing on chromosome 12 causes the abnormalities in the lefty genes because of the left-right asymmetry defects [8,9]. Situs inversus with dextrocardia may also be associated with Kartagener syndrome [10], so a history of chronic sinusitis and bronchiectasis is a must which may be difficult to elicit in the pediatric age group. This group of patients may land up with pulmonary complications. Hence, pulmonary consultation and optimization including antibiotics, nebulization, bronchodilation, and chest physiotherapy may be required perioperatively.

There have been few case reports available in the literature regarding the successful anesthetic management of such patients. Koc *et al*. reported successful management of emergency anesthetic management of a patient with situs inversus undergoing an emergency appendectomy under general anesthesia. They also emphasized that, although the patient may be asymptomatic preoperatively, they still need extensive preoperative examination, along with a diligent physical examination, cardiological consultation, and the exclusion of probable additional abnormalities [11].

Although the anesthetic technique is not much different, one has to be vigilant while putting monitors such as for dextrocardia, ECG leads, and defibrillator pads must be placed accordingly. Incorrect placed ECG leads may hinder the diagnosis of perioperative ischemia. While intubation, one must be careful about left-sided endobronchial intubation in contrast to right-sided endobronchial intubation in normal individuals. While choosing regional anesthesia, one should be aware of associated spinal canal abnormalities such as spina bifida, split cord myengomyelocoel, and scoliosis [12].

From a surgical point of view, in situs inversus, as anatomy is reversed in the abdomen and thorax, so typical presentation of the surgical conditions may not be there. Instead, patients' clinical symptoms may confuse the clinician and need to be interpreted in terms of CT or ultrasonographic findings. Anatomical positions also change the surgical approach. Hence, one needs to be aware of the type of dextrocardia and its associated anatomical and physiological variations to plan and execute safe and successful anesthesia.

CONCLUSION

Situs solitus is a rare congenital disorder that may be associated with cardiac, pulmonary, and spinal abnormalities and needs careful preoperative evaluation and preparation for the successful conduct of anesthesia.

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Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Kaur H, Singh A, Kumar V, Chaudhry D. Anesthetic management of a pediatric patient with situs solitus and dextrocardia. Indian J Case Reports. 2023;9(5):150-152.