Anesthetic Management in a Case of Situs Inversus Totalis

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ABSTRACT

Situs inversus totalis is a rare syndrome, with an estimated prevalence of 1/10,000 births, characterized by the inverted position of the thoracic and abdominal organs with respect to the sagittal plane. The condition affects all major structures within the thorax and the abdomen. The etiological factors for which are still not completely understood. In a patient with situs inversus totalis, the anesthetic management during the respective surgical procedure is challenging. We are reporting a patient who was incidentally detected with situs inversus totalis with dextrocardia and was operated for Angle’s grade 3 malocclusion. The present case report lays an emphasis on the potential difficulties during anesthetic management and its various implications.

Keywords: Dextrocardia, Kartagener’s syndrome, Situs inversus.

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INTRODUCTION

The absence of heart sounds on auscultation at the precordial area raises the suspicion of one rare condition, dextrocardia, which could have been associated with situs inversus totalis.

During the embryological development, a 270° clockwise rotation instead of normal 270° anticlockwise of the developing thoracoabdominal organs results in mirror image positioning of the abdominal and thoracic viscera. 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 anesthesiologist.

The apex of the heart is located on the right side of the thorax, the stomach, and the spleen on the right hypochondriac region in the abdomen and the large lobe of the liver and gallbladder on the left side. The left lung is tri-lobbed and the right lung bi-lobbed and blood vessels, nerves, lymphatics, and the intestines are also transposed.

This rare condition is well described by a few surgical and medical journals, but the anesthetic implications and considerations have not been thoroughly explained by any anesthesia specialty journal. We are reporting a case of situs inversus totalis who was operated for Angle's grade 3 malocclusion with an aim of discussion of the anesthetic considerations and implications associated with such anatomical abnormalities.

CASE REPORT

• A 20-year-old female patient presented to oral and maxillofacial surgery outpatient department (OPD) with chief complaints of forwardly placed lower front teeth.

• It was only with the help of radiological investigation reports and electrocardiogram that we were able to confirm it as a case of situs inversus totalis. Her chest X-ray (Fig. 1) typically depicted dextrocardia and spiral-computed tomography (CT, Fig. 2) scan showed transposition of all major abdominal organs. Echocardiography was carried out (Fig. 3) and revealed normal cardiac parameters with an ejection fraction of 63%.

• Preanesthetic evaluation revealed mallampati class II patient with a progynathia, a pulse rate of 90/min, and blood pressure of 130/80 mm Hg. Systemic examination was unremarkable.

• Hematological investigations were normal.

Fig. 1: X-ray chest

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1 hour prior to the surgery injection ranitidine 50 mg and injection emeset 4 mg were given intravenously.

- The electrocardiogram (ECG) electrodes have to be applied in the opposite direction as the changed surface electric polarity may give a false picture of perioperative ischemia.
- Other standard monitoring techniques were employed.
- Glycopyrrolate, midazolam, and fentanyl were used for premedication.
- General anesthesia was induced with propofol i.v., fentanyl, and sevoflurane 2%.
- Induction of anesthesia was achieved with O₂, sevoflurane, propofol, fentanyl, and vecuronium.
- Nasal intubation was done using a 7.0-mm cuffed north pole endotracheal tube.
- Intubation was easy and the laryngoscopic view could be best labeled as Cormack–Lehane grade I.
- Bilateral air entry was checked and tube was fixed and maintained with O₂ + N₂O + sevoflurane.
- Injection hydrocortisone 100 mg was given prophylactically to prevent any bronchospasm.
- Maintenance of anesthesia was carried out with divided doses of injection vecuronium and sevoflurane in oxygen and nitrous oxide gas mixture were used.
- Air warmer was used to maintain normothermia.

RESULTS

- Bilateral sagittal split osteotomy: mandibular setback of 7 mm was performed.
- All vital parameters were stable throughout the procedure.
- At the end of surgery, reversal of neuromuscular block was done with inj. neostigmine and inj. glycopyrrolate.
- Patient was extubated on noting adequate ventilatory efforts.
- Room air SPO₂ following extubation was 98% and patient was conscious, oriented, phonating, and moving all four limbs.
- Patient was observed in the postoperative ward for 24 hours with uneventful stay.

DISCUSSION

- Situs inversus totalis is a rare condition in which the organs of the chest and abdomen are arranged in a perfect mirror image reversal of the normal positioning, the etiologic factors for which are still not completely understood.
- The most preferred diagnostic technique involves chest and abdominal skiagrams as well as CT scans.
- The diagnostic parameters of simple skiagrams include the presence of dextrocardia, the stomach bubble under the right dome of diaphragm, and the liver shadow on the left side.
- The association of situs inversus with other syndromes and diseases such as Kartagener’s syndrome, mucociliary dysfunction, and airway anomalies, which may predispose the patient to numerous varieties of airway difficulties and pulmonary infections that can have considerable implications during induction of anesthesia and intubation.
- The syndrome is associated with numerous cardiac anomalies such as atrial septal defects, ventricular septal defects, transposition of great vessels, absent coronary sinus, double-outlet right ventricle, total pulmonary anomalous venous defect, and pulmonary valve stenosis either singly or in combination.
- The ECG electrodes have to be applied in the opposite direction as the changed surface electric polarity may give a false picture of perioperative ischemia.
In case of cardiothoracic surgery, lung separation throws a challenging task due to transposition of thoracic viscera. Insertion of a double-lumen tube will pose a multitude of challenges and the successful intubation and separation of lungs cannot be accomplished without the aid of fibreoptic bronchoscope.

Situs inversus also complicates organ transplantation operation as donor organs will almost certainly come from situs sollicitus (normal), as the heart and the liver have geometric problems while placing the organs into cavity shaped in mirror image. Orientation of these blood vessels also reversed necessitating steps to be taken so that blood vessels join properly.

Situs inversus in Kartagener’s syndrome is invariably associated with mucociliary dysfunction. Primary ciliary dyskinesia is present in 25% of the patients with situs inversus totalis with an increased probability of developing respiratory complications. Therefore, moist and filtered mixture of gases should be administered during mechanical ventilation. The role of bronchodilators, chest physiotherapy, postural drainage, antibiotics, and incentive spirometry cannot be underestimated and is mandatory in optimizing the pulmonary status before any surgical procedure.

In case of cardiac arrhythmias and cardiac arrest, great care has to be taken while applying direct current with defibrillator pads on the right side.

**Conclusion**

The precise diagnosis of situs inversus totalis and a thorough preoperative evaluation can minimize, to a large extent, the difficulties and the various potential challenges associated with its anesthetic management.

Keeping in mind all these considerations and implications, it becomes easier and safer to successfully manage the patients of situs inversus totalis in the operation theater and intensive care units.

**References**
