The challenging aspects and successful anaesthetic management in a case of situs inversus totalis

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ABSTRACT
Situs inversus totalis is a rare condition with a predicted incidence of one in 10,000 among the general population, the aetiologic factors for which are still not completely understood. In a patient with situs inversus totalis, not just the diagnosis of any acute abdomen pathology is difficult due to distorted anatomy and transposition of thoraco-abdominal viscera but equally challenging is the anaesthetic management during the respective surgical procedure. We are reporting a patient who had situs inversus totalis and was operated for open cholecystectomy. The present case report lays an emphasis on the potential difficulties during anaesthetic management and its various implications.

Key words: Cholecystectomy, dextrocardia, Kartagener’s syndrome, situs inversus

INTRODUCTION
The absence of heart sounds on auscultation at the pre-cordial area, confusion during the diagnosis of acute cholecystitis, acute appendicitis and splenetic lesion on the basis of clinical examination raises the suspicion of one rare condition, the situs inversus totalis. This rare condition is characterized by total transposition of thoracic and abdominal viscera, and the predicted incidence is one in 10,000 among the general population.[1-4]

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.[5] 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]

This rare condition is well described by a few surgical and medical journals, but the anaesthetic implications and considerations have not been thoroughly explained by any anaesthesia speciality journal. We are reporting a case of situs inversus totalis who was operated for symptomatic cholelithiasis, with an aim of thorough discussion of the anaesthetic considerations and implications associated with such anatomical abnormalities.

CASE REPORT
A 55-year-old female patient presented to surgical out-patient department (OPD) with chief complaints of pain and left hypochondrium for the last 3 days accompanied by nausea and vomiting. She presented a big dilemma to the attending surgeon as all the clinical signs and symptoms pointed towards symptomatic cholelithiasis, but the pain in the left hypochondrium totally contradicted the diagnosis. It was only with the help of radiological investigation reports that we were able to confirm it as a case of situs inversus totalis. Her chest X-ray typically depicted dextrocardia and spiral computed tomography (CT) scan showed transposition of all major abdominal organs. Echocardiography was carried out and revealed normal cardiac parameters with an ejection fraction of 60%.
Pre-anaesthetic evaluation revealed Mallampatti class II patient with a slight retrognathia, a pulse rate of 74/min, blood pressure of 130/78 mmHg and normal profile of laboratory investigations. She had a history of intermittent respiratory tract infections, especially during the winter season. At present, the patient had no systemic complaint pertaining to any organ system except for the gall bladder disease. A decision to operate the patient was taken and she was posted for open cholecystectomy.

Tablet ranitidine 150 mg and tablet alprazolam 0.25 mg were administered as premedication a night before and 1 h prior to the surgical procedure with a sip of water. In the operation theatre, a good intravenous (IV) access was secured with a wide bore cannula. Induction of anaesthesia was achieved with O₂, isoflurane, propofol 120 mg, butorphanol 1 mg and succinylcholine 100 mg. During induction, we encountered masseter spasm immediately after the administration of succinylcholine, which lasted for approximately 45 s, and disappeared spontaneously, but we were able to ventilate the patient with bag and mask during this episode. Intubation was extremely difficult as the larynx was placed quite anteriorly and the laryngoscopic view could be best labelled as Cormack Lehane grade-III. Maintenance of anaesthesia was carried out with injection vecuronium and low concentration of isoflurane in oxygen and nitrous oxide mixture. The peri-operative period was characterized by frequent copious secretions from the respiratory tract that had to be cleared regularly for smooth ventilation. Inj. Hydrocortisone 100 mg was given prophylactically to prevent any bronchospasm. The surgical procedure was smooth and uneventful, and lasted for about 1 h. At the end of surgery, inj. Neostigmine and glycopyrrolate were administered but extubation was delayed as the patient developed prolonged post-operative apnoea period. Therefore, extubation was performed only after establishing the return of all protective airway reflexes and establishment of regular breathing pattern. Infiltration of surgical incision site was carried out with 8 ml 0.375% of ropivacaine. The recovery period and hospital stay were uneventful and the patient was discharged in a satisfactory condition on the eighth post-operative day and was called for follow-up after 2 weeks.

**DISCUSSION**

Situs inversus totalis is a rare condition, the etiologic factors for which are still not completely understood. There is no established gender or racial difference in its incidence, but genetic predisposition and familial occurrence point towards multiple inheritance patterns.[2-4] The symptomatic pain of cholelithiasis in these anomalies presents in the left hypochondrium and may mimic other acute abdomen presentations such as hiatus hernia, pancreatitis, duodenal perforation, etc. The scenario of clinical diagnostic dilemma was similar in our patient also, for which we had to take assistance of the radiology department. 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, stomach bubble under the right dome of diaphragm, liver shadow on the left side and the findings of barium meal.[7-9]

There was prolonged post-operative apnoea period in our patient. It was only after 30 min of administration of reversal agents that the patient developed a regular breathing pattern with the return of protective airway reflexes that we were able to successfully extubate the patient. Our institute does not have the facilities of diagnosing the atypical cholinesterase levels, and this was a limiting factor in this presentation. Hence, we recommended the patient to get these investigations done from the top-most research centre and to bring the requisite reports during the follow-up visits. A case of prolonged paralysis after administration of succinylcholine has been reported earlier also in a patient with situs inversus totalis.[10]

The challenging aspects for anaesthesiologists in successful management of patients with situs inversus totalis should be thoroughly evaluated, such as:

1. The association of situs inversus with other syndromes and diseases such as Kartagener’s syndrome, mucociliary dysfunction, airway anomalies, etc., which may predispose the patient to numerous varieties of airway difficulties and pulmonary infections that can have considerable implications during induction of anaesthesia and intubation. We encountered masseter spasm immediately after administration of succinylcholine, which was relieved spontaneously in less than 1 min, and there is no evidence in the literature where masseter spasm developed after injection of depolarizing neuromuscular blockers in patients with situs inversus totalis.

2. 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.[11]

3. The spinal deformities like split cord, spina bifida, meningomyelocele, scoliosis, etc. have been described in the literature, and one has to evaluate the patient carefully if any surgery is planned under neuraxial anaesthesia.[12]

4. The ECG electrodes have to be applied in the opposite direction as the changed surface electric polarity may give a false picture of peri-operative ischaemia.

5. 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 fibroptic bronchoscope. The transposition of the thoracic viscera also alters the various anatomical landmarks, and one has to be well acquainted with ultrasound-guided procedures if in case a need arises for invasive central venous cannulation and brachial plexus blockade.

6. 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.[13] Therefore, moist and filtered mixture of gases should be administered during mechanical ventilation. There were plenty of mucoserous secretions during the peri-operative period, even though our patient had no active respiratory tract infection. We administered injection hydrocortisone 100 mg, anticipating bronchospasm due to any subclinical infective pathology. 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.

7. 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. A successful resuscitation of such patients requires a thorough knowledge and skills on the part of the attending anaesthesiologist and intensivists.

From the mentioned implications and considerations in a case of situs inversus totalis, it can be safely established that regional anaesthesia is the preferred choice for any infra-umbilical surgery as compared with administration of general anaesthesia, provided that there is no spinal anomaly or dysraphism.

CONCLUSION

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

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