Safe Anesthetic Management and Anesthetic Considerations in a Patient with Situs Inversus Totalis

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INTRODUCTION
Situs inversus (SI) is an uncommon congenital anomaly characterized by a mirror image orientation of the abdominal and thoracic viscera relative to the midline during the embryological development, a 270-degree clockwise rotation instead of normal 270-degree counterclockwise rotation of the developing thoracoabdominal organs, and if it is associated with right-sided heart (dextrocardia), it is called situs inversus totalis (SIT) (1-6). SI was first described by Fabricus, and SIT was first described by Mathew Baillie (1, 4). Incidence of SIT varies from 1:5000 to 1:20000 births (1,5). Except for positional anomaly, cardiac functions are normal (1). Patients with SIT are asymptomatic and have a normal life expectancy (1, 4, 5). It is usually inherited by autosomal recessive, but sometimes it is inherited by X chromosome (4). Radiography, ultrasonography, and computed tomography can be used for diagnosis, but the most preferred method is computed tomography and it is considered as the best diagnostic method for SIT (1, 3, 4). Diagnostic features in plain chest X-ray include dextrocardia, stomach bubble in right side, and liver shadow on left side (3). SI is one of the components of Kartagener’s syndrome (primary ciliary dyskinesia), which is a combination of SIT, bronchiectasis, male infertility, and sinusitis (1, 2, 4). We report a case of SIT posted for emergency perforation after we obtained patient’s consent from his relatives.

CASE PRESENTATION
A 72-year-old-man with known case of SIT, coronary arterial disease, and lung cancer was presented to emergency operation for ileal perforation. He had been treated by radiotherapy and chemotherapy for lung cancer 5 months ago. He had gastric operation for ulceration 20 years ago. He was under treatment with acetylsalicylic acid and proton pump inhibitor. In the pre-anesthetic evaluation, his Glasgow Coma Scale score was E3M5V4. Physical examination revealed consciousness; tend to fall asleep; closed eyes; normal-sized, normally reactive pupils; apex beat was on the right fifth intercostal space in the midclavicular line; heart was auscultated on the right side of the chest; and lung auscultation was evaluated as decreased bilaterally. He had respiratory failure. Abdominal examination revealed tenderness and rigidity in all four quadrants of his abdomen. His pulse rate was 115 beat/min, blood pressure 110/50 mmHg, and body temperature was 36°C. Chest X-ray (Figure 1) and computed tomography (Figure 2) showed dextrocardia with fundal gas shadow on the right side. Electrocardiography (ECG) showed marked right axis deviation (Figure 3); ECG with reverse lead placement showed left axis deviation, Ashman phenomenon, and pathological q-wave in inferior leads (Figure 4); and 2D ECG confirmed dextrocardia with first-degree aortic and tricuspid failure.
In the operating room, routine monitoring (electrocardiogram, pulse oximeter, noninvasive blood pressure) was attached and a pre-induction arterial line was placed. The electrocardiogram lead placement was reversed because of the patient’s dextrocardia. Anesthesia was induced with pentothal 5 mg/kg, fentanyl 1 µg/kg, and cis-atracurium 0.2 mg/kg uneventfully, and a single lumen endotracheal tube was placed. Anesthesia was maintained with air in O₂ (50%-50%), desflurane (5%-6%); muscle relaxation was maintained with intermittent cis-atracurium boluses. Capnography was also initiated. Because of the history of radiotherapy and chemotherapy, we had difficulty in intravenous cannulation and thus we placed central venous catheter
cautiously. He was hemodynamically stable during the perioperative period. At the end of the surgery, he was transferred to the intensive care unit. Postoperative analgesia was maintained with tramadol three times 75 mg intravenously. On the second day of the operation, he was extubated, and he was discharged from the intensive care unit 5 days after the operation.

DISCUSSION

We report a safe anesthetic management of a patient with SIT for emergency ileal perforation. Our patient had diagnostic findings in plain chest X-ray. ECG with reverse lead placement showed no abnormality except arrhythmia.

Situs inversus totalis does not cause any significant morbidity. Patients with SIT are asymptomatic and have a normal life expectancy (1, 4, 5). But SIT can coexist with other congenital anomalies, such as cardiovascular, respiratory, digestive system, etc. (6). Also, although SIT is well described by a few medical journals, the anesthetic considerations have not been thoroughly explained. By this case, we want to report precautions in the anesthetic management of the cases with SIT. Precautions that should be taken during the anesthetic management of patients with SIT are:

1. Mainstem intubation can occur on left side of the trachea and should be kept in mind while intubating the trachea (1, 4, 5)
2. If central venous catheter cannulation is planned, preference should be given to left internal jugular vein (to avoid thoracic duct and to ensure direct access to right atrium) (1, 3, 5)
3. All invasive lines such as central venous cannulation, arterial cannulation in major artery, should always be US-guided (3, 5, 6)
4. ECG electrodes and defibrillation pads should be placed in reverse orientation (1-6)
5. Kartagener’s syndrome should be considered in these patients (1, 2, 6)
6. Detection and documentation of SI is important to prevent adverse events (1)
7. The intraoperative (6) and postoperative oxygen supplementation should always be humidified especially in cases with Kartagener’s syndrome (2, 3)
8. The patient’s hydration should be well maintained to prevent any inspissation of secretions in the airway and potential incidence of airway compromise (2)
9. Adequate analgesia should be provided (Inadequate analgesia may lead to respiratory insufficiency in the presence of Kartagener’s syndrome) (2)
10. Surgery requires relatively more duration in patients with SIT because tissue handling and orientation were difficult to interpret (2)
11. In prolonged surgery, body temperature should be monitored and normothermia should be actively maintained (2)
12. In thoracic surgery, while choosing a double lumen tube, the anatomy of the bronchi should be considered (6)
13. If Echo/USG is available they may be used briefly to exclude any flaw in a situation of proper Cardiopulmonary Resuscitation (CPR). Don’t hesitate to interrupt, it may save your ineffective CPR time and help in early return of spontaneous circulation (3)
14. Modification in CPR and defibrillation techniques (3, 5, 6)
15. Prolonged paralysis is reported with succinylcholine in SIT (due to decrease in pseudocholinesterase levels) (3, 5, 6)
16. Kartagener’s syndrome is associated with airway anomalies, which may cause difficult laryngoscopy and intubation (3, 4, 6)
17. Since it has no effect on thoracic/respiratory muscles and spontaneous respiration, regional anesthesia is a suitable method when indicated (5). But the spinal deformities such as split cord, spina bifida, meningocele, scoliosis, etc., have been described in the literature. When surgery is planned under regional anesthesia, the patient must be evaluated very carefully (10).
18. Pregnant patient maneuvers should be done considering uterus position. It is possible that uterus is relocated on the right side (5).

Prediagnosed SIT patients should be evaluated and examined very carefully. If any sign of SIT is present in any preoperative imaging, examination, or laboratory tests, the diagnosis should be confirmed or excluded. Although there is no distinctive approach for SIT, monitoring should be done accordingly, and any concomitant disease or possible complications should be under consideration.

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