



Anaesthetic Management for Appendectomy in a Patient with Situs Inversus Totalis

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Situs inversus totalis is a congenital syndrome, in which all the internal organs are in the opposite position, including dextrokardia. Most patients are asymptomatic and maintain their normal life. Kartagener syndrome may accompany situs inversus totalis. Diagnosis may be overlooked in patients with situs inversus totalis in emergency situations. Patients with this syndrome should inform the clinician as this will facilitate the diagnosis. Acute appendicitis is an emergency situation that would require urgent intervention. Appendicitis symptoms can be observed in the left lower quadrant of patients. We present a case concerning the anaesthetic management of a patient with situs inversus undergoing an emergency appendectomy.

Keywords: Situs inversus totalis, kartagener syndrome, acute appendicitis, anaesthesia

Introduction

Situs inversus totalis is the condition where all internal organs are in a mirror image, and this includes dextrocardia. It is known as a hereditary syndrome. The normal order of organs is called situs solitus. Situs inversus totalis is usually an autosomal recessive disorder, and sometimes may be X chromosome-related. It was first described by Matthew Baillie. Although its incidence in the community is low, it was reported as one in 5000–20000 births in the sources (1, 2). Most of the patients are asymptomatic and maintain their normal life. Because Kartagener syndrome may also accompany, it should be kept in mind and be excluded with a carefully-taken medical history. Kartagener syndrome is an autosomal recessive syndrome that occurs with the trilogy of dextrocardia, sinusitis and bronchiectasis. Its frequency is between 1: 30000 and 1: 40000 (3).

The diagnosis can be difficult to make in situs inversus totalis patients in emergency cases. Informing the patient about his/her own situation and his/her informing the clinician about it will make the diagnosis easier. Otherwise, it will be difficult to make the diagnosis because of the reversed positions of the organs. This is especially true in many emergencies, including acute myocardial infarction, because of the increased mortality. Documenting situs inversus totalis in a person is important to interpret the signs of the disease that may occur as well as to avoid any adverse clinical and surgical complications.

Acute appendicitis is the first among the situations in terms of admitting to the emergency department and requiring immediate intervention. The Incidence is between 4% and 8% (4). Symptoms of appendicitis, which appear in the right in normal patients, may be seen in the lower left quadrant of patients with situs inversus totalis, and the diagnosis can be difficult. In this case report, we aim to explain the anaesthesia management in a patient with situs inversus totalis, in whom an emergency appendectomy was performed.

Case Presentation

Pain and sensitivity in the left lower quadrant was detected in the examination of a 46-year-old female patient that was brought to the emergency department with the complaint of abdominal pain. Dextrocardia was detected on a chest radiograph (Figure 1).

The view resembling the appendix in the abdominal ultrasonography and the liver on the left side resulted in the suspicion of situs inversus totalis. This diagnosis was also evident in the history of the patient. Acute appendicitis localized in the left was confirmed using computed tomography, and the decision for surgery was made by general surgeon. In the pre-anaesthesia evaluation of the patient, she was found to be asymptomatic in her normal life. She gave a history of intermittent hypertension, but she did not use the prescribed drugs properly. She was not followed up at any clinic. There were no abnormalities in routine laboratory analysis. Consultation was held with the cardiology department about the patient's condition, and no pathology was encountered except for the dextrocardia. Ejection fraction (EF) was found to be 65% on echocardiography. Kartagener syndrome was not suspected in the patient because she did not give any history of sinusitis and bronchiectasis. Pregnancy was eliminated with beta HCG and ultrasound examinations. General anaesthesia was planned for the patient. Informed written consent was received after information was provided about the surgery and anaesthesia. The patient was taken to the operating room. Non-invasive arterial blood pressure (BP), cardiac apex beat (CAB), peripheral oxygen saturation (SpO₂) and electrocardiography (ECG) monitoring was performed. ECG electrode location was determined by paying attention to the dextrocardia in monitoring. The fluid of 0.9% NaCl 1000 mL was administered intravenously. Preoperative BP was measured as 167/87 mm Hg, CAB as 126 min⁻¹ and SpO₂ as 98%. After the patient was sedated with 1 mg of midazolam, 75 mcg of fentanyl was administered. Then, thiopental 7 mg kg⁻¹ was intravenously administered as a hypnotic. In the 1st minute of the induction, BP was measured as 153/82 mm Hg, CAB as 104 min⁻¹ and SpO₂ as 100%. When it was observed that there was no issue with mask ventilation, rocuronium 0.6 mg kg⁻¹ was administered. Intubation was performed with an endotracheal tube with a 7.0-mm internal diameter. In verification of the intubation,



Figure 1. Patient's chest X-ray obtained in the emergency service

it was observed that the left lung was ventilated more when the tube was inserted forward; equal ventilation of both lungs was achieved by pulling back the tube a bit. Immediately after the intubation, BP was measured as 186/95 mm Hg, CAB as 119 min⁻¹ and SpO₂ as 100%. Anaesthesia was maintained with a mixture of 50% O₂ + 50% nitrous oxide (N₂O) at a 4 L min⁻¹ flow and 2% of sevoflurane (sevorane). In the perioperative 10th minute, BP was 116/55, CAB was 71/min and SpO₂ was 100%. Intraoperative and postoperative vital signs were stable. Because there wasn't any problem in the postoperative follow-up, the patient was delivered to inpatient service. The patient with no problems in follow-up was discharged without any complication on day the 2nd of admission.

Discussion

Situs inversus is a condition that occurs as a mirror image of the chest and abdominal organs. Situs inversus totalis is a rare autosomal recessively inherited syndrome, and there is no difference among races in terms of incidence. Distribution is the same for both sexes. Anatomical asymmetry in human and vertebrates occurs during embryogenesis. It is determined at the beginning of embryonic development with the formation of the left-right, dorsoventral and cephalocaudal axes. The cardiac tube curve to the right is the first sign of asymmetry. The left-right gradient has been established at cellular level. The left-right relationship of asymmetric internal organs is preserved and is known as situs solitus. The displacement of the organs as a mirror image is known as situs inversus totalis (5, 6). Recent studies suggest that the mutation in the node gene, ZIC 3, ACVR2B, Pitx2 genes and the genes existing on chromosome 12 causes the abnormalities in the lefty genes because of left-right asymmetry defects (7, 8).

The recognition of situs inversus and knowing the reverse anatomy are important to avoid complications in cases such as surgical gallstones or appendicitis. The diagnosis can be made easily through radiography and ultrasonography. However, computed tomography is a preferred diagnostic tool. Usually, situs inversus patients are asymptomatic and have a normal life expectancy (9).

Acute appendicitis is one of the most common situations occurring in the emergency department and requires immediate initiative. The Incidence is between 4% and 8% (10). The most common symptom in the acute appendicitis is pain in the right lower abdomen. Therefore, acute appendicitis is not a typical differential diagnosis in patients with left lower quadrant pain. However, in patients with situs inversus, left lower quadrant pain may be a symptom of appendicitis and if the diagnosis is delayed, the appendix perforation may occur.

The evaluation for anaesthesia in these patients should be performed together with the consideration of existing symptoms and findings. Cardiology consultation, ECG and

echocardiography performed may give us information in asymptomatic patients. Chronic sinusitis and bronchiectasis should be queried in history, and Kartagener syndrome should be excluded. It should be supported by extra pulmonary function testing and chest diseases consultation if necessary. Although there is not a different approach in the administration of anaesthesia, special care should be paid to monitoring and ECG electrodes should be placed in accordance with dextrocardia. In addition, the evaluation for Mallampati should be done in terms of difficulty with intubation and the pre-mental level should be well considered. In the control of equal ventilation of the lungs after intubation, after further insertion of the tube, it should be kept in mind that the left lung will be well ventilated, not the right one. Because there were not any cardiac symptoms and abnormal findings in our case, we did not change the anaesthetic management. The patient was delivered to the inpatient department without any surgical or anaesthetic complications.

Conclusion

This case report has demonstrated the difficulty of diagnosis in situs inversus totalis patients who require emergency surgery and the importance of informing the patients about their condition. In the management of anaesthesia in such cases, the history of the patient should be taken carefully in preoperative examination, along with a diligent physical examination. The evaluation with cardiological consultation and the exclusion of probable additional diseases are important. Patients should be queried for features of Kartagener syndrome. Pulmonary function test, chest CT and chest diseases consultation may provide additional information. In our case, the patient was asymptomatic. In addition, she didn't have a history of ongoing respiratory problems and chronic sinusitis since childhood. Routine procedures of general anaesthesia were administered in the patient and a different drug was not administered.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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