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CASE REPORT

Management of Acute Appendicitis and Left Paraovarian Cyst in a Case of Situs Inversus Totalis by Laparoscopy

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ABSTRACT

Introduction: Situs inversus totalis (SIT) is an entity in which there is transposition of both the abdominal and thoracic organs. Presentation of acute abdomen in a case of SIT poses a challenge to the treating surgeon. We present a rare case report in which we identify the role of laparoscopy in confirming acute appendicitis with a simultaneous left paraovarian hemorrhagic cyst as a cause of left iliac fossa pain.

Case report: A 17-year-old female presented with pain in the left lower abdominal quadrant. Ultrasonography and computed tomography confirmed a left-sided inflamed appendix and a left paraovarian hemorrhagic cyst along with transposition of other organs. A diagnostic laparoscopy was done to confirm the diagnosis with subsequent appendectomy and cyst enucleation as a definitive treatment for left iliac fossa pain.

Discussion: Management of acute abdomen in a case of SIT can be challenging, keeping in mind the transposed organs and also that the nervous supply may still be normal in up to 50% of the cases. The role of diagnostic laparoscopy is pertinent in arriving at diagnosing and treatment of the underlying pathology or pathologies as in our case.

Keywords: Abdominal pain, Appendicitis, Paraovarian cyst, Situs inversus totalis.

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INTRODUCTION

Situs inversus (SI) is a rare congenital developmental anomaly occurring with an incidence of 1:5,000 to 1:10,000 live births in which abdominal organs are placed as mirror image of each other.[1] There is no racial predilection, and sex incidence is 1:1. Situs inversus totalis (SIT) is a condition in which both thoracic and abdominal organs are transposed. The incidence of SIT is approximately 1/1,400 to 1/35,000 in the general population. Acute abdomen in SIT possesses a big diagnostic dilemma as though the viscus is transposed, but their nervous innervation may follow the normal original distribution as in 50% of the individuals. The incidence of acute appendicitis associated with SIT is reported to be between 0.016 and 0.024%.[2,3] Laparoscopy is a valuable tool in surgeon's hand in case of SIT with acute abdomen, which provides confirmatory and definitive surgery of involved pathology.[4]

To the best of our knowledge, there are no reported cases of acute abdomen due to acute appendicitis associated with left paraovarian hemorrhagic cyst in patients with SIT.

CASE REPORT

A 17-year-old female presented in the casualty ward with complaints of left lower quadrant nonradiating pain for 1 day. The patient had history of nausea, but no fever, vomiting, or diarrhea. There was no history of associated abdominal or any previous similar attack of pain in abdomen. The patient had a normal menstrual history with no comorbid conditions like tuberculosis, diabetes, etc. She was hemodynamically stable and on physical examination revealed tenderness in the left iliac fossa with localized guarding. Laboratory investigations revealed a raised total leukocyte count of 13,500 cells/mL, with a differential neutrophil count of 85% of the total cells. The liver function tests and renal function tests were within the normal limits. A plain chest radiograph showed the presence of dextrocardia with a gastric bubble on the right side. Ultrasonography (USG) revealed a tubular, aperistaltic structure in the left iliac fossa with a diameter of 9 mm along with a paraovarian cyst of size 8 × 12 mm and transposition of the abdominal organs. The abdominal computed tomography (CT) revealed the presence of a contrast-enhanced appendix with the maximum diameter of 9.5 mm and adjacent fat stranding along with a left paraovarian cyst of diameter 9 × 12 mm (Fig. 1) in size with transposed intraabdominal organs (Fig. 2). A diagnostic laparoscopy was planned in view of localized guarding and raised white blood cell count. A 10 mm umbilical port was placed with open technique. The second 10 mm working port was placed

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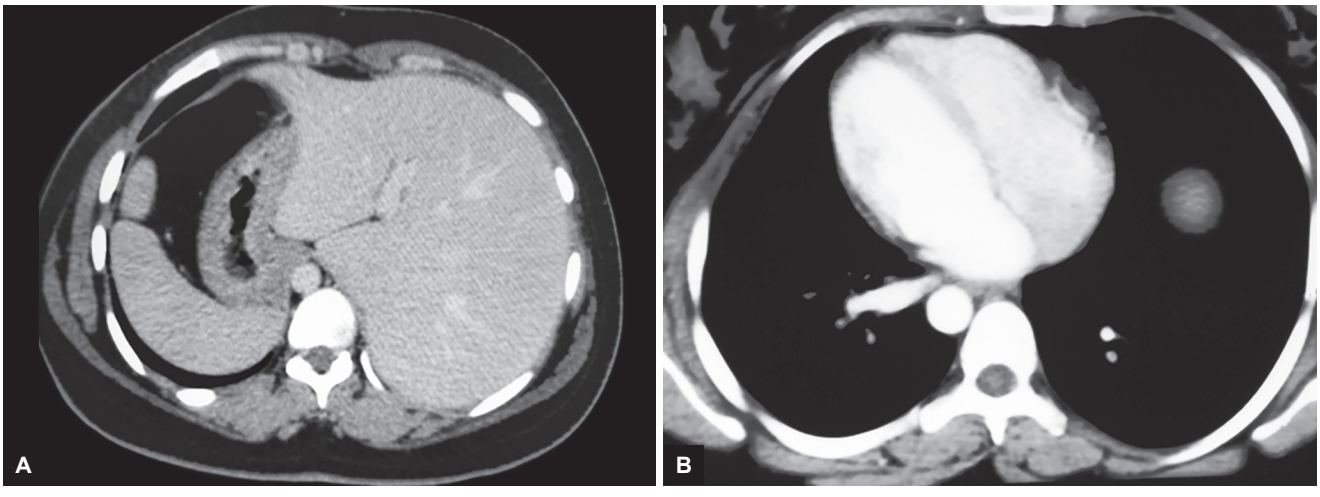
Fig. 1: Computed tomography scan with a left-sided inflamed appendix

in right iliac fossa on the midclavicular line and the third 5 mm trocar with the working port was placed in the suprapubic region. The operative findings showed an inflamed turgid appendix with a normal base (Fig. 3). There was also a paraovarian cyst present along the left ovary with minimal free fluid collection in the pouch of Douglas (Fig. 4). Transposition of the various abdominal organs was also confirmed (Fig. 5). A laparoscopic appendectomy with enucleation of paraovarian cyst was done. An inflamed appendix with inflammatory infiltrates was confirmed in the histopathology report. Postoperative recovery was uneventful with patient discharged on full diet on 4th postoperative day without any complications.

DISCUSSION

Matthew Baillie[5] first demonstrated the complete mirror image, reversal of thoracic and abdominal organs in SI in the 18th century. It is a rare congenital anomaly where abdominal organs are placed as a mirror image of each other. In SIT, both abdominal and thoracic organs are transposed. The SI results due to incomplete penetration



Figs 2A and B: Computed tomography scan with transposition of the visceral abdominal organs: (A) A dextrocardia and (B) confirming SIT

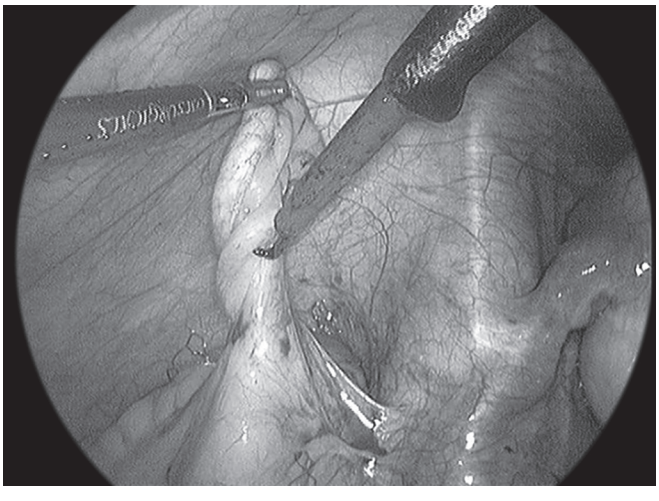


Fig. 3: Diagnostic laparoscopy showing an inflamed left-sided appendix with a normal base

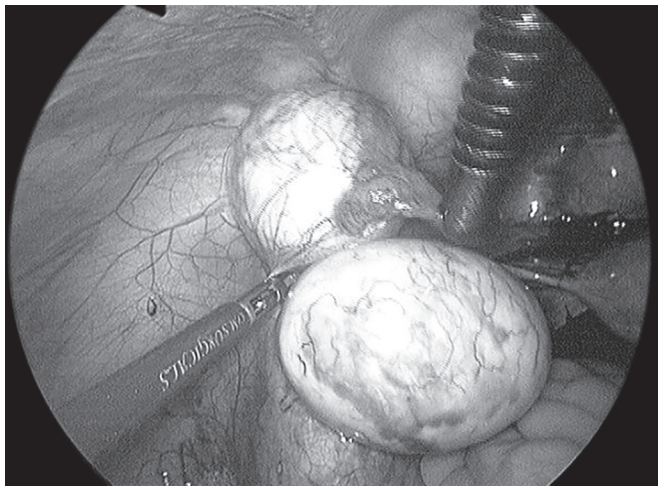
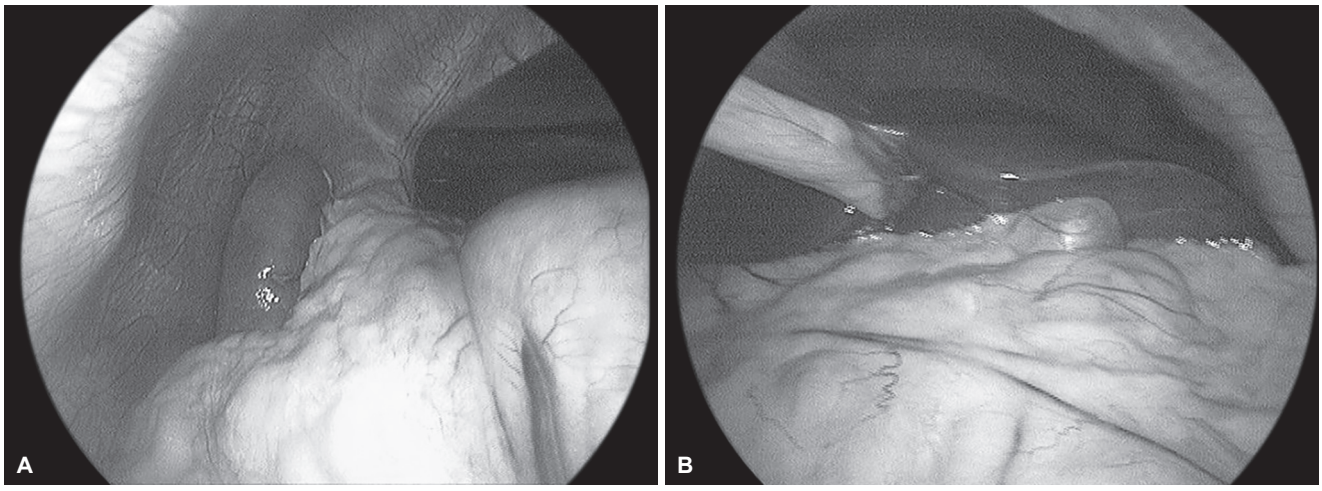


Fig. 4: Left-sided paraovarian cyst during diagnostic laparoscopy



Figs 5A and B: Transposed abdominal organs with spleen in the right hypochondrium: (A) Liver along the left hypochondrium; and (B) during diagnostic laparoscopy

of single autosomal recessive gene located on the long arm of chromosome 14.[6] Acute abdomen in SI is a major clinical challenge for surgeons for diagnosing and treating the pathology. In fact, this is the reason why in 40% of cases, the incision for surgical pathology is planned inappropriately.[7,8]

The differential diagnosis of left lower quadrant pain in SIT patients includes diverticulitis, epididymitis, incarcerated or strangulated hernia, bowel obstruction, regional enteritis, psoas abscess, mesenteric ischemia, right- and left-sided acute appendicitis (LSAA), pelvic inflammatory disease, endometriosis (in females), and others.[9,10]

The diagnosis of acute appendicitis in patients with SIT can be based on clinical examination, blood investigations, X-rays, USG, electrocardiogram, barium studies, CT scan, and diagnostic laparoscopy. However, in SIT with multiple pathologies, cause of symptoms is difficult to correlate and in these circumstances, the laparoscopy offers a big role in diagnosis as well as definite treatment of pathology. Even if the surgery is not possible to be completed laparoscopically due to any reason, it allows the incision to be properly placed at the site of pathology, as in about 18.4 to 31% of patients with SIT, the pain caused by LSAA has been replaced in right lower quadrant. Incision has been reported as inappropriate sites in greater than 40% of cases.[7,8]

Regarding the pain location of LSAA, Akbulut et al[11] reported that 62% of patients presented with left lower quadrant pain, 14% with right lower quadrant pain, and 7% with bilateral pain. Because the nervous system may not show corresponding transposition, pain location may be confusing, so preoperative diagnosis has been made only in 51% of patients.

The authors believe that any SIT patients who present with lower abdominal pain, especially female, should undergo diagnostic laparoscopy as many gynecological pathologies may also mimic appendicitis. Removing the appendix even if appearing normal may eliminate any possibility of misdiagnosis in future. It will also exclude the risk of complications that come with delayed diagnosis, such as appendiceal rupture, which can be fatal in young female patients.[12]

There is no standard port position for laparoscopic appendectomy in SIT patients, and it is placed according to basic principles of laparoscopy and ergonomics.[13]

CONCLUSION

With the case report, the authors wish to suggest that in patients of SI with acute abdomen, the laparoscopy is an important tool in surgeons' hand in diagnosing as well as in doing definitive management of patient. Even in the circumstances of normal looking appendix, it is better to do appendectomy to avoid future misdiagnosis and decrease morbidity associated with the same.

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