

Acute Appendicitis In Situs Inversus Totalis : Report of two cases

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[Received : 22/02/2014, Revised : 20/03/2014, Accepted : 23/04/2014]

Abstract :

Situs inversus totalis is a relatively rare congenital anatomic anomaly in which there is complete right to left reversal (transposition) of the thoracic and abdominal organs. The incidence of acute appendicitis with situs inversus totalis is between 0.016% and 0.024%. We present two cases of situs inversus totalis with acute appendicitis. Appendectomy was performed on both patients as an emergency procedure. Postoperative recovery was uneventful in both of the patients and were free of their symptoms in their follow up period. Acute appendicitis in situs inversus is a rare association that can present a diagnostic problem. Morphologic exploration methods such as ultrasonography, computed tomography, magnetic resonance imaging, and laparoscopy may contribute to the early management of the disease and give guidance in choosing the most appropriate treatment for patients.

Introduction:

Situs inversus is a congenital anomaly characterized by the transposition of the abdominal viscera. It may or may not be associated with dextrocardia, also known as situs inversus totalis^{1,2} Generally, this rare genetic anomaly is discovered incidentally, often when a radiographic assessment of a patient is undertaken, particularly to investigate an abdominal infection. We report two cases of situs inversus discovered while evaluating acute pain in left iliac fossa and diagnosed to be acute appendicitis. This is particularly interesting because of the scarcity of this association and the diagnostic difficulties that may arise because of unusual site of pain.

Case Reports

CASE 1 : A 22 year-old female patient presented to our hospital with 2-days history of acute abdominal pain in the left iliac region. This pain was associated with fever. She had a fever of 100°F, a pulse rate of 100/minute and blood pressure of 120/70 mm Hg. Her physical examination revealed abdominal tenderness predominantly over left iliac fossa.

Sonography of abdomen and lower chest revealed a situs inversus totalis with dextrocardia with acute appendicitis. An X-ray chest confirmed dextrocardia. CT abdomen confirmed findings of situs inversus totalis with acute appendicitis.

Laboratory investigations showed a white blood cell count of 13,900/mm³ with 93% neutrophils, 42% hematocrit, and platelets at 323,000/mm³.

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Patient was taken up for an emergency surgery. Exploration showed inflamed oedematous left sided appendix. Appendectomy was performed. Histopathology confirmed acute appendicitis. No post-operative complication was noted and she was discharged home five days after her operation.

CASE 2 : A 17 year old male patient presented to our hospital with one day history of acute abdominal pain in the left iliac region. This pain was associated with nausea and vomiting. He was afebrile, had a pulse rate of 90/minute and blood pressure of 100/70 mm Hg. His physical examination revealed abdominal tenderness predominantly over left iliac fossa.



Figure 1-Peroperative view of left sided caecum and appendix.

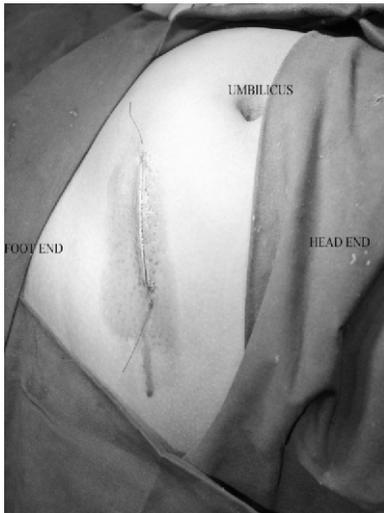


Figure 2-Sutured site.

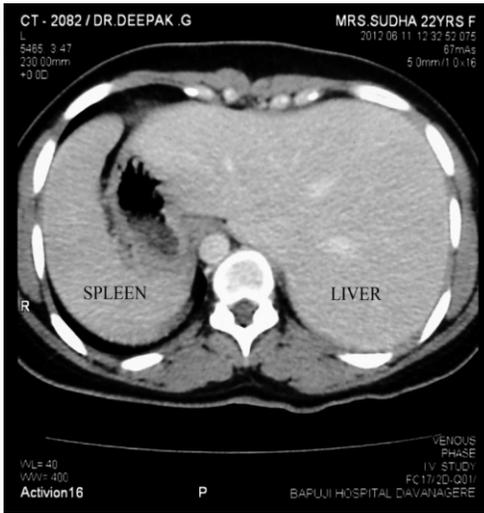


Figure 3-CT abdomen showing left sided liver and right-sided spleen.

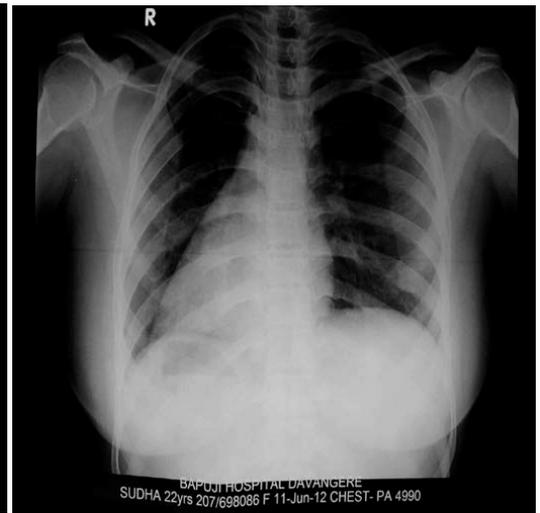


Figure 4-Chest x-ray showing right sided cardiac shadow.

Patient was given antibiotics and analgesics and ornidazole, but there was no improvement.

The patient was further investigated with sonography of abdomen and lower chest which revealed a situs inversus totalis with dextrocardia with Acute Appendicitis. An X-ray chest confirmed dextrocardia. CT abdomen confirmed findings of situs inversus totalis with acute appendicitis.

Laboratory investigations showed a white blood cell count of $11,200/\text{mm}^3$ with 88% neutrophils, 42% hematocrit, and platelets at $3,43,000/\text{mm}^3$.

Patient was taken up for an emergency surgery. Exploration showed inflamed oedematous left sided appendix. Appendectomy was performed. Histopathology confirmed acute appendicitis. No post-operative complication was noted and he was discharged home five days after his operation.

Discussion

Situs inversus is a positional anomaly that rotates the abdominal internal viscera. It is known as situs inversus totalis when it is associated with a transposition of the thoracic organs. Situs inversus is a rare congenital anomaly with an incidence in the population of only 0.001% to 0.01%^{1,2} with a male-to-female ratio of 3:2³ Its transmission mode is autosomal recessive, but its precise genetic mechanism has yet to be identified.^{1,3}

Situs inversus results from a rotation in the opposite direction of the viscera during the development of the embryo.^{2,3} Patients with situs inversus may face diagnostic problems because of the unusual localizations of their symptoms.

In the case of our patients, pain was in the left iliac fossa, hence the diagnosis was difficult. Even in patients without situs inversus, the right iliac appendicular symptoms would be found in only 60% of cases.^{1,3} The

presence of symptoms in the left iliac fossa in the absence of situs inversus may be due to an abnormally long appendix projected to the left, or to intestinal hyperkinesis.

A study of 71,000 patients, with appendicular symptoms,



Figure 5-Chest x-ray showing right sided cardiac shadow.

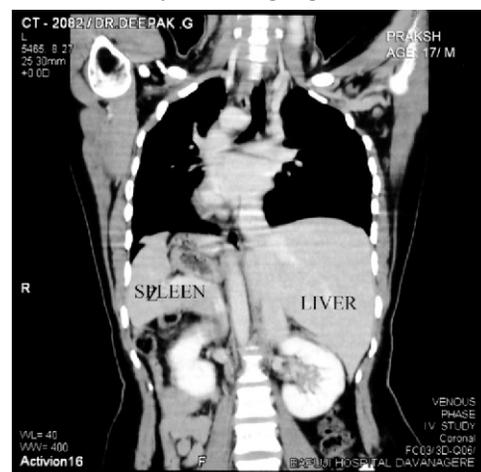


Figure 6-Left-sided liver and right-sided spleen.

found that 0.04% of cases involved left iliac localization, comprising 0.024% with abdominal situs inversus and 0.016% with situs inversus totalis^{3,4} Until 2008, fewer than 10 cases of appendicitis associated with situs inversus were reported in the literature³ Half of these patients reported pain in their right iliac fossa despite the presence of situs inversus¹ Therefore, given the scarcity of this association, the diagnosis of appendicitis with situs inversus is not automatically evoked, which delays the appropriate management of patients. As a consequence, peritoneal diffusion may eventually develop.

Meanwhile, the usual differential diagnosis of left lower quadrant abdominal pain in an adult man includes, among others, sigmoid diverticulitis, epididymitis, bowel obstruction, psoas abscess, and in this rare instance, situs inversus with acute appendicitis. Medical imaging can help clinicians to arrive at a correct diagnosis. Abdominal X-ray, ultrasonography, and CT abdomen can facilitate an accurate and early diagnosis if a patient is unaware of this positional anomaly.^{1,3,4} Medical imaging can also guide the appropriate therapeutic choice, surgical indication, and type and location of the incision.⁴ The contribution of laparoscopy is undeniably useful in these situations, as it

favors a minimally invasive surgical approach in diagnostics and treatment.⁵

Conclusion

The occurrence of acute appendicitis with situs inversus is very rare. Very few cases have been reported in the literature. This condition poses a diagnostic problem that can be decreased by ultrasonography and laparoscopy. These procedures allow the early management of the disease and guide therapeutic choices.

Consent

Written informed consent was obtained from our patients for publication of this case report and any accompanying images.

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How to Cite this article :

Udapudi D G, Karne R. Acute Appendicitis In Situs Inversus Totalis : Report of two cases. *J Pub Health Med Res*, 2014;2(1):41-3.

Funding: Declared none

Conflict of interest: Declared none