

Left-Sided Acute Appendicitis With Situs Inversus Totalis In A Nigerian Male – A Case Report And Review Of Literature

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Abstract

Situs inversus is a rare congenital abnormality which occurs in 1:20,000 of the general population. Left-sided acute appendicitis is associated with two types of congenital abnormalities, situs inversus and malrotation. This condition is often diagnosed incidentally while investigating or treating a patient for some other conditions as in our index patient.

We present a case report of a 22-year-old Nigerian male who was admitted through the emergency unit of a military hospital in South Nigeria on account of an acute abdomen. A diagnosis of acute appendicitis was made after clinical evaluation but at surgery, the inflamed appendix and cecum were found in the left iliac fossa. At the time of his presentation, an abdominal ultrasound scan could not be done for logistic reasons. However, an abdominal ultrasound scan and a chest X-ray done after surgery confirmed situs inversus totalis.

This case is presented to highlight the rarity of this condition (the first reported case in Calabar) and the need to do an abdominal ultrasound scan routinely in cases of acute abdomen where indicated to avoid making a wrong incision at surgery with its attendant morbidity and poor cosmesis. Laparoscopic surgery, where possible, is of immense benefit in this condition.

INTRODUCTION

Situs inversus was first described in the 16th century by Mathew Baillie¹. Situs inversus (also called situs transversus or oppositus¹) is a rare congenital anomaly characterized by the transposition of the abdominal viscera. When associated with dextrocardia, it is referred to as situs inversus totalis². It has an autosomal recessive pattern of inheritance with an incidence of 1:20,000^{1,3} in the general population.

It is termed situs inversus with dextrocardia (situs inversus totalis) if the heart is swapped to the right side of the thorax or situs inversus with levocardia (situs inversus incompletus) if the heart remains in the normal left side of the thorax (a much rarer condition). Left-sided appendicitis occurs in two types of congenital anomalies namely situs inversus and intestinal malrotation³. The situs anomalies are rare and often unrecognized until they are incidentally detected during imaging for other conditions, emergency surgeries (as in the index case) or during laparoscopy⁴.

CASE REPORT

A 22-year-old Nigerian male presented to the emergency unit of the Nigerian Navy Hospital, Calabar, Cross River State, Nigeria with a 6-hour history of severe right-sided colicky abdominal pain associated with nausea but no vomiting or fever. He was previously unaware of his situs anomaly. Two years earlier, he had a similar episode of abdominal pain which was managed conservatively. At presentation, he was acutely ill-looking, afebrile (temperature 36.6°C), with positive pointing sign and maximal tenderness at McBurney's point. His packed cell volume was 44% and total white blood cell count was 5.1 x 10⁹/L with a neutrophil count of 50%. Urinalysis was normal. A diagnosis of acute appendicitis was made and he was booked for emergency appendectomy.

Surgery was commenced with a Lanz incision over the right iliac fossa but this incision was abandoned when the cecum and appendix were not found in the right iliac fossa. A mid-line sub-umbilical laparotomy incision was then made and a normal looking cecum found in the left iliac fossa with an

inflamed retrocecal appendix bound down by fibrous adhesions (figures 1-3).

Figure 1

shows the cecum and the appendix in the left iliac fossa.



Figure 2

shows the left-sided appendix after adhesiolysis.

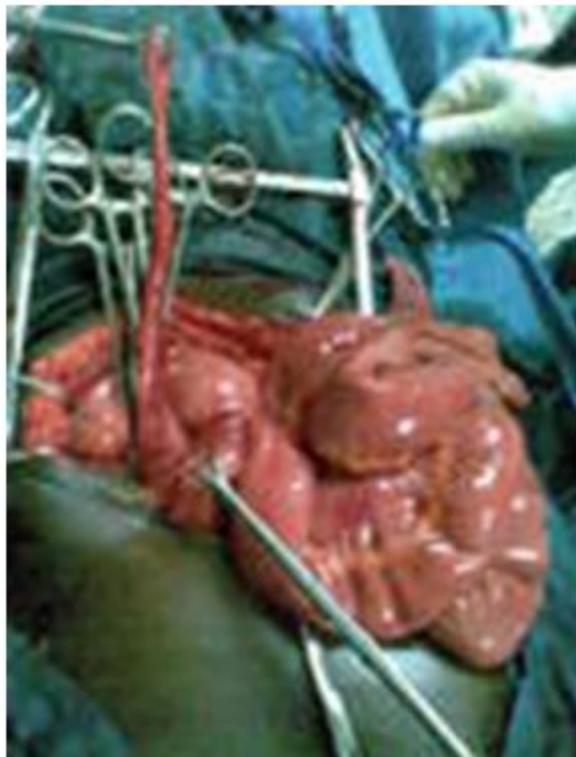


Figure 3

shows the initial Lanz incision and the subsequent midline subumbilical incision.



Figure 4

Chest X-ray showing dextrocardia



Appendicectomy was done after careful adhesiolysis. The patient had an unremarkable post-operative recovery. A post-operative abdominal ultrasound scan done by a consultant radiologist confirmed situs inversus totalis. Also a plain chest radiograph and an ECG, done post-operatively, confirmed dextrocardia (figure 4). Histology of the appendix confirmed inflammation of the appendix with obstruction of the lumen by fecolith.

DISCUSSION

Situs anomalies are rare and their incidence, as documented in the literature, varies from 0.001 to 0.01% in the general population⁴. Diagnosis of a left-sided acute appendicitis in these patients is rather challenging due to altered anatomy¹⁻⁵.

Acute appendicitis is the commonest surgical emergency the world over⁶⁻⁹ with an annual incidence of 1:1000 of the population. The classical presentation occurs in only 60% of patients and comprises vague peri-umbilical pain (which later localizes in the right lower quadrant of the abdomen) with associated nausea, vomiting and anorexia, amongst other symptoms. This is the first documented case of situs inversus in the literature from this environment, hence the need to draw attention to this rare condition.

Diagnosis of acute appendicitis in situs inversus totalis is often difficult because of abnormal pain localization³. It has been noted that although the viscera are transposed, it is thought that the central nervous system may not share the reverse transposition thus leading to confusing symptoms of signs in these patients. Thus, the pain and rebound tenderness of left-sided acute appendicitis is reported to be on the right iliac fossa in about 31-50% of patients with situs inversus totalis³⁻⁵ (as in our index patient). This has led to an incorrect incision in 45% of cases and in a third a second correct incision had to be made like in our index patient³. In their case report, Ucar et al.³ made a correct initial incision because the diagnosis of situs inversus totalis and left-sided acute appendicitis was made using abdominal ultrasound scan and chest radiograph before surgery, whereas Golash⁴ established the diagnosis incidentally at laparoscopy in a patient with suspected diverticulitis and localized peritonitis. In Turkey, Fuat et al.¹⁰ also reported a pre-operative diagnosis of acute appendicitis in situs inversus based on the clinical finding of a right-sided heart confirmed radiologically (dextroposition of the heart and gastric fundus). Though the diagnosis of acute appendicitis is largely clinical, the place of abdominal ultrasound scan in diagnosis of acute abdomen cannot be overemphasized.^{6,7} In our index patient, a correct incision may have been made at the onset if situs inversus had been confirmed by an abdominal ultrasound scan pre-operatively. We strongly recommend that abdominal ultrasound scan should be done for suspected acute appendicitis as this may reveal the existence of this condition and guide surgery accordingly. In centers where laparoscopic surgery is possible, it can be used as the preferred approach. Laparoscopy, apart from helping to confirm the situs inversus at surgery, also provides a better cosmetic outcome in addition to the reduced morbidity it offers.

It is also important to educate patients with this condition once they have been diagnosed and advise them to volunteer this information to the doctor whenever the need arises to minimize the risk of misdiagnosis and treatment. Some authorities advocate that these patients should carry a name tag to this effect¹.

Finally, situs inversus has significant implications on organ transplantation since such organs may be from situs solitus

(normal) donors because of geometric problems¹.

CONCLUSION

Left-sided acute appendicitis is a rare but important condition and is usually associated with situs inversus. Diagnosis is often incidental and the use of abdominal ultrasound scan, which is non-invasive, is advocated routinely in patients suspected to have acute appendicitis as this will demonstrate the situs abnormality and guide the surgeon accordingly. Laparoscopy is of immense benefit where facilities and expertise exist. These patients should be made aware of their abnormal anatomy and advised to volunteer it subsequently to any physician as this will aid diagnosis and treatment in future.

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