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An unusual presentation of left sided ruptured appendix in a child with situs inversus totalis

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Introduction

Left sided appendicitis is a rare phenomenon which may occur due to two main types of congenital abnormalities; situs inversus totalis (SIT) and midgut malrotation (1). It is widely accepted that SIT occurs due to ciliary dyskinesia, whereas midgut malrotation occurs due to abnormal development of the primitive gut, specifically due to abnormal re-entry of physiologically herniated bowel loop during early embryogenesis (1). The incidence of acute appendicitis associated with SIT is estimated to be 0.016% to 0.024% in the general population and the knowledge regarding specific aetiology and predisposing factors is limited (1).

Situs inversus totalis (SIT) is a rare (1 in 10,000 live births) autosomal recessive disorder characterised by transposition of visceral organs, including the appendix, to the contralateral side (2). Therefore, visceral pathologies present with atypical clinical manifestations. Furthermore, the nerve supply to the transposed organs may be the same as in normal development, adding further dilemma to the clinical diagnosis (1). There is a lack of knowledge in the neural pathways of the transposed organs and further, the molecular regulation of brain asymmetry and its role in visceral lateralization needs to be investigated.

We report a child with previously undiagnosed SIT presenting with dysuria and lower abdominal pain and later diagnosed with a ruptured left sided appendix.

Case presentation

A 10-year-old girl with a history of congenital heart disease presented with fever, loss of appetite, severe diffuse lower abdominal pain, and dysuria for 2 days duration. There was no history of consanguinity of the parents, exposure to teratogens in the intra-uterine life nor family history of congenital heart disease. She had no dysmorphic features and had a normal development. On examination, she was ill-looking and the abdominal examination revealed diffuse lower abdominal tenderness with guarding of bilateral lower quadrants. There was no hepatosplenomegaly, umbilical hernia or free fluid in the abdomen. Her pulse rate was 120 beats per minutes with low volume and blood pressure was 90/60 mmHg. There was no radio-radial or radio-femoral delay. Examination of the chest revealed normal heart sounds and a palpable apex on the right side (4^{th} intercostal space, midclavicular line) without any associated murmurs. There were no precordial deformities. Her previous records revealed a perimembranous ventricular septal defect with dextrocardia and she was on follow up with annual echocardiography. She was not worked up for situs inversus as she had normal development and bowel function.

Her inflammatory markers were elevated (white cell count: 11,000/microlitre with neutrophil predominance and C-reactive protein: 110 mg/L). Her urinalysis was normal. Therefore, based on the clinical and biochemical findings, a diagnosis of acute appendicitis with possible perforation was

made and antibiotics were commenced (Intravenous cefuroxime 750 mg twice daily). A complicated urinary tract infection was entertained as a differential diagnosis. Chest-X ray showed dextrocardia with bowel shadows on the right upper quadrant (Figure 1). Abdominal ultrasonography was performed before surgery which revealed SIT with right lobe of the liver on the left hypochondrium, spleen on the right hypochondrium and inflamed appendix in the left iliac fossa.

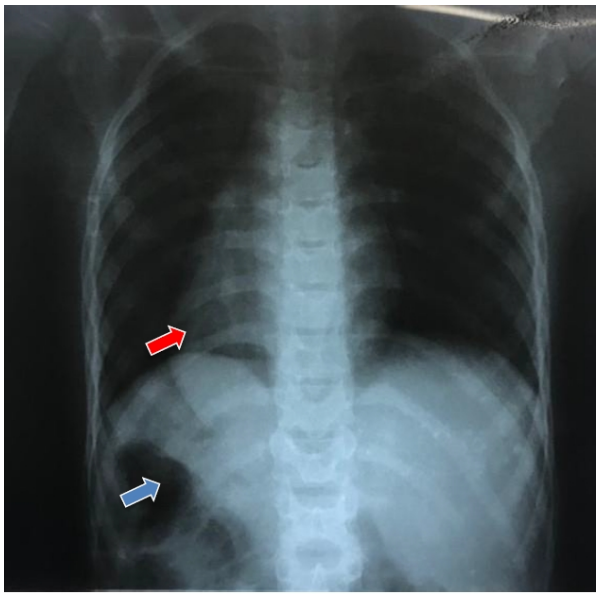

Figure 1: Chest-X ray showing dextrocardia (red-arrow) with bowel shadows (blue arrow) on the right upper quadrant

Surgery was performed via a lower midline laparotomy due to the clinical suspicion of perforation. A mirror image of the usual approach was performed. Intra-operative findings included a grossly inflamed appendix with a perforated tip and omental adhesions (Figure 1). The appendix was identified easily as it was located in the pre-ileal position with the tip attached to the omentum and was the mirror image of the normal anatomy. Appendicectomy was done and intravenous antibiotics were continued for 3 days. Her post-operative period was uneventful and was discharged on day 3 with oral antibiotics.

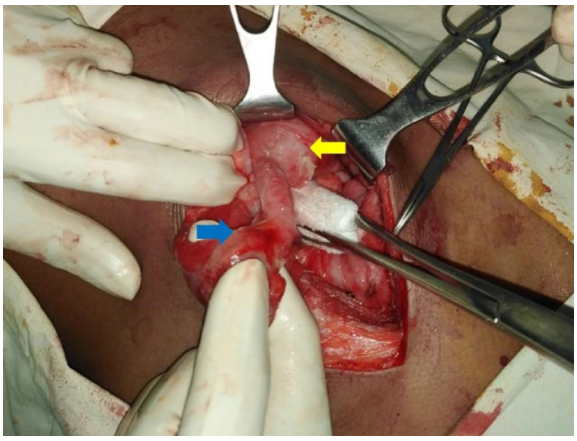

Figure 2: Intra-operative image. Caecum in the left iliac fossa (yellow arrow). Inflamed appendix with a perforated tip and omental adhesions (blue arrow).

Discussion

Our patient was diagnosed with dextrocardia but she was not investigated for transposed viscera prior to current presentation. Majority of patients with dextrocardia have isolated transposition of the heart to the right side while a minority may have associated transposition of visceral organs (2). This could have been a reason for not investigating her for transposition of viscera in her previous presentations. She had not had any other complaints which suggests transposition of other viscera in any of the previous visits.

The presentation of left-sided appendicitis is often diverse and atypical. Presentations may include left (62%), right (15%) or even bilateral lower quadrant pain (7%) as in our patient (1). Further, the pain can rarely originate in the pelvis (2%), left-upper quadrant (7%) or umbilical regions (6%) (1). Whereas, when the appendix is right sided, the findings are usually periumbilical pain that later shifts to right lower quadrant with associated local tenderness, rebound tenderness and guarding. Our patient presented with diffuse lower abdominal pain with guarding of both lower quadrants. Furthermore, the presence of dysuria added to the clinical dilemma as a complicated urinary tract infection which does not usually need surgical intervention was entertained as a differential diagnosis. However, the abdominal

signs with elevated inflammatory markers were very suggestive of acute appendicitis. Imaging was a key investigation before surgery which confirmed SIT and was suggestive of acute appendicitis.

The management of left sided appendicitis is essentially similar to conventional appendicitis. Although both laparoscopy and open techniques have been utilised in left sided appendicectomy, we chose to proceed with a lower midline laparotomy (,). A strong clinical suspicion of a ruptured appendix, atypical anatomy in SIT, lack of paediatric laparoscopic instruments and a small made abdomen were reasons for choosing the open approach. We used a lower midline laparotomy instead of the Lanz or Gridiron incision due to the same reasons.

There are several learning points in this case study. The atypical presentation with dysuria and lower abdominal pain with guarding of bilateral lower quadrants which is highly suspicious of an abnormality. Examination for evidence of dextrocardia should be performed routinely, especially in a patient with congenital heart disease as parents may be unaware of the diagnosis of dextrocardia. Although, clinical diagnosis of acute appendicitis is certain, imaging is extremely important to look for SIT in a patient with dextrocardia. This is helpful for the diagnosis and pre-operative planning.

Conclusions

We report a child with an unusual presentation of left sided ruptured appendix in the background of SIT. It is paramount to look for dextrocardia in all patients, specifically with a background of congenital heart disease. Routine imaging is imperative in a SIT patient with dextrocardia both for the diagnosis and planning of therapeutic interventions.

Informed written consent was obtained from the mother for the publication of the case report with photographs.

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