CASE REPORT

Left-Sided Appendicitis in a 14-Year Girl with Midgut Malrotation

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ABSTRACT

We present a case of 14-year girl with left-sided acute appendicitis who presented with lower abdominal pain. Midgut malrotation occurs at a rate of 1 in 500 live births. The condition is incidentally diagnosed during various radiological investigations done for other purposes. However, such patients may present with conditions like acute appendicitis, which poses a diagnostic dilemma if a high index of suspicion is not kept. The purpose of this case report is to increase awareness in the emergency physicians and young surgeons of this rare presentation; and the importance of radiological investigations in the diagnosis of left-sided appendicitis, to decrease morbidity and mortality.

Key Words: *Appendicitis. Malrotation. Midgut congenital.*

INTRODUCTION

Acute appendicitis is the most common surgical emergency presenting as 'acute abdomen' in the emergency department in patients less than 50 years, with a peak incidence in the second and third decades of life.[1] Acute appendicitis is mainly diagnosed on the basis of history, clinical evaluation, and physical examination. Despite the extraordinary advances and development of newer modalities of investigation, the accurate diagnosis of acute appendicitis remains an enigma. The diagnostic error rate ranges from 12% to 23% in men, and 24% to 42% in women.[2]

Left-sided appendicitis poses an even greater diagnostic conundrum. There are about 97 reported cases of left-sided acute appendicitis as of December 2015. Left-sided location of the appendix occurs in two conditions, i.e. midgut malrotation or in Situs Inversus Totalis.[3] Midgut malrotation is a rare congenital anomaly resulting from incomplete rotation or failure of midgut rotation and fixation, which mostly presents with bowel obstruction or volvulus in infancy with rare occurrence after first year of life.[4]

We present a case of a 14-year girl, with previously undiagnosed midgut malrotation with a left-sided acute appendicitis, with the aim of highlighting the diagnostic challenges faced.

CASE REPORT

A 14-year girl was brought into Accident and Emergency Department, Military Hospital, Rawalpindi in October 2016 with a history of lower abdominal pain for the last four days, and multiple episodes of vomiting, fever and anorexia for the past 24 hours. The pain was of gradual onset, involving the lower abdomen, more in the left iliac fossa than the right. She had low grade continuous fever for the past one day and skipped her breakfast due to anorexia. There were no urinary or gynecological complaints.

She was treated on the lines of urinary tract infection at a nearby hospital, but her symptoms gradually worsened. An ultrasound of abdomen at another setup was suggestive of acute appendicitis, after which the father of the girl brought her to Military Hospital, Rawalpindi.

Her past history revealed that she was a diagnosed case of Tetralogy of Fallot and underwent surgery (Rastelli's Repair) at Armed Forces Institute of Cardiology in June 2015. She had been taking tablet Warfarin 5mg, and Tablet Loprin 75mg, once daily since January 2016. She had stopped taking medicines two days back on her own. Her childhood immunisations were upto date. Family history was insignificant.

On examination, she was a young girl with a sick look, conscious, cooperative and well orientated in time, place and person with a pulse of 120 beats/min, temperature of 100°F, blood pressure of 115/70 mmHg and respiratory rate of 24 breaths/min. The oxygen saturation on room air was 89%. On abdominal examination, it was found distended with tenderness in both lower quadrants, left more than right. Rebound tenderness was positive in both iliac fossae with guarding in lower abdomen. Shunt murmur was auscultated on cardiovascular examination. Respiratory, central nervous system and genital examinations were normal.

Her baseline investigations were requested, and she was kept nil per oral. Her blood complete picture revealed a white cell count (WCC) of 17,400/mm3, hemoglobin of 12.8 g/L, and platelet count of 285 × 109/L. Urine routine examination, liver and renal function

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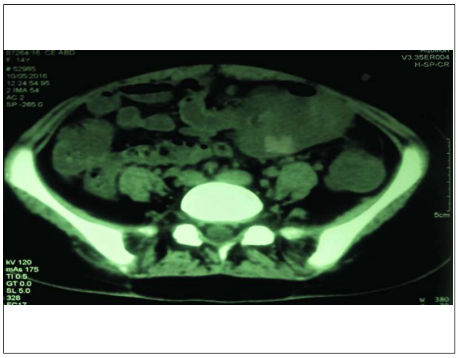

Figure 1: CT scan image of the patient showing left-sided caecum and appendix.

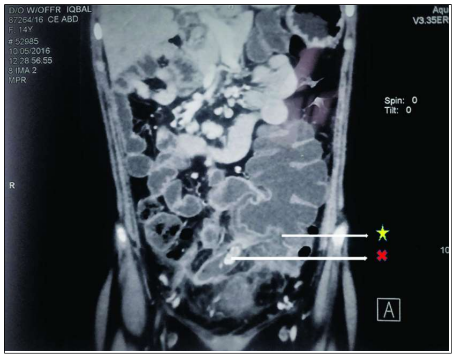

Figure 2: CT scan of the patient.
Yellow star = Left-sided caecum.
Red cross = Appendix with appendicolith.

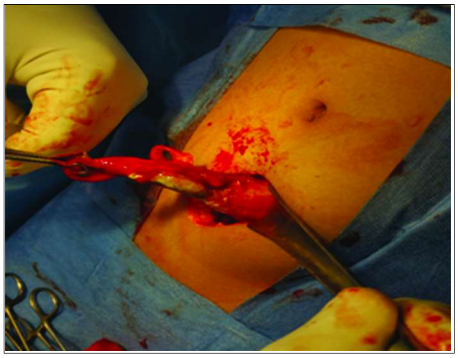

Figure 3: Peroperative picture of perforated inflamed appendix.

tests were within normal range. Her prothrombin time (PT) was 15 seconds and activated partial thromboplastin time (aPPT) was 32 seconds with international normalised ratio (INR) of 1.1. Repeat ultrasound abdomen was done which suggested appendicolith left with free fluid in right iliac fossa. Both kidneys appeared normal. Her contrast-enhanced computerised tomography (CECT) scan abdomen was done which revealed midgut malrotation with features of left-sided appendicitis with an appendicolith at the base of inflamed left-sided appendix. The inflamed appendix was found to be extending from left iliac fossa to midline with adjacent mesenteric fat stranding (Figures 1 and 2).

She underwent lower midline laparotomy for the signs of developing perotinitis. Intraoperatively, there was an inflamed appendix with omentum adherent to it. Her cecum was present in the left iliac fossa with a long appendix, which was perforated near the base extending from the left iliac fossa to the midline (Figure 3). Appendicectomy was done. Postoperatively, recovery was uneventful. Histopathology of the appendix revealed perforated acute suppurative appendicitis.

DISCUSSION

Visceral transposition is a well-established autosomal recessive disorder. It can coexist with various other congenital cardiovascular, respiratory, and digestive tract anomalies. The examples of cardiovascular anomalies include ventricular septal defect, atrial septal defect, Tetralogy of Fallot and transposition of great arteries. The congenital digestive tract anomalies linked with visceral transposition include anal atresia, duodenal stenosis, absent appendix, and megacolon. It can also occur with paranasal sinus deformities.[5]

There are two anatomical abnormalities associated with left-sided acute appendicitis, i.e. Situs Inversus, and midgut malrotation, the latter being less common. During the normal embryological development, the gut herniates out from the abdominal cavity, where it undergoes a 270° counter-clockwise rotation around the superior mesenteric artery. Following the rotation, the bowels return into the abdominal cavity, with fixation of the duodenojejunal loop in the left upper quadrant and the cecum with appendix in the right lower quadrant. Intestinal malrotation is the term used to describe a spectrum of congenital positional anomalies of the intestine, caused by non-rotation or incomplete rotation of the primitive loop around the axis of the superior mesenteric artery during fetal life. The incidence of midgut malrotation varies from 0.03% to 0.5% live births.[6]

Due to the rarity of cases and ambiguity of presenting complaints, the diagnosis of a left-sided appendicitis is challenging and delayed. This child presented with abdominal pain and vomiting. She was of school-going age; and the initial presumption of a gastroenteritis is a sound differential. Another element that delayed diagnosis was the decision to proceed with a repeat ultrasound abdomen instead of a CT scan during the second day of admission. Radiation was limited to a low dose CT scan in this patient. Imaging studies are important to diagnose atypical cases, such as this one, and it is pertinent to know the options available.[7]

Radiological imaging especially CT scan, plays a vital role in the evaluation and management of patients presenting with non-traumatic acute abdomen. CECT scan has a positive predictive value of 95% for diagnosing acute appendicitis. The most specific sign of acute appendicitis on CT examination is a dilated, fluid-filled tubular structure that measures more than 6 mm in diameter, with a thickened enhancing wall in the right lower quadrant, the expected location of the appendix or in rare cases in the left lower quadrant as was present in our patient. Calcified appendicoliths and mesenteric fat stranding are useful secondary imaging findings.[8]

The most extensive study on left-sided acute appendicitis was published in 2010 by Akbulut *et al*. which reviewed 64 case reports of 95 cases.[6] Incidence of acute appendicitis associated with midgut malrotation was reported at 24.2%, and incidence with situs inversus totalis was 69.4%. The study concluded that because the appendix is located abnormally, acute appendicitis

represents a diagnostic dilemma and that ultrasound and CT can provide useful information.[6] These differentials can be differentiated from the likely cause with careful history and physical examination, along with biochemical and imaging investigations. Laparoscopy could not be done in our case as it is not available in our setup in emergency.

Left-sided acute appendicitis is a diagnostic dilemma, which can lead to management delays causing morbidity or mortality in otherwise a completely curable pathology. A high index of suspicion, alertness and cautiousness by the medical team in the diagnostic approach of these patients, especially in children having history of other congenital anomalies, is vital for clinching the diagnosis. Mandatory tools for an accurate diagnosis include thorough medical history, detailed physical examination, laboratory and imaging investigation including ultrasound and CT scan, balancing the risk-benefit-ratio of radiation exposure, especially in pediatric cases. Laparoscopy can be both diagnostic and therapeutic in such difficult cases.

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