

Acute Appendicitis in Situs Inversus Totalis: A Case Report and Review of Literature

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ABSTRACT

Acute appendicitis is the most common surgical condition seen in the emergency unit. In most cases, the diagnosis is straightforward, but sometimes it is not easy as it may be presented in an atypical manner like unusual pain in the left iliac fossa. A 10-years-old boy was presented with an acute abdomen started in the periumbilical area and thereafter, shifted to the left iliac fossa. Chest X-ray and abdominal CT scan was confirmed the diagnosis of acute appendicitis with situs inversus totalis (SIT). Conventional appendectomy was performed with a good outcome. In addition to the detailed history and proper examination, a high index of suspicion and radiological investigations are mandatory to diagnose acute appendicitis in subjects with SIT.

Keywords: Situs inversus totalis; Acute abdomen; Acute appendicitis; Left-sided acute appendicitis.

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INTRODUCTION

Acute appendicitis is the most frequent disease requiring urgent surgery in the world [1]. It could be defined as an acute inflammation of the appendix due to obstructive or non-obstructive factors [2]. It occurs particularly in younger age groups, with a risk of 8.6% in females and 6.7% in males. While appendectomy have performed in 25% of females and 12% of males [3]. Despite an improvement in the diagnostic tools and management of this disease, there are considerable cases that end with postoperative complications (10%) and even death (1–5%) [4]. The rate of these problems will be increased if the condition is not treated within the appropriate time, which is most likely to happen in subjects with atypical presentation. The diagnosis of these conditions depends on the clinical features, radiological imaging, and experience of the dealing surgeon [5].

Situs inversus totalis (SIT) can be defined as a rare autosomal recessive disease characterized by transposition of the organs of the chest and abdomen [6]. Despite the differential diagnosis of acute appendicitis is a huge list, but they are well known to surgeons. However, certain uncommon conditions

may create a wrong diagnosis, like midgut malrotation and SIT. Besides, these may delay the time of starting treatment with uneventful complications owing to their physical findings are obscure [5]. Laparoscopic surgery can be safely used in such a difficult situation to reach an accurate diagnosis. It decreases the chance of delaying diagnosis and it is appropriate for each patient especially in the hand of an expert surgeon who can use the trocars to reach various parts particularly when there is an anatomical variation [7].

This case presentation will discuss the diagnosis and treatment process of the 10-year-old boy with SIT who was diagnosed with acute appendicitis.

CASE PRESENTATION

A 10-years-old boy was presented to the emergency unit of Al-Muqdadhiya general hospital, Al Muqdadhiya city, Diala, Iraq. The case was suffered of acute abdomen with sudden onset of abdominal ache located around the umbilicus and then settled in the left iliac fossa associated with nausea, vomiting, and fever. Physical examination was revealed that the pulse rate was 110 beats/minute, body temperature 38.2°C, and tenderness and rebound tenderness maximum at the left lower abdomen. The patient was admitted to the surgical ward with nil by mouth, intravenous fluids, and antibiotics. Laboratory tests were showed leukocytosis (white cells count 18000/mm³) and C-reactive protein titer 96 in addition of

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chest X-ray that showed dextrocardia as shown in Figure 1. Ultrasound examination of the abdomen was showed an echogenic lesion located at the left lower abdomen with 7 mm in diameter. Computerized tomography (CT) scan of the abdomen was performed revealed situs inversus with features of left-sided acute appendicitis as shown in Figure 2.

Owing to the lack of full equipment for laparoscopic surgery, a conventional appendectomy was performed through a left grid-iron incision. The boy was discharged on the 2nd postoperative day without complications. Informed consent was taken from his father for the publication of this case.

DISCUSSION

A 60% of patients with acute appendicitis are usually presented with sudden onset of pain originating from the umbilical region, which later migrates to the lower right quadrant, loss of appetite, nausea, vomiting, constipation, and sometimes diarrhea [8]. Around 1/3rd of subjects with acute appendicitis have presented with acute abdominal pain in an atypical site due to different anatomical variation in the position of the appendix [9, 10]. However, this is not constant, such as pelvic, subcecal, retrocecal, postilial, preilial, and in rare occasions, subhepatic, mesocolic, left-sided, long right-sided appendix extends into the left lower quadrant region [11]. Left-sided acute appendicitis mimics various conditions that causing left iliac fossa pain like diverticular disease, acute pancreatitis, mesenteric ischemia, and left-sided primary epiploic appendagitis. In addition to gastrointestinal conditions, genitourinary diseases like ectopic pregnancy, ovarian torsion, pelvic inflammatory disease, cystitis, epididymitis, prostatitis, and testicular torsion could cause acute left-sided lower abdominal pain [5, 12, 13]. Therefore, great care is of the utmost importance when dealing with left lower quadrant acute abdominal pain to prevent serious complications as a result



Figure 1. Shows dextrocardia with the gastric gas shadow at the right side.

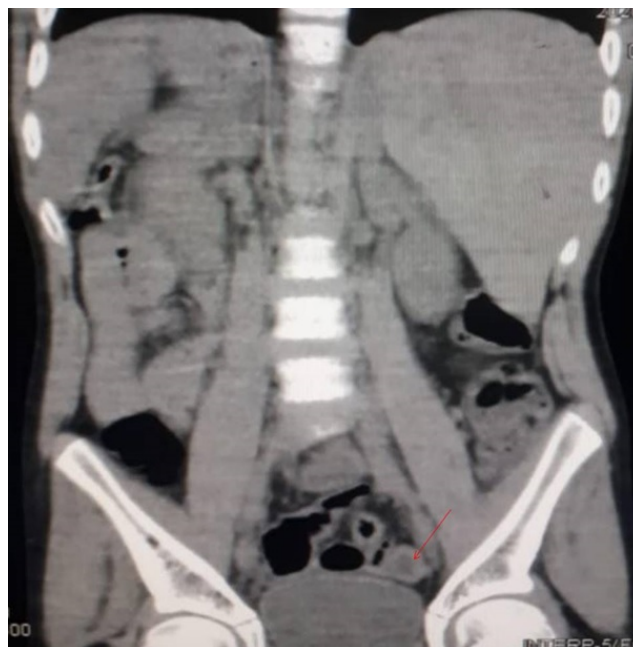


Figure 2. Coronal CT scan of the abdomen showed transposition of the abdominal organs and inflamed appendix (red arrow).

of delayed diagnosis.

Situs inversus is a rare autosomal recessive disease characterized by transposition of the internal organs, with a prevalence rate of 0.01% in the USA population [14]. There are two types either total (both chest and abdominal organs are affected) or partial (chest or abdominal organs are involved) [9]. Our patient had a SIT as proved by the radiological investigations.

Our case was presented with classical acute appendicitis as a sudden abdominal pain which was periumbilical with radiation to the left iliac fossa and the maximum tenderness was in the left iliac fossa. Therefore, an accurate diagnosis was performed by CT scan of the abdomen and chest X-ray. The advantages of the chest X-ray in the case of the acute abdomen are the exclusion of the hollow organs perforation and to rule out respiratory causes. Besides, it was the key to the diagnosis of our case. Despite, some investigations have conclude that chest X-rays in the majority of patients with acute abdomen are of no benefit, the presenting case was diagnosed depends on chest X-ray [15]. Furthermore, a study by Singla et al. in 2015, have reported a rare case of a 73-year-old gentleman diagnosed with acute appendicitis, axial and coronal CT scan of the abdomen and pelvis confirms the diagnosis of asymptomatic midgut malrotation. The study have concluded that radiological imaging is of benefit for early diagnosis and definitive treatment [16]. Moreover, in around 31% of cases with acute appendicitis in the left side, there are still the initial features of pain and tenderness and rebound tenderness in the right side. This is even to the transposition of the abdominal organs, the innervation remains the same. Therefore, imaging techniques can avoid false incisions in 45% of the patients [17, 18].

A high index of suspicion of the situs inversus and midgut malformations should be included in the differential diagnosis

of acute pain with atypical localization, which is suspicious of an acute appendicitis [9, 19]. The high rate of suspicion and thorough assessment are necessary for the diagnosis in subjects with acute abdominal pain and SIT in the emergency unit. In addition, electrocardiography and radiological imaging are important too.

Acute appendicitis is seldom to see in SIT patients with an incidence of 0.016%–0.024% [5, 18]. Laparoscopic surgery is the gold standard technique for the removal of the vermiform appendix in patients with acute appendicitis. It has many benefits in comparison with conventional appendectomies such as a quick healing process, shorter admission period, less operative stress and complications [20]. Therefore, this technique is of utmost importance in conditions when the clinical and imaging investigations are not helpful in the diagnosis of acute appendicitis. The technique can visualize the whole abdominal cavity in comparison to the conventional appendectomy through a McBurney's incision that allows only limited access [11]. In addition to the confirmation of the suspected cases of acute appendicitis, laparoscopy can iden-

tify other abnormalities and treat them simultaneously with the removal of the appendix [20]. Unfortunately, due to the unavailability of the laparoscopy tools in our hospital, open appendectomy was performed in the presenting case and without postoperative complications. Therefore, we can conclude that preoperative confirmation of the diagnosis is necessary for subjects with acute appendicitis with an atypical presentation to achieve a good outcome even treated with conventional appendectomy.

In conclusion, acute appendicitis might be presented in non-classical form, as in SIT and congenital intestinal malformations like midgut malrotation. A thorough history, proper physical examination, and a high index of suspicion are very important in the diagnosis. Furthermore, X-ray of chest, ultrasound, and abdominal CT scan are also invaluable assisting tools for final diagnosis.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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