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Case Report

A 25-year-old male with appendicular agenesis: A case report and literature review

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لملخص

إن عدم تكون الزائدة الدودية منذ الولادة خلقيا أو عدم وجودها هو نادر للغاية. تعرض هذه الحالة مريضا ذكرا عمره ٢٥ عاما ظهرت عليه أعراض وعلامات التهاب الزائدة الدودية الحاد. تعذر العثور على الزائدة الدودية على الرغم من عملية الاستكشاف الجراحي الواسع. ولم تظهر فحوص مابعد الجراحة شيئا غير طبيعي، وتم تشخيص المريض كحالة آلام في البطن غير محددة وعدم تخلق الزائدة الدودية.

الكلمات المفتاحية: آلام البطن غير المحددة؛ عدم تخلق الزائدة الدودية؛ آلام البطن؛ الزائدة الدودية؛ ألم الحفرة الحرقفية اليمنى

Abstract

Congenital agenesis or absence of vermiform appendix is extremely rare. This case report entails a 25-year-old male who developed symptoms and signs of acute appendicitis. Despite an extensive surgical exploration, the vermiform appendix could not be found. The postoperative investigation did not reveal any abnormality, and the patient was diagnosed as a case of nonspecific abdominal pain (NSAP) and appendicular agenesis.

Keywords: Abdominal pain; Appendicular agenesis; Nonspecific abdominal pain; Right iliac fossa pain; Vermiform appendix

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Introduction

Absent vermiform appendix is a very rare finding.[1] Worldwide, the incidence of absent vermiform appendix is 1:100,000 surgical cases for suspected acute appendicitis.[2] Symptoms and signs that can clearly signal the diagnosis of suspected agenesis of the vermiform appendix are absent. Vermiform appendix has no definitive essential functions in the regulation of body functions that can be analysed in serum. The diagnosis of acute appendicitis largely relies upon clinical presentation and the physician's judgement. This case report aims to illustrate a patient who presented with clinical features of acute appendicitis, but the surgical exploration could not identify the vermiform appendix. Further postoperative imaging endorsed the operative finding of appendicular agenesis.

Case report

A 25-year-old male Pakistani labourer working in Rafha, a town on the Northern border of KSA, presented to the emergency room with acute colicky abdominal pain for a duration of 8 h. This pain shifted to the right iliac fossa and was associated with nausea and a single episode of vomiting. The patient denied a past history of abdominal surgery. On examination, his pulse was 88 beats/min, and his temperature was 38 °C. He experienced tenderness in the right iliac fossa without guarding, but rebound tenderness was present. The Rovsing's sign, obturator sign and psoas sign were negative. The white blood cell count was 7.6×10^3 mcL, and the neutrophil count was 85% (shift to left). Complete urine analysis was normal. The Alvarado score for diagnosis of acute appendicitis was 7, which strongly favoured surgical intervention. The patient was diagnosed as a case of acute appendicitis and was explored through a Grid Iron incision by the surgical specialist. Initial exploration did not reveal

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the appendix, which necessitated involvement of two senior consultants who extended the incision for a wider surgical field. A careful search for a sub-serosal, sub-hepatic appendix and Meckel's diverticulum was performed, but still the appendix was not visualized. Interestingly, enlarged mesenteric lymphadenopathy was not observed. A perioperative view of the surgical field is shown in Figure 1.

The abdomen was closed, and the patient was provisionally diagnosed as a case of genesis or congenital absence of vermiform appendix with NSAP. Postoperatively, the patient was managed with antibiotics and analgesics, and his postoperative recovery was smooth. A CT scan with IV and oral contrast on the 3rd postoperative day did not show any abdominal abnormality. The patient was discharged on the 4th postoperative day in stable condition. The patient was followed up for one year with examination and ultrasound every three month postoperatively. The patient remained symptom free during this follow-up, and he was finally diagnosed as a case of congenital agenesis or absent vermiform appendix and NSAP.

Discussion

Embryologically, the vermiform appendix is a diverticulum of the caecum that does not keep pace with the growth of the caecum and takes the shape of an elongated tubular structure with a blind distal end. The appendicular tip varies in position,[3] and its base is almost fixed at the junction of three taenia coli of the caecum. The range of positions of the tip of the appendix described in the literature are as follows: retrocaecal (approximately 38%), retrocolic (26%), subcaecal (14%), pelvic (8%) and preileal (3%).[4] Before diagnosing agenesis or absent appendix, it is imperative to understand that the vermiform appendix is a vestigial remnant that varies in size from 2 cm to 20 cm and, in very rare cases, the appendicular tip may be found embedded inside the lumen of the caecum, often referred to as intussusception of the vermiform appendix. The appendicular agenesis is presumed to be the result of intrauterine vascular accidents, auto amputations due to fibrous bands and appendicular atresia.[5]

The congenital absence of the appendix was reported by Morgagni in 1718,[6] and is well documented earlier in worldwide laparotomies for acute appendicitis. The absent appendix rate is 1:100,000[2] cases.

Being a blind structure and established lymphocytic organ, the appendix may be plugged by faecal matter or by proliferation or hyperplasia of submucosal lymphatic aggregates, which may lead to obstruction of the lumen of the vermiform appendix. This event may compromise its blood supply and may jeopardize the venous return. Thus, a vicious cascade of inflammation, necrosis, gangrene and perforation may ensue. The diagnosis of acute appendicitis is based largely on clinical observation as it is well described by the Alvarado scoring system.[7] This scoring system has a total score of 10; one mark each is assigned to shifting abdominal pain in the right iliac fossa, loss of appetite (anorexia), nausea or vomiting, rebound tenderness, temperature 37.3 °C or more and shift to left (neutrophilia), and two marks each are assigned for tenderness in the right iliac fossa and white blood cell count of 10,000 mcL or more. An Alvarado score of 7 or higher carries 78% sensitivity and 100% specificity.[8] Based on this score, the majority of surgeons recommend appendectomy especially in men without any further investigation, but females may require ultrasonography for exclusion of gynaecological diseases. Patients with a score of 5 or 6 may benefit from other modes of investigation for the diagnosis of acute appendicitis, such as ultrasound of the abdomen, CT Scan,[9] MRI and laparoscopy. In the described case report, an Alvarado score of 7 urged and signalled a straightforward decision for appendectomy. However, a negative exploration revealed a limitation of the Alvarado scoring system.

Appendectomy is the gold standard and definitive treatment for acute appendicitis. Recently, a popular trend is to treat acute appendicitis by conservative management with antibiotics,[10] although surgery has a higher efficacy than antibiotics with no difference in perforated cases; however, a higher complication rate is associated with the surgical option.

Despite the availability of a range of modern and state-of-the-art investigation modalities, the reported negative appendectomy rate ranges from 8 to 15%.[11] In the current case, even with a meticulous surgical exploration and search, the vermiform appendix was not found. The possibility for sub-serosal, sub-hepatic appendix and Meckel's diverticulum was ruled out, and interestingly, no mesenteric lymphadenopathy was observed perioperatively.

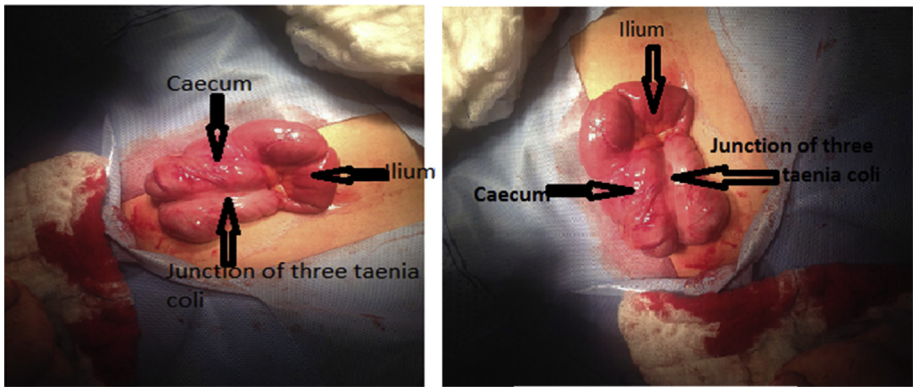

Figure 1: A perioperative view of the surgical field showing the loops of the distal ileum and caecum with no appendix.

Provisionally, the patient was diagnosed as a case of congenital agenesis or absent vermiform appendix and NSAP. Postoperatively, the patient was managed with antibiotics and analgesics, and postoperative recovery was smooth. USG abdomen was performed on the 2nd postoperative day, which was non-diagnostic. On the following day, IV and oral contrast CT of the abdomen and pelvis did not reveal any intra-abdominal abnormality.

NSAP is defined as a condition that refers to abdominal pain of more than 6 h and less than seven days in duration.[12] The causes of NSAP are diseases of the gastrointestinal tract, urinary tract in males and gynaecological disorders in females. Postoperatively, after a negative exploration for presumed appendicitis, NSAP can be further investigated by an ultrasound of the abdomen with overall efficacy of 70%–83%.[13] CT scan has a sensitivity from 68% to 75%,[14] and MRI and laparoscopy have a diagnostic accuracy of 85.2%.[15] As many as 14.7%[16] of patients still remained undiagnosed despite all of the described imaging modalities. A review of several international studies has revealed that NSAP was observed as the most common presentation for patients admitted to emergency surgical wards, with an estimated incidence of 13%–40%.[12]

Conclusion

Congenital agenesis or absence of the vermiform appendix is a very rare condition in the general population. Generally, the diagnosis is incidental. Although it is difficult to evaluate or diagnose preoperatively, a careful search should be performed perioperatively. Additionally, postoperative investigations for final diagnosis should be performed to confirm non-appendicular causes of NSAP. This case report mandates that the treating surgeons maintain a low threshold for considering non-appendiceal causes of abdominal pain, particularly in the presence of a significantly high Alvarado score.

Conflict of interest

The author has no conflict of interest to declare.

Author's contribution

All the components are done by TAS.

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