

Case Report

Appendicular agenesis: a case report

Alaa Sedik^{1*}, Tarek Al-Dawoodi¹, Salwa Elhoushy²

¹Department of General Surgery, ²Department of Internal Medicine, King Khalid Hospital Hail, Saudi Arabia

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*Correspondence:

Dr. Alaa Sedik,

E-mail: asedik59@yahoo.com

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ABSTRACT

Congenital agenesis or absence of vermiform appendix is very rare. Herein, author reported a case of a 28 years old Vietnamese lady who presented with a picture of acute appendicitis. During surgery, author failed to find the appendix. Postoperatively, she made uneventful recovery and she was diagnosed as a case of nonspecific abdominal pain (NSAP) with appendicular agenesis.

Keywords: Abdominal pain, Appendicular agenesis, Nonspecific abdominal pain

INTRODUCTION

Absent vermiform appendix is a very rare finding.^{1,2} The incidence of accounts for 1:100,000 surgical cases undergoing surgery for suspected acute appendicitis. The suspicion of acute appendicitis is made mainly on clinical basis.

This case report aims to discuss a patient who presented with clinical and radiological features of suspected acute appendicitis, but the surgical exploration could not identify the vermiform appendix.

CASE REPORT

Twenty-eight years old Vietnamese lady, working as a housemaid for 2 months in Hail, Saudi Arabia, presented to the emergency room with few days ?? history of vague abdominal pain, fever and vomiting.

There was a big language barrier to deal with her. Author couldn't know details about change of bowel habits, urinary or gynecological problems. Hardly, author found no history of previous trauma or surgery.

Clinically, blood pressure (120/80mmHg), pulse (95/min), temp (38°C) were found and shows no dehydration or cyanosis.

Abdominal examination was mildly distended abdomen with right lower quadrant tenderness. No scars, mild guarding over right lower quadrant, no masses, bowel sounds were exaggerated. Rectal examination revealed some stools.

Lab-works showed WBCs (13,000/cmm), otherwise CBC was normal. Normal urine analysis, liver and renal functions. Urine pregnancy test was normal. Abdominal Ultra-sonography was unremarkable. Contrast enhanced abdominal computerized scan (CECT) showed evidence of 14mm tubular structure in right iliac fossa with fat stranding around with suspicious acute appendicitis. The patient was consented for open appendectomy. During surgery, author couldn't find the appendix; even after extending the wound and searching in sub-hepatic or ileocecal area and retrocecal sub-serous positions. Also, no other pathology, like ovarian cysts or Meckel's diverticulum had been found (Figure 1). Author left a drain and abdomen closed.

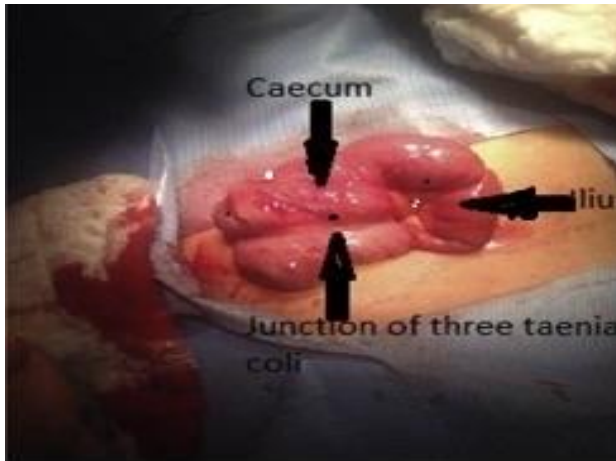


Figure 1: Intraoperative findings showing no appendix was found. The terminal ileum and cecum shown with the convergence of three tinae coli (black dot).

The CECT was discussed again postoperatively with radiology department and all other radiologists interpreted the study as nothing abnormal. The case was followed postoperatively and discharged home on 7th postoperative day in good condition. Ultrasonography came normal before discharge. The case was diagnosed as nonspecific abdominal pain (NSAP). Over the last 6 months, she was seen for follow up twice in the outpatient department and was symptom free. She had normal ultrasonography during the last visit.

DISCUSSION

Embryologically, in 10th week of gestation; the appendix arises from the caecum as an elongated tubular structure with a blind distal end. Its base is constant in position and is 2-5cm below the ileo-cecal valve on the posteromedial aspect of the cecum. The appendicular tip varies in position.³ and its base is almost fixed at the junction of three taenia coli of the caecum. The positions of the tip of the appendix described in the literature are retrocaecal (38%), retrocolic (26%), subcaecal (14%), pelvic (8%) and preileal (3%).⁴ The length varies from 2cm to 20cm 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 Arrest of development may occur at any stage and give rise to either absence of cecum and appendix (type 1), blunt conical cecum without appendix (type 2), longitudinal symmetrical cecum with longitudinal muscle bands converging toward its apex, but without appendix (type 3), or asymmetric cecum without appendix (type 4).⁶ The present case discussed here was type 3 variety of congenital absence of appendix.

Agenesis of the appendix was firstly reported by Morgagni.⁷ The absent appendix rate is 1:100,000 cases. The diagnosis of acute appendicitis is based largely on clinical observation as it is well described by the Alvarado scoring system.⁸

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 10,000cmm or more. An Alvarado score of 7 (as in the case) or higher carries 78% sensitivity and 100% specificity.⁹ Based on this score, the majority of surgeons recommend appendectomy especially in men without any further investigation, but in females ultrasonography may be required to exclude any gynaecological diseases. Equivocal cases with a score of 5 or 6 may benefit from other investigations such as abdomen ultrasonography, CT Scan, MRI and laparoscopy.¹⁰ In the present case, Alvarado score was 7 and that made the decision for appendectomy mandatory. However, a negative exploration revealed a limitation of the Alvarado scoring system.^{11,12}

NSAP is defined as a condition that refers to abdominal pain of more than 6 h and less than seven days in duration.¹³ 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%.¹⁴ CT scan has a sensitivity from 68% to 75%, and MRI and laparoscopy have a diagnostic accuracy of 85.2%.¹⁵ As many as 14.7% 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%.¹³

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. Postoperative investigations for final diagnosis should be performed to confirm non-appendiceal causes of NSAP. This case report mandates that the treating surgeons maintain a low threshold for considering non-appendiceal causes of abdominal pain, specially in the presence of a significantly high Alvarado score.

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