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Case Report

Congenital absence of the appendix in a 21-year-old male with clinical presentation of acute appendicitis: a diagnostic challenge at Prince Ali Military Hospital

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ABSTRACT

Acute appendicitis is commonly associated with right lower quadrant abdominal pain and is a leading cause of emergent abdominal surgeries, carrying a significant morbidity risk. The rarity of anatomical deviations, such as appendix agenesis, complicates diagnosis and can exacerbate clinical outcomes due to diagnostic delays or mismanagement. We report the case of a 21-year-old male, who was surgically explored at prince Ali military hospital for suspected acute appendicitis based on clinical symptoms and an Alvarado score of 7. Despite strong clinical indications, imaging studies, including ultrasound and computed tomography, failed to visualize the appendix. Intraoperatively, appendix agenesis was confirmed, a finding seldom reported in medical literature. This case underscores the challenges of diagnosing congenital appendix absence and highlights the need for heightened awareness among clinicians to prevent unnecessary surgical interventions. Such awareness could potentially improve patient outcomes by sparing them from unnecessary surgical risks and complications.

Keywords: Appendix agenesis, Acute appendicitis, Surgery, Diagnostic imaging, Alvarado score

INTRODUCTION

Appendix agenesis is an exceedingly rare congenital condition marked by the absence of the vermiform appendix. Although generally considered vestigial, the absence of the appendix poses diagnostic challenges, especially in cases presenting with symptoms indicative of acute appendicitis.[1] The prevalence of appendix agenesis is not well-documented, with estimates suggesting it occurs in fewer than 1 in 100,000 individuals.[2] Diagnosis is often incidental, discovered during surgeries for other abdominal issues or through investigations for suspected appendicitis.[3]

Symptoms of acute appendicitis, such as lower right quadrant abdominal pain, nausea, vomiting, and elevated inflammatory markers, can mislead clinicians when imaging fails to visualize the appendix, potentially due to its congenital absence.[4] Many patients with non-visualized appendices undergo surgical exploration based on strong clinical suspicion, which may lead to unnecessary surgeries.[5] Recognizing appendix agenesis is crucial to refining diagnostic protocols and avoiding unwarranted interventions.[6]

Medical professionals need to consider appendix agenesis in differential diagnoses to prevent diagnostic errors and

unnecessary procedures.[7] Misdiagnosis can lead to extended hospital stays, increased healthcare costs, and significant patient distress.[8] Therefore, improving diagnostic accuracy is essential for optimizing healthcare resources.[9]

Enhanced educational efforts aimed at healthcare providers could diminish these diagnostic errors. Training to better interpret imaging where the appendix is not visualized and increasing awareness of this rare condition among clinicians are pivotal.[10]

CASE REPORT

A 21-year-old male patient, with no previous medical or surgical history, presented to the emergency department of prince Ali military hospital with acute abdominal pain. The pain, which was initially peri-umbilical and later localized to the right lower quadrant, began 4 hours before admission. The patient reported associated symptoms including nausea and two episodes of diarrhea, as well as anorexia. There were no accompanying symptoms of vomiting or urinary disturbances.

Upon physical examination, the patient appeared well and was afebrile with stable vital signs. Abdominal examination revealed a soft, lax abdomen with localized tenderness in the right lower quadrant. There was rebound tenderness, and Rovsing's sign was positive. Psoas's sign was positive, suggesting irritation in the area of the iliopsoas muscle, commonly associated with appendicitis, while Murphy's sign was negative, ruling out acute cholecystitis. On auscultation bowel sounds were present, patient Alvarado's score was 7.

Laboratory tests showed an elevated white blood cell count of 15,000/µl, with a neutrophil predominance (9.4/µl) and lymphocytes at 4.8/µl, indicative of an acute inflammatory response.

Imaging studies were pursued further to evaluate the cause of the patient's symptoms. An ultrasound scan of the abdomen was performed, which could not visualize the appendix. To rule out other potential intra-abdominal pathologies and further assess for appendicitis, a contrast-enhanced abdominal CT scan was also conducted. Similar to the ultrasound, the CT scan failed to visualize the appendix, and there were no signs of free fluid or fat stranding around the cecum, which are typical indicators of appendiceal inflammation or perforation.

Given the clinical suspicion of acute appendicitis based on the physical findings and laboratory results, coupled with the non-visualization of the appendix on multiple imaging modalities, a decision was made to proceed with surgical exploration.

During the exploratory laparotomy aimed at appendectomy, the procedure progressed systematically. After the skin incision, the surgical team dissected through the subcutaneous tissue and opened the fascia to access the peritoneal cavity. The cecum and taenia coli were identified along with the terminal ileum. The cecum was mobilized to ensure a comprehensive examination, particularly checking the retrocecal area where the appendix is sometimes hidden.

The terminal ileum was meticulously examined up to 70 cm proximally to rule out the presence of Meckel's diverticulum, a common congenital anomaly that could mimic symptoms of appendicitis. No such diverticulum was found.

Despite extensive dissection around the cecum and terminal ileum, the appendix was not visualized, confirming its absence. The search extended to other possible ectopic locations where the appendix might be located, including the pelvic area, the mesentery of the small intestine, and near the liver. No vermiform appendix was found in any of these areas, supporting the diagnosis of congenital appendix agenesis (Figure 1).

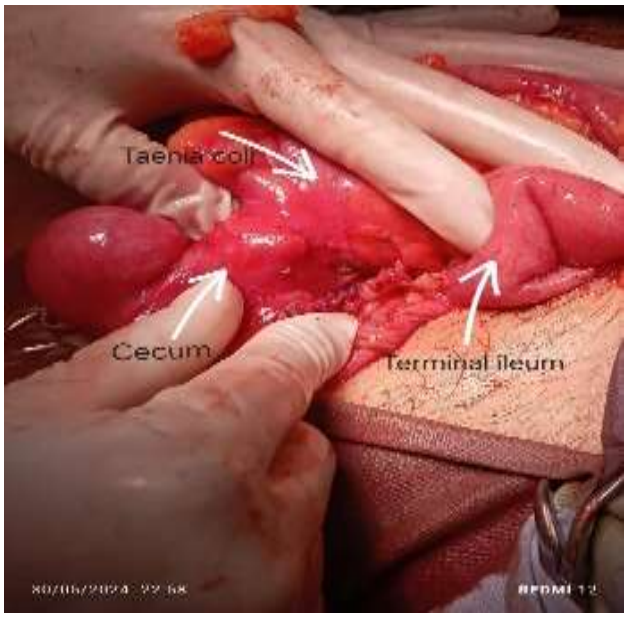

Figure 1: Real-time intraoperative view of the cecoileal region in a 21-year-old male during exploratory laparotomy, confirming the congenital absence of the appendix. No signs of inflammation or other abnormalities are present, providing a clear visual confirmation of appendix agenesis.

During the surgical exploration, two swollen lymph nodes were identified in the vicinity of the terminal ileum. These lymph nodes were excised and sent to histopathology for further analysis to rule out any underlying pathology that might explain the patient's symptoms and inflammatory response.

The abdominal wall layers were then carefully closed in reverse order of opening: the fascia was sutured, followed by the subcutaneous tissue, and finally, the skin, ensuring

proper alignment and suture for optimal healing and minimal scarring. Postoperative management was directed towards recovery, with close monitoring for any potential complications from the surgery.

DISCUSSION

The diagnosis of acute appendicitis is primarily clinical, supported by imaging modalities such as ultrasound and CT scans. However, the absence of the appendix, a condition known as appendix agenesis, presents a significant diagnostic challenge, as demonstrated in this case. Appendix agenesis is an extremely rare congenital anomaly with an estimated prevalence of fewer than 1 in 100,000 individuals.[1] This rarity often leads to diagnostic dilemmas in cases presenting with typical symptoms of appendicitis.[2]

In this case, the clinical presentation of right lower quadrant pain, elevated white blood cells, and positive psoas sign strongly suggested appendicitis. The Alvarado score, which was 7 in this case, further supported the likelihood of appendicitis. The Alvarado scoring system ranges from 0 to 10, and a score of 7 or higher generally indicates a high probability of appendicitis, with surgical intervention considered.[8] However, the inability to visualize the appendix on ultrasound and CT necessitated a surgical exploration, which is consistent with the standard management protocol when imaging results are inconclusive but clinical suspicion remains high.[3] This approach aligns with other documented cases where patients underwent unnecessary surgical interventions due to the non-visualization of the appendix.[4]

The surgical finding of appendix agenesis highlights the limitations of current diagnostic tools in identifying this anomaly. While imaging techniques are highly effective in diagnosing appendicitis, their ability to confirm appendix agenesis is not as reliable. This can lead to unnecessary surgical procedures, which not only pose risks to the patient but also increase healthcare costs.[5]

Moreover, the incidental finding of swollen lymph nodes and the absence of Meckel's diverticulum, which were additional considerations in the differential diagnosis, underscore the complexity of diagnosing abdominal pain. It emphasizes the need for a comprehensive surgical and pathological evaluation when standard diagnostic procedures fail to provide clarity.[6]

Ectopic appendices, though uncommon, are another complicating factor in the diagnosis of appendicitis. It is estimated that ectopic appendices occur in approximately 0.2% to 0.4% of the population. These can reside in various locations including subhepatic, pelvic, pre ileal, and post ileal, which can mislead both clinical assessment and imaging.[9]

In all cases of surgery for suspected appendicitis where the appendix is found to be normal, it is a common practice to search for other potential sources of infection or pathology that could mimic the symptoms of acute appendicitis. These alternate diagnoses include Meckel's diverticulitis, mesenteric lymphadenitis (especially prevalent among children), salpingo-oophoritis, ovarian cysts and complications, and ruptured ectopic pregnancy.[10] Recognizing these potential differential diagnoses is crucial, as they require different management strategies and can significantly impact patient outcomes.

The case adds to the small but growing body of literature that suggests appendix agenesis should be considered in the differential diagnosis of patients with symptoms of acute appendicitis but with negative imaging findings. Developing more sophisticated imaging techniques or diagnostic criteria may help reduce the rate of unnecessary surgeries and improve patient outcomes.[7]

CONCLUSION

This case highlights the need for awareness of appendix agenesis as a differential diagnosis for acute appendicitis, especially in patients with typical clinical signs but negative imaging findings. Further studies are needed to establish guidelines to avoid unnecessary surgeries in such cases.

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