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Case report

Agenesis of vermiform appendix; a case report with literature review

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ABSTRACT

Introduction: Agenesis of the vermiform appendix (AVA) is a very rare finding. The report aims to present a case of suspected acute appendicitis who lacked vermiform appendix during surgical exploration.
Case presentation: A 25-year-old pregnant lady was presented with abdominal pain for a period of 2 days. Her past history was unremarkable. After admission the pain exacerbated. Upon examination; there was tenderness and rebound tenderness in the right iliac fossa. Under general anesthesia the right iliac fossa was explored through right grid-iron incision. No appendix could be found,. Two days after admission the patient was discharged in a good health.
Discussion: Appendix has been considered as a vestigial organ with little or no relevant function. AVA shouldn't be confused with the absence of appendix due to atrophy or any other causes. Usually, AVA is diagnosed in adults. There are no clinical manifestations that can clearly signify AVA prior to surgery, and the appendix has no regulatory function that can be identified in the serum.
Conclusion: The vermiform appendix is considered as a vestigial organ in the body, its congenital absence has rarely been observed and does not seem to have any known impact on the body's function.

1. Introduction

Vermiform appendix is regarded as a rudimentary organ with little or no relevant function in the body [1]. Agenesis of the vermiform appendix (AVA) is a highly rare condition in which the appendix is absent and may be encountered during operation and autopsy, which lacks specific clinical manifestations [2,3]. It has an incidence of 1 in 100,000 laparotomies for suspected appendicitis cases [4]. According to a previous report by Collins, the incidence of AVA is estimated to be 0.0009% of abdominal surgeries and 0.006% in autopsies of suspected appendicitis cases [5].

The aim of the current report is to present a case of suspected acute appendicitis who lacked vermiform appendix during surgical exploration, with a brief literature review. The report has been written in accordance with SCARE 2020 guidelines [6].

2. Patient information

A 25-year-old female who was 7-week pregnant was presented with abdominal pain for a period of 2 days. The pain started at the periumbilical region, later, shifted to the right iliac fossa. She was admitted to the emergency department. She had no history of chronic disease or previous surgery. After admission, the abdominal pain gradually increased and then became localized in the right iliac fossa, in addition, the case also developed anorexia, nausea and vomiting with low grade fever.

3. Clinical findings

Upon examination, there was tenderness and rebound tenderness in the right iliac fossa. Bowel sound was positive. ALVERADO score was nine.

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4. Diagnostic assessment

Laboratory findings were generally normal with the exception of elevated WBC (19.9 109/L). Abdominal ultrasonography showed normal organs with the possibility of acute appendicitis to be correlated clinically. There was no free fluid in the abdomen. Based on the clinical examination, the most differential diagnosis was acute appendicitis.

5. Therapeutic intervention

Under general anesthesia, in supine position, the appendix was explored through right Gridiron incision. No appendix could be found, all adjacent organs (cecum, terminal ileum and both ovaries) were found to be normal with no fluid collection (Fig. 1). After good hemostasis, the wound was closed. The patient was then put on antibiotics, intravascular fluid, and analgesia.

6. Follow up

Two days after admission, the patient was discharged in a good state. Three months later, she was found to be healthy.

7. Discussion

Vermiform appendix (or appendix) is a long, narrow, blind ended and worm shaped organ extending from the posteromedial surface of the caecum, with a distance of 2 cm below the ileocecal valve [7]. The appendix has been considered as a vestigial organ with little or no relevant function [1]. However, there is an abundant number of lymphoid tissues in the wall of the appendix that may provide a kind of immune function [8]. Several congenital abnormalities have been reported to be associated with the appendix with varying frequency, such as; appendix helices (an appendix with helical configuration), appendix atresia (occlusion of the appendix lumen), appendix duplication (duplication of the appendix), appendix triplex (a giant appendix), and AVA (lack of appendix) [2,9,10,11]. Congenital AVA is an exceedingly rare anomaly that was first described by Morgagni in 1716 [12].

AVA shouldn't be confused with the absence of the appendix due to atrophy or any other causes [13]. Sometimes the absent of the appendix is secondary to an intrauterine vascular accident, such as in case of intestinal atresia that occludes blood and cause atrophy of the affected site [14]. Autoamputation of the appendix is another cause of the absent of appendix, which can be suspected by the presence of fibrotic bands [15]. Administration of thalidomide during the first trimester of pregnancy is also associated with the congenital absence of the appendix due to its antiangiogenic property [2]. The current patient was not associated with any prior relevant surgery or diseases, indicating a congenital absence.

According to Collins's classification, there are five types of congenital absence of the appendix; I- cecum and the appendix are absent, II- rudimentary cecum with no appendix, III- normal cecum with the absent of the appendix, IV- normal cecum with not fully developed appendix, V- greatly enlarged cecum distal to ileocecal valve with no appendix, with the third type being most frequently reported. All the types except for the fourth type, occur due to the unsuccessful differentiation of the cecal swelling, while the fourth type occurs as the result of intrauterine atrophy of an initially well-developed fetal appendix [16]. This case was type III.

Usually, AVA is diagnosed in adults [17]. However, there are some extremely rare cases that are diagnosed in children. Vincet et al. reported a case of AVA in a 14-year-old boy [12]. Tripathy also reported a 5-year-old case [18]. The current case was a 25-year-old adult female.

There are no clinical manifestations that can clearly signify AVA prior to surgery, and the appendix has no regulatory function that can be identified in the serum [3]. Which was also the case in this study. During surgery of the relevant site, while attempting to diagnose AVA, the patient must be checked for any sign of previous vermiform appendix, such as, the scar of previous operation, any residual tissue, and the torsion of the appendix should be excluded [10]. In addition; the tinea coli junction along with ileal and retrocecal areas must be explored [17].

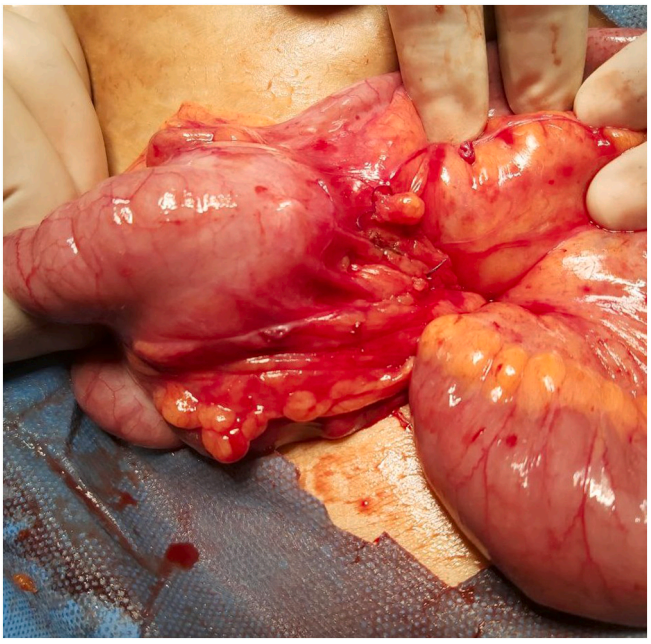

Fig. 1. Intraoperative picture shows absence of the appendix with normal looking cecum.

In conclusion, the vermiform appendix is considered as a vestigial organ in the body, with no known function, its congenital absence has rarely been observed and does not seem to have any known impact on the body's function.

Consent

Written informed consent was obtained from the patient's family for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Declaration of competing interest

None to be declare.

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