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Case Report

Ileocecal stenosis with agenesis of vermiform appendix: A rare cause of congenital bowel obstruction

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ABSTRACT

A 3-month old male patient, with chronic constipation and abdominal distension complaints, was intraoperatively diagnosed with ileocecal stenosis with agenesis of the vermiform appendix. Agenesis of the vermiform appendix is a very rare anatomical finding with an incidence rate about 0.001% in patients who receive laparotomy due to suspected acute appendicitis. No previous cases of ileocecal stenosis with agenesis of the vermiform appendix were reported in the literature. Ileocecal stenosis with agenesis of the vermiform appendix, which present with more shallow symptoms as opposed to ileocecal atresia, can be a cause of congenital bowel obstruction during neonatal period. Stenotic segment was resected without ileocecal valve reconstruction and then ileocolic anastomosis was performed. Development of the baby was normal during post-op 6-month follow-up period. The main objective of this case report is to attract the attention of pediatric surgeons in differential diagnosis of ileus and treatment strategies during laparotomy.

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1. Introduction

Congenital bowel obstructions seen during early childhood or neonatal period can be caused by a variety of conditions such as atresia and stenosis, annular pancreas, malrotation, duplication cyst, meconium ileus, Hirschsprung's disease, neoplasia or trauma.[1–3] The most common cause of this condition is atresia and stenosis.[3] In the literature, no previous cases of congenital bowel obstruction caused by ileocecal stenosis with agenesis of the vermiform appendix were found. Agenesis of the vermiform appendix is a very rare anatomical anomaly which is seen in only 1 case out of 100.000 laparotomy cases with acute appendicitis.[2,4] In this case report, we would like to report a ileocecal stenosis case with agenesis of the vermiform appendix to attract the attention of pediatric surgeons for helping in the differential diagnosis of chronic bowel obstructions.

2. Case report

A 3-month old male patient with a weight of 2800 g presented to Pediatric Surgery department of Faculty of Medicine, with complaints of nutrition intolerance, developmental retardation, delayed meconium, abdominal distension and bilious emesis (Fig. 1). No genetic disease or medication usage during pregnancy was reported by the family. Since abdominal distension did not resolve and vomiting continued during conservative follow-up, the patient was taken to laparotomy. Rudiment cecum with ileocecal stenosis and agenesis of vermiform appendix was seen during the surgery (Fig. 2a and b). Ileocolic anastomosis was done by resecting narrow segment of terminal ileum and cecum. The patient was discharged on the 7th day of operation. No problems or complications were reported during 6-month post-op follow-up period.

3. Discussion

Chronic bowel obstruction can be caused by a variety of conditions.[3] A delayed diagnosis of this condition during neonatal or early childhood periods can cause difficulties in surgical treatment, increased morbidity and mortality rates and complications.[3] Atresia and stenosis cases make up about 1/3 of the congenital bowel obstructions of neonates. During the treatment of the cases, ileocecal valve and appendix anomalies play an important role in planning surgery.[1,5] In the literature, although previous cases of ileocecal atresia with agenesis of the vermiform appendix were reported, no cases of ileocecal stenosis with agenesis of the vermiform appendix were found.

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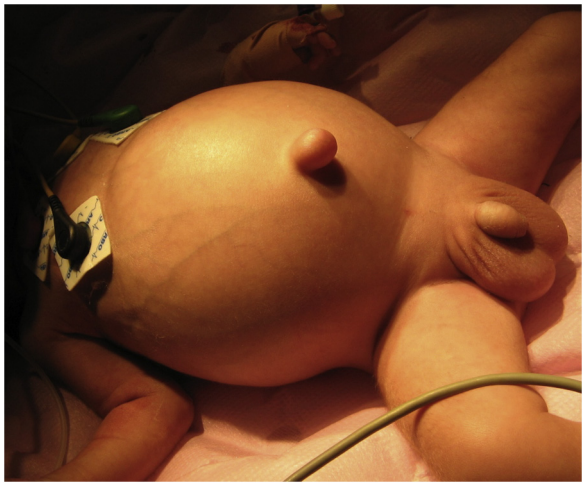

Fig. 1. The photograph shows the patient.

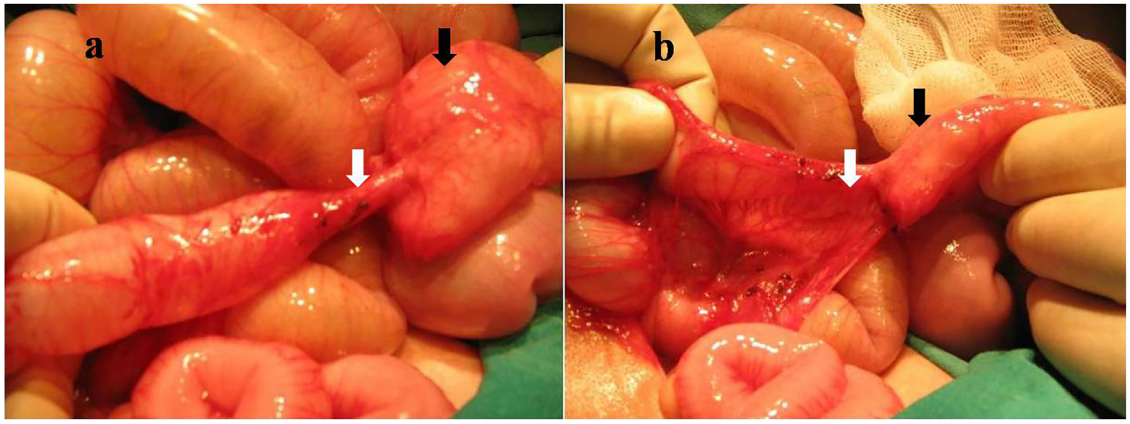

Fig. 2. The photographs show ileocecal stenosis and agenesis of vermiform appendix. 2a and 2b: white arrow shows ileocecal stenosis and black arrow show rudiment cecum with agenesis of vermiform appendix.

First described by Morgagni in 1718, agenesis of the vermiform appendix was reported to have an incidence rate of about 0.006% in autopsy studies, which makes it a very rare anatomical anomaly.[2,6] The etiology of agenesis of the vermiform appendix is still not clear. Appendix, which is the apex of the embryonic cecum, becomes visible in 8th week of gestation and moves from its original location towards posteromedial aspect.[2,7] Appendix becomes visible on the 10th week of fetal development.[7] Any interruptions during this developmental phase can cause congenital anomalies of cecum and appendix.[2] In addition, there are also reported child cases of agenesis of the vermiform appendix due to thalidomide usage during pregnancy.[8] Anomalies of vermiform appendix can have a wide range of manifestations such as variations in localization, duplication, atresia or agenesis.[5] Those rare anomalies of vermiform appendix can be localized or also a part of a larger ileocecal pathology. Collins divided cecum and appendix anomalies in 5 types,[9] and our case is classified as a Type 2 (rudimentary cecum and missing vermiform appendix).

Because there was no ileocecal atresia, the patient showed a clinical representation in a relatively long period which is 3 months. Ileocolic anastomosis was achieved by resecting stenotic segment and cecum without ileocecal valve reconstruction. The patient showed normal development during 6-month post-op period. Ileocolic anastomosis is usually sufficient in short intestinal segment losses without valve replacement.[4]

4. Conclusion

In this paper, we would like to present a case of ileocecal stenosis with agenesis of the vermiform appendix in order to understand the anatomical anomalies which might cause congenital bowel obstructions, making a correct differential diagnosis and using the proper surgical treatment techniques. Ileocecal stenosis with agenesis of the vermiform appendix, which show more shallow signs compared to ileocecal atresia, can also be a cause of congenital bowel obstructions.

Conflict of interest

None.

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