

**LEFT-SIDED
APPENDICITIS IN
A PATIENT WITH
CONGENITAL
INTESTINAL
MALROTATION IN A
BRAZILIAN CENTER:
A CASE REPORT AND
REVIEW OF LITERATURE**

Edgar F. Ndunduma Samoge

Theresa Rodriguez

Afonso DS Bunga

Sophia Garcia

William F. de Araújo Willmer

All content in this magazine is licensed under a Creative Commons Attribution License. Attribution-Non-Commercial-Non-Derivatives 4.0 International (CC BY-NC-ND 4.0).



Abstract: Acute appendicitis (AA) is one of the most frequent surgical emergencies. Typical symptoms and compatible laboratory changes may be absent. In these cases, imaging investigation is useful to establish a correct diagnosis. Intestinal malrotation is a rare congenital condition caused by the absence or incomplete rotation of the small intestine during the embryonic period in which the primitive intestine is arranged in front of the abdominal cavity, and upon returning, the large intestine normally rotates counterclockwise, while the cecum sits in the lower right quadrant. In some patients, malrotation is discovered incidentally as part of the evaluation of another problem. We report and discuss the case of a 21-year-old female patient with a history of peri-umbilical abdominal pain accompanied by nausea; in the emergency room she underwent an abdominal tomography and was diagnosed with acute appendicitis and intestinal malrotation.

Keywords: Acute Appendicitis, Intestinal Malrotation, General Surgery, Case Report.

INTRODUCTION

Acute abdomen accounts for about 7-10% of all admissions to the emergency department, with acute appendicitis (AA) being the most common type. AA has a rate of 5.7-50 patients per 100,000 inhabitants per year, with a greater predominance in the age group of 10-30 years in developed countries [1]. The clinical manifestations of acute appendicitis include abdominal pain, anorexia, nausea, constipation/diarrhea, and fever. Pain is typically located in the periumbilical and epigastric regions, and later migrates to the lower right quadrant, although this only occurs in 50-60% of cases. A leukocyte count greater than 10,000 cells/mm³ and left shift with C-reactive protein greater than 1.5 mg/L are considered diagnostic

indicators. One of the most accurate imaging studies for diagnosing acute appendicitis is computed tomography (CT). Radiological signs of acute appendicitis on CT include an appendix diameter greater than 6mm, peri-appendiceal fat stranding, wall thickening, and the presence of an appendicolith. CT can also distinguish between perforated and non-perforated appendicitis. Magnetic resonance imaging (MRI) and ultrasound can also be used to diagnose appendicitis, but CT is the most used modality [2]. Diagnosing AA is challenging due to its varied presentation. Clinical and laboratory parameters alone have little diagnostic accuracy, and different scoring systems have been proposed for correct diagnosis and risk stratification of patients [3].

On the other hand, mid-gut malrotation is a rare congenital anomaly with an estimated incidence of 1 in 6000 live births, with an unknown true incidence as most cases are asymptomatic. Adult presentation of malrotation is rare, with only 0.2 to 0.5% of cases occurring in adults [4]. When patients with malrotation and acute appendicitis have their cecum and appendix in an abnormal location, they may experience left-sided abdominal pain, which is usually associated with diverticulitis. This can create a problem for doctors trying to diagnose the cause of the pain [5]. We present a case in which the relatively common entity of appendicitis presented itself in a very atypical way. Imaging studies incidentally revealed gastrointestinal malrotation, significantly altering the clinical management.

CASE REPORT

A 21-year-old female patient presented to our Emergency Department with a 6-hour history of sudden-onset peri-umbilical abdominal pain, without irradiation, accompanied by nausea, vomiting, and

fever. Her gynecological history was G1P0A, and her surgical history was a previous surgery done at 15 days of age for probable congenital gastrointestinal anomaly. Her diagnosis was unknown, duodenal stenosis/atresia Vs malrotation was suspected. Any other comorbidities were denied. Patient smoked and was a social drinker. On physical examination, the patient's abdomen was atypical, flaccid and peristaltic, tympanic on percussion, painless on palpation, and without any signs of peritoneal irritation. Laboratory work came out with leukocytosis of 15710/mm and C reactive protein of 0.9 mg/dl. CT of the upper abdomen and pelvis was performed without intravenous administration of contrast. It showcased intestinal malrotation with the cecum located in the left flank (Fig.1). An enlarged cecal appendix in diameter (measuring 10 mm), with parietal thickening and an appendicolith at its base. In addition, it showed densification of adjacent adipose planes (Fig.2). The imaging test was consistent with uncomplicated acute appendicitis.

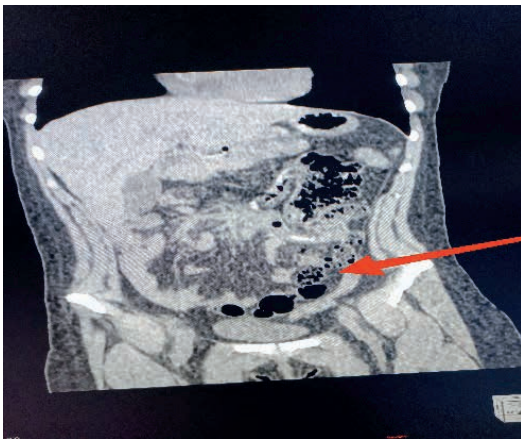


Fig 1: Coronal CT of the abdomen without contrast showing the cecum in the left iliac fossa with obliteration of adjacent fat (red arrow).

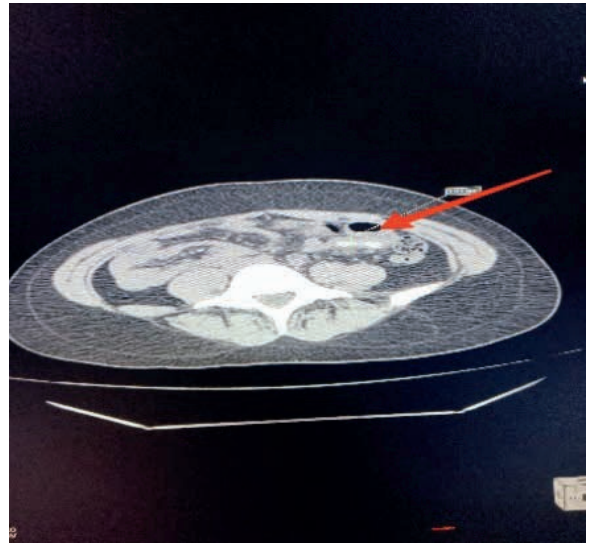


Figure 2. Abdominal CT without contrast in the axial section demonstrating an enlarged cecal appendix in diameter, with parietal thickening and appendicolith at its base.

A videolaparoscopic appendectomy was performed. During the procedure, the cecum identified in the left iliac fossa. A plastron was evident, and the appendix was perforated. After a successful surgery the patient recovered promptly. She was treated with amoxicillin and potassium clavulanate for 7 days to avoid any infections.

DISCUSSION

Midgut malrotation is caused by an abnormality in the embryonic development of the small bowel, which usually rotates 270 degrees counterclockwise around the superior mesenteric artery axis between the fourth to eight weeks of gestation. There are different types of midgut malrotation, including non-rotation, incomplete rotation, reverse rotation, and anomalous fixation of the mesentery [4]. The reported incidence of rotation abnormalities varies widely depending on how they are defined. Autopsy studies suggest an incidence of 0.5%, while a population-based study estimates an incidence of approximately 15 per 1 million in children less than 1 year

old and 10 per 1 million in children aged 1 to 2 years. The incidence decreases after that age group. The presentation of intestinal malrotation in adults is rare and occurs in approximately 0.2%. In some patients, malrotation is discovered incidentally as part of the evaluation of another problem [6].

When malrotation occurs, fibrous bands, called Ladd's bands, develop attaching the cecum to the retroperitoneum, causing volvulus. In those cases, surgical management is necessary and involves detorsion of any volvulus present, division of Ladd's bands, widening of the mesenteric root, proper positioning of the small and large bowels, and a prophylactic appendectomy. An appendectomy is performed to prevent delayed or missed diagnosis of acute appendicitis and to avoid damage to the appendiceal vessels during dissection of Ladd's bands [7]. Our patient underwent surgery at 15 days of life, presumably because of intestinal malrotation or another congenital intestinal anomaly. And, although it is more common for older children and adults to have recurrence of mild symptoms (approximately 30% have vomiting and 20% have recurrent nonspecific abdominal pain) [8], the patient in question was asymptomatic her whole life. It is unknown exactly which procedure she undertook, but it is remarkable that she didn't have an appendectomy during that procedure.

Imaging studies are key to be able to diagnose these cases of intestinal malrotation in adults, Cross-sectional imaging, especially CTs, are increasingly detecting cases of malrotation in adults who have no symptoms. CTs not only show the abnormal positioning of the intestines but also reveal other related conditions that cannot be seen on traditional tests [9]. A CT, in combination with typical appendicitis signs and symptoms, led our team to a prompt and easy diagnosis. Nevertheless, that is not always the case. On another similar

case report on the subject, authors described the necessity to perform a laparoscopic exploration procedure. They had to perform a surgery without a proper diagnosis due to unspecific symptoms and severe abdominal pain [10]. This case report highlights the importance of a proper examination, accurate testing, and imaging.

CONCLUSION

Left-sided appendicitis is a rare condition that can be easily misdiagnosed, especially when its symptoms are non-specific. A thorough physical examination, laboratory tests and proper imaging is mandatory to be able to reach the correct diagnosis without worsening patient's prognosis. Patients diagnosed with symptomatic intestinal malrotation as newborns should also undergo a prophylactic appendectomy during Ladd's procedure to avoid this future complication.

REFERENCES

1. Di Saverio, S., Podda, M., De Simone, B. et al. Diagnóstico e tratamento da apendicite aguda: atualização de 2020 das diretrizes da WSES Jerusalém. *World J Emerg Surg* 15, 27 (2020). <https://doi.org/10.1186/s13017-020-00306-3>
2. Hernández-Cortez Jorge, León-Rendón Jorge Luis De, Martínez-Luna Martha Silvia, Guzmán-Ortiz Jesús David, Palomeque-López Antonio, Cruz-López Néstor et al. Apendicitis aguda: revisión de la literatura. *Cir. gen [revista en la Internet]*. 2019 Mar [citado 2023 Mayo 03] ; 41(1): 33-38. Disponible en: http://www.scielo.org.mx/scielo.php?script=sci_arttext&pid=S1405-00992019000100033&lng=es. Epub 02-Oct-2020.
3. Echevarria† S, Rauf† F, Hussain† N, et al. (April 02, 2023) Typical and Atypical Presentations of Appendicitis and Their Implications for Diagnosis and Treatment: A Literature Review. *Cureus* 15(4): e37024. doi:10.7759/cureus.37024
4. Alani M, Rentea RM. Midgut Malrotation. [Updated 2022 Aug 1]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK560888/>
5. Welte FJ, Grosso M. Left-sided appendicitis in a patient with congenital gastrointestinal malrotation: a case report. *J Med Case Rep*. 2007 Sep 19;1:92. doi: 10.1186/1752-1947-1-92. PMID: 17880685; PMCID: PMC2034390.
6. Langer JC: Intestinal rotation abnormalities and midgut volvulus. *Surg Clin N Am* 97(1):147–159, 2017. doi: 10.1016/j.suc.2016.08.011
7. Al Smady MN, Hendi SB, AlJeboury S, Al Mazrooei H, Naji H. Appendectomy as part of Ladd's procedure: a systematic review and survey analysis. *Pediatr Surg Int*. 2023 Apr 3;39(1):164. doi: 10.1007/s00383-023-05437-7. PMID: 37010655; PMCID: PMC10070202.
8. Kotze, Paulo Gustavo et al. Procedimento de ladd para má rotação intestinal no adulto: relato de caso. *ABCD. Arquivos Brasileiros de Cirurgia Digestiva (São Paulo)* [online]. 2011, v. 24, n. 1 [Acessado 26 Julho 2022], pp. 89-91. Disponível em: <<https://doi.org/10.1590/S0102-67202011000100020>>. Epub 11 Abr 2011. ISSN 2317-6326. <https://doi.org/10.1590/S0102-67202011000100020>
9. Pickhardt PJ, Bhalla S. Intestinal malrotation in adolescents and adults: spectrum of clinical and imaging features. *American Journal of Roentgenology*. 2002 Dec;179(6):1429-35.
10. Hu Q, Shi J and Sun Y (2022) Left-sided appendicitis due to anatomical variation: A case report. *Front. Surg*. 9:896116. doi: 10.3389/fsurg.2022.896116