Acute appendicitis in a patient with intestinal malrotation

Apendicitis aguda en un paciente con malrotación intestinal

Acute appendicitis is the most frequent acute surgical illness in the adult. Typical symptoms include: periumbilical pain that migrates to the right iliac fossa, anorexia, fever, and signs of peritoneal irritation.1 Due to the variety of appendicular positions, one third of the patients with acute appendicitis have pain that is located outside of the right lower quadrant. Left side pain is rarely observed.2 In both children and adults, the left cecal appendix can present in situ inversus totalis or in intestinal malrotation.

A 49-year-old man had a history of morbid obesity, laparoscopic gastric band placement, and pulmonary thromboembolism 7 years earlier. He came to the emergency service complaining of 24-h progression of sudden, colicky, generalized abdominal pain predominantly in the left lower quadrant, abdominal distension, nausea, and pasty stools. Physical examination showed him to be afebrile, dehydrated, with important abdominal distension, reduced peristalsis, involuntary muscle resistance, and pain upon palpation and decompression of the left iliac fossa. His biochemical parameters were: leukocytes of 9.7 10^3/ml, neutrophils of 70%, hemoglobin of 17.2 g/dl, hematocrit of 52.7%, and platelets of 449 10^3/ml. Blood chemistry, serum electrolytes, and urinalysis were normal. Initial diagnostic suspicion was diverticular disease. An abdominal computed tomography scan revealed a probable sigmoid volvulus vs colonic malrotation. A barium enema was then carried out that confirmed colonic malrotation (fig. 1). In a new analysis of the tomography scan, acute appendicitis was identified (fig. 2), with a delay in the diagnosis and treatment of approximately 8 hours. Laparoscopic appendectomy was performed, washing the abdominal cavity and placing drains. The findings were perforated appendicitis in the left iliac fossa and generalized purulent peritonitis. The patient’s progression was adequate and he was released from the hospital on the 4th postoperative day. The histopathologic study confirmed perforated acute appendicitis.

Whereas appendicitis is the abdominal pathology that most commonly requires surgical intervention, intestinal malrotation in the adult is rare. Gastrointestinal tract rotation and fixation abnormalities are frequently associated with abdominal wall anomalies and diaphragmatic hernia. Filston and Kirls3 reported an association with lesions, such as atresias and upper gastrointestinal tract stricture, intussusception, and Hirschsprung disease, of up to 62%.4,5

Intestinal malrotation is an anatomic variant defined as the lack of rotation or incomplete rotation of the intestine. This is caused by a defective rotation of the middle intestine around the axis of the superior mesenteric artery, between weeks 4-12 of fetal life, and the subsequent abnormal fixation to the parietal peritoneum.6,7 It is a broad term that encompasses a wide variety of abnormalities of intestinal rotation and fixation. This alteration can be asymptomatic throughout the patient’s lifetime or can produce a fatal acute abdomen, if not appropriately diagnosed and treated. The clinical presentation of malrotation is: intermittent colicky pain, vomiting, chronic diarrhea, and malabsorption. In cases of exacerbation of another pathology, the typical symptoms appear, as occurred with our patient.

To understand malrotation it is necessary to be aware of the normal embryonal development of the intestine. During fetal development, the digestive tract is a short and straight tube. This tube then becomes elongated and takes up its orderly and stable arrangement in the peritoneal cavity. This process is known as intestinal rotation and fixation, according to the classic description by Snyder and Chaffin.7,8

To better understand the abnormalities of rotation and fixation, they are grouped according to the stage of development in which they were produced, as follows:

- Non-rotation. The small bowel is found in the right part of the abdomen and the colon and cecum in the left part. The distal ileum crosses the midline from right to left to reach the cecum. Our patient had this type of rotation.
Incomplete rotation. This is produced during the final counter-clockwise 180° rotation of the small bowel or the final counter-clockwise 180° rotation of the colon. The intestine is in an intermediate position between non-rotation and the normal postnatal arrangement.

Reversed rotation. The duodenum is in front of the superior mesenteric artery and the transverse colon is behind the superior mesenteric artery.

Different cases of appendicitis with atypical location have been described, and the same as in our case, initial diagnosis is often incorrect, delaying diagnosis and treatment. Abdominal tomography provides sufficient information for the surgical plan. It is not necessary to perform dynamic studies to evaluate intestinal malrotation. The typical tomographic findings in malrotation are: duodenal-jejunal junction on the right side, colon on the left side, and an abnormal orientation of the mesenteric vein and artery. \(^1,2,5,6\) The treatment options for left-side appendicitis are the same as those in cases of its habitual location, and open or laparoscopic appendectomy can be performed. \(^9\) The laparoscopic approach is viable with good results. It is also useful for evaluating the differential diagnoses and resolving different pathologies. \(^9,10\) With respect to the treatment of intestinal malrotation, there is still no consensus on the asymptomatic patient. \(^6\)

Intestinal malrotation in adults is diagnosed incidentally when detected in imaging studies or during the evaluation of another intra-abdominal pathology (as was the case of our patient). \(^1,3,4,9\) The diagnosis and treatment of an acute surgical pathology, such as diverticulitis, or in this case, acute appendicitis, should not be delayed.

**Ethical responsibilities**

**Protection of persons and animals.** The authors declare that no experiments were performed on humans or animals for this study.
Data confidentiality. The authors declare that they have followed the protocols of their work center in relation to the publication of patient data.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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Conflict of interest

The authors declare that there is no conflict of interest.

References


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Primary signet ring cell carcinoma of the colon: A rare condition with a poor prognosis. A report on two cases

Carcinoma primario de colon con células en anillo de sello: una rara entidad de mal pronóstico. Comunicación de 2 casos

Signet ring cell carcinoma of the colon is a rare subtype of mucinous adenocarcinoma, making up less than 1% of all tumors of the colon and rectum. There must be a primary origin in the colon or rectum and at least 50% of the tumor must have a signet ring cell pattern to make the diagnosis. Its presentation is usually late and associated with a high degree of aggressiveness.

We describe 2 cases herein of patients with signet ring cell carcinoma of the colon.

The first case was an 89-year-old woman with no family history of colon cancer. She presented with insidious clinical symptoms of 3-month progression that included anorexia, weight loss, and abdominal pain. Laboratory work-up results reported hypochromic anemia, leukocytosis with no neutrophilia, and elevated C-reactive protein. A computed tomography (CT) scan identified irregular thickening of the cecum, with infiltration of the pericocolic fat and the terminal ileum wall, and no signs of obstruction, as well as numerous regional and retroperitoneal adenopathies (fig. 1A). Long colonoscopy detected an ulcerated and structured mass that took up almost the entire cecal lumen and impeded the passage of the endoscope. Biopsies were positive for signet ring cell adenocarcinoma. Right oncologic hemicolecction was performed and infiltration at the level of the right parietocolic peritoneum and the mesocolon was observed.

Anatomopathologic report: poorly differentiated adenocarcinoma of the colon with > 50% signet ring cell pattern (fig. 1B) with multiple tumor nodules and countless lymphatic embolisms, disperse tumor implants, and stage T4aN2bM1b (2010 ICC/AJCC TNM classification, 7th Edition). Immunohistochemistry showed microsatellite instability in