Intestinal malrotation with appendicitis caused by appendicolith: literature review with an illustrative case.

Poster No.: C-0315
Congress: ECR 2014
Type: Educational Exhibit
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Keywords: Education and training, Congenital, Technical aspects, Intraoperative, Education, Teleradiology, Image manipulation / Reconstruction, CT-High Resolution, Emergency, Anatomy, Abdomen, Image registration

DOI: 10.1594/ecr2014/C-0315

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Learning objectives

Elucidate a diagnose of inflammatory acute abdomen associated with a congenital malformation with unusual symptoms.
Background

Acute appendicitis is one of the most common conditions requiring emergency surgery (1,2), with, however, a high risk of complications if the diagnosis is not performed at an early stage (4). The typical presentation consists of periumbilical pain radiating to the right lower quadrant with peritoneal reaction to palpation - McBurney sign (2).

However, when pain in the left lower abdominal quadrant is extremely rare (1,4), the majority of reported cases associated with congenital malrotation and situs inversus (9) or even a very long appendix (4,13). When patients with asymptomatic gastrointestinal malrotation diagnosed clinically present with abdominal pain, accurate diagnosis and definitive treatment may be delayed, possibly increasing the risk of morbidity and mortality (8).

The intestinal malrotation is a rare pathological condition (10,11,19) consisting of complete or incomplete rotation and fixation of the primitive gut during fetal life (3,6,9,20), appearing in the first weeks of life, but can persist into adulthood in a non-recognized condition (6). Due to the incidence of intestinal malrotation be 0,2 % of the population, complete diagnostic evaluation should be used in patients with acute atypical abdominal pain (18). This abnormal cecal position caused by malrotation (12) makes the acute appendicitis diagnosis difficult (3).

A physician should not be fooled by an atypical presentation of acute appendicitis and should keep in mind the diagnosis to prevent serious complications (2,4,19). Several case reports describe an atypical presentation of appendicitis show that late diagnosis is a common feature among them (6).
Findings and procedure details

Conventional radiography is not sensitive or specific for malrotation, although marks on the right side of the jejunum and no colon full of feces in the right lower quadrant may be suggestive of this finding (14).

Ultrasonography (USG) and CT scan imaging techniques are relevant and complementary to the establishment of a diagnosis (3). The deviation from the normal relationship between the superior mesenteric artery and superior mesenteric vein, is a useful indicator of intestinal malrotation (15), with, in most patients, a vertical relationship or show left-right reversal (16).

In patients with abdominal abnormality unrelated to the malrotation coexistence, the anatomy changes may result in an atypical clinical presentation and appendicitis may have symptoms that are located more on the left side (17). CT scan is useful not only in providing an accurate diagnosis of appendicitis on the left, but also for detecting anomalies associated rotation, which may require surgical correction (6,11).

The diagnosis is confirmed by a set of clinical signs, blood tests, imaging tests (2) and the physician experience who provide care (5).

Imaging tests, particularly CT scan, play an important role in establishing an accurate and rapid diagnosis (1,8), as the delay in diagnosis may occur due to lack of uniformity of the clinical signs and symptoms (1). Careful analysis of the CT scan images, however, must demonstrate the correct diagnosis, including unsuspected malrotation (17). In practice, malrotation may be incidentally discovered superimposed on virtually any abdominal disease (17).

The emergency physician and the radiologist should be aware of these atypical clinical presentations, so that appropriate surgical intervention can be initiated rapidly (7).

CASE REPORT

Female patient, 39 years old, caucasian, Brazilian, born in Santos, lives in São Vicente (São Paulo), constant diffuse abdominal pain at the beginning of intense cramping, evolving progressively worsening for the left iliac fossa and lower abdomen, followed by vomiting with food aspect for 05 days. Denies smoking and drinking, as well as abortions and use of oral contraceptives, referring only two previous cesareans.
At the physical examination the abdomen was distended, with involuntary muscle tension and pain in lower abdomen palpation and percussion and positive rebound hydro-aerial noise. Don’t have comorbidities, referring only childhood asthma and normal menstrual cycle. Father, mother and their two children healthy.

B - HCG was negative and blood count 14,500 leukocytes, requested computed tomography scan (CT scan) that founds:

- Intestinal malrotation identifying the cecum in the left iliac fossa and ascending colon crossing the midline (Figures 01 to 05).
- Obliteration of the fat in the left lower abdomen, adjacent to the cecum, associated with a blind intestinal segment with appendicolith (Figures 01 to 05).

During surgery, the appendix was found necrotic punched in the left iliac excavation, showing plastron (Figures 06 and 07). The surgical procedure was performed without complications and the patient was released in 04 days.
Fig. 1: Intestinal malrotation identifying the cecum in the left iliac fossa with the appendicolith in appendix.

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Fig. 2: Obliteration of fat in the lower left abdomen, adjacent to the cecum, associated intestinal loop segment blindly with appendicolith.

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Fig. 3: After contrast CT scan.

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**Fig. 4:** After contrast CT scan.

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Fig. 5: Intestinal malrotation identifying the cecum in the left iliac fossa and ascending colon crossing the midline.

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Fig. 6: Surgical image of inflamed appendix.

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Fig. 7: Appendicolith surgical image.

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Conclusion

Although its rarity, appendicitis associated with intestinal malrotation should be amidst the differential diagnosis of pain in the left iliac fossa in emergency care, and imaging studies, particularly CT scan, is essential for the correct disease diagnosis.
References