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Acute Appendicitis in Situs Inversus Totalis: A Case Report and Literature Review

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Abstract

Situs inversus is a rare condition caused by a congenital anomaly that consists of the inverted position of thoracic or abdominal organs with respect to the sagittal plane, with a low incidence in the general population and usually incidental finding. On the other hand, acute appendicitis is the acute inflammation of the vermiform appendix, being the most common abdominal surgical pathology in the world. We report the case of a 43-year-old female patient with no medical history, who comes to a hospital in the course of abdominal pain of onset in the epigastrium with subsequent migration of the mesogastrium and right iliac fossa, associated with SIRS, anorexia and signs of irritation. Peritoneal, in whom an open appendectomy was performed with the particularity of showing in the intraoperative ileocecal complex with macroscopic inflammatory characteristics of the left location and inverted position of the rest of the intra-abdominal organs, constituting a rare case of acute appendicitis in complete situs in versus.

Keywords: Acute appendicitis; Situs in versus; Congenital anomaly

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Introduction

Situs inversus is a rare condition caused by an autosomal recessive congenital anomaly caused by mutations located on the long arm of chromosome 14 and deletions that affect chromosomes 7 or 8. Classically, two types of presentations are described, situs in versus totals or complete, the most usual form, characterized by the mirror image location of intra-abdominal and thoracic organs including the heart, or partial or incomplete situs inversus, with a prevalence of 1:22,000 births, determined by the mirror image of the organs intra-abdominal and thoracic, but the heart in normal disposition [1].

Acute appendicitis is one of the most common intra-abdominal diseases and the main abdominal surgical emergency. It accounts for approximately 5% of all emergency department visits for patients under 65 years of age and 30% of acute surgical abdominal emergencies in patients fewer than 50 years of age worldwide [1]. Acute appendicitis with situs inversus totals is reported between 0.016 and 0.024%, becoming a true diagnostic dilemma due to the abnormal position of the appendix and the inconsistency with clinical signs and symptoms. In this article we report a case of acute appendicitis with complete situs inversus.

Case presentation

We present the case of a 43-year-old female patient. She went

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to the emergency room due to a clinical picture of 28 hours of evolution at admission, characterized by predominantly epigastric abdominal pain with migration to the mesogastrium and later to the hypogastrium with lateralization to the right iliac fossa. Associated with quantified thermal increases, anorexia and intolerance to the oral route. Within her personal history, she denies pathological, toxic, and allergic components, but she states that she had performed a caesarean section 8 years ago.

In her initial assessment, a patient was found to be in fair general condition, algid, with vital signs (TA 110/80 mmHG, HR 110 L*m, RR 18 R*m, T 38.5°C, So2 96%), with data from systemic inflammatory response (SIRS)

On physical examination she has clear signs of peritoneal irritation (blumberg, psoas, obturator, dunphy positive), without pain on

movement of the cervix or pathological genital discharge. Blood chemistry is performed with evidence of normochromic anaemia, normocytic WHO GI. Marked leucocytosis at the expense of neutrophilia, with elevated CRP-type acute phase reactant, negative BHCG and coagulation times within normal limits.

Given the manifestations described above and a high-risk Alvarado score for acute appendicitis, it was decided to transfer to the operating room to perform an open appendectomy. Prior administration of prophylactic antibiotics and optimization of medical management.

Initially, a Rocky-Davis-type incision was made, dissecting by planes until reaching the abdominal cavity without showing evidence of an ileocecal-appendicular complex after an exhaustive search. For this reason, an infraumbilical medial approach was performed, requiring blunt digital adherenciolysis due to established Zuhlke grade III omentum-parietal adhesions, drainage of peritoneal reaction fluid. It is evidenced with the ileocecal complex of the left location. An appendectomy was performed with extraction of the surgical piece (image 1) with management of the appendicle stump with 2.0 Pouchet-type silk and a systematic review of the abdominal cavity showing the spleen, stomach, Treitz ligament on the right side and second portion of the duodenum, liver together with the gallbladder, ascending colon on the left side without concomitant injuries. Haemostasis is verified, the parabolic slides and the pelvic cavity are cleaned. We proceed to perform closure by planes, up to the skin. With completed surgical procedure, without immediate complications (Figure 1).

In his postoperative period he evolves satisfactorily, returning positive and effective intestinal transit, with resumption of the oral route early. Thoracoabdominal tomography Figure 2 is performed, showing dextrocardia, confirming sites inversus totalis (Figure 2).

Patient currently without late postoperative complications, without hospital readmissions during the first 30 days.

Literature review

Acute appendicitis is an extremely common abdominal disease or surgical pathology worldwide, which represents one of the main

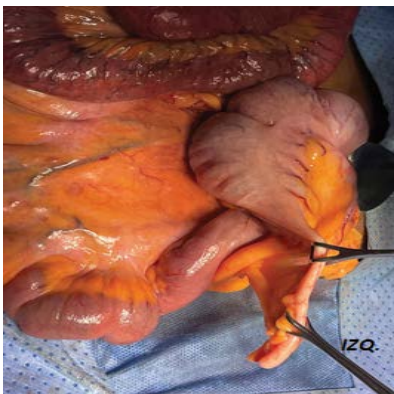


Figure 1 Open appendectomy via laparotomy.

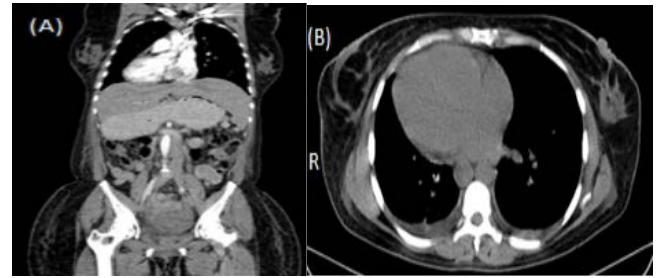


Figure 2 (a) Thoracoabdominal CT in coronal view, showing inversion of the position of the thoracic and abdominal organs. (b) Chest CT showing dextrocardia.

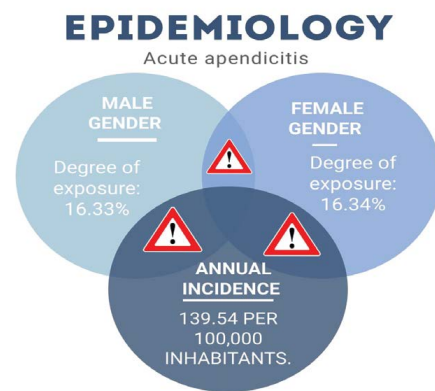


Figure 3 Epidemiology of acute appendicitis.

causes of emergency abdominal surgery. It has been reported that its highest incidence is among the population between 20 and 30 years old, emphasizing that this pathology does not show gender predominance; although in some reports certain percentages are shown. In addition, its clinical manifestation is versatile on certain occasions; therefore the use of imaging studies must be used to establish an effective diagnosis in this way. Similarly, it should be noted that the treatment by which acute appendicitis is managed is surgery with a laparoscopic or open approach [2].

Epidemiology

Acute appendicitis is the most common cause in emergency non-traumatic abdominal surgery, as mentioned above. This presents a degree of exposure of 16.33% in males and 16.34 in females. Its annual incidence is 139.54 per 100,000 inhabitants (Figure 3) [8].

Clinical manifestations

Abdominal pain is the most common symptom manifested in patients, although we can also find other signs such as: nausea, anorexia, diarrhoea and fever. It is necessary to mention that this pain is per umbilical and epigastric, and then expands to the lower right quadrant; however this is a classic symptom that occurs only in 50 to 60% of patients.

The manifestation of nausea and vomiting usually occurs after the onset of pain, pyrexia becomes apparent after six hours of the clinical picture. But to a certain extent these clinical manifestations can change from one person to another [3].

In the physical examination of patients with this pathology, vital signs are usually taken, where we can find a temperature greater than 38°C, heart rate above 100, that is, tachycardia and/or in some cases tachypnea also occurs. On the other hand, it is essential to bear in mind that the first signs and symptoms of acute appendicitis are nonspecific, but that through the irradiation that occurs in the lower right quadrant, it is detectable to a certain extent on physical examination, but they must perform certain studies to corroborate their diagnosis [4].

The maximum point of pain in the abdomen almost always corresponds to McBurney's point, which is located two-thirds of the way from the umbilicus on a line drawn from the umbilicus to the right anterior superior iliac spine [5] (Figure 4).

Diagnosis

To establish an effective diagnosis of acute appendicitis, it is necessary or relevant to take into account certain criteria, such as the physical examination of the patient, the results of relevant studies (laboratories, images, among others) and findings obtained in the interview. Likewise, it should be clarified that through the physical examination a judgment of said pathology can be obtained, but this depends on the experience of the health personnel who carry it out, therefore the importance of carrying out a whole study together and in this way offer an accurate diagnosis [6].

Situs inversus total's (SIT)

Situs inversus total's is a rare congenital malformation, which involves the transposition of thoracic and abdominal organs; which in turn complicates the diagnosis and management of the acute abdomen. The presence of and ITS does not mean a serious risk to health, but its early detection is essential, especially when we are facing certain pathologies that require immediate surgical intervention [7].

Discussion

Acute appendicitis is one of the most frequent intra-abdominal diseases and the main abdominal surgical emergency. It accounts

for approximately 5% of all emergency department visits for patients under 65 years of age and 30% of acute surgical abdominal emergencies in patients under 50 years of age worldwide [8].

Appendicitis accounted for 318,000 hospital admissions in the US in 2006, and has an overall incidence of approximately 82 to 110/100,000 populations in North America. The lifetime risk of appendicitis is 8.6% for men and 6.7% for women in Western countries. And may be double this risk in Asia [9].

It usually presents clinically as a dull epigastric or periumbilical pain, accompanied by nausea or emesis, fever, and hyporexia. Hours later, it radiates and is located in the lower right quadrant [1]. However, its presentation in the left lower quadrant is infrequent, and may be due to congenital or anatomical causes.

Four disorders can cause the appendix to be left-sided, and in order of frequency they are: 1.) visceral transposition (situs inversus), 2.) Intestinal malrotation, 3.) Wandering cecum with a long mesentery, and 4.) Appendage of excessive length, crossing the midline [3]. For reasons of the present case, we will focus on the first note.

Situs inversus is a rare condition caused by an autosomal recessive gene with incomplete penetrance and occurs in 1/10,000 to 1/35,000 live births [4, 5].

This condition can be complete when both the thoracic and abdominal organs are transposed, or partial when only one of these cavities is affected [5].

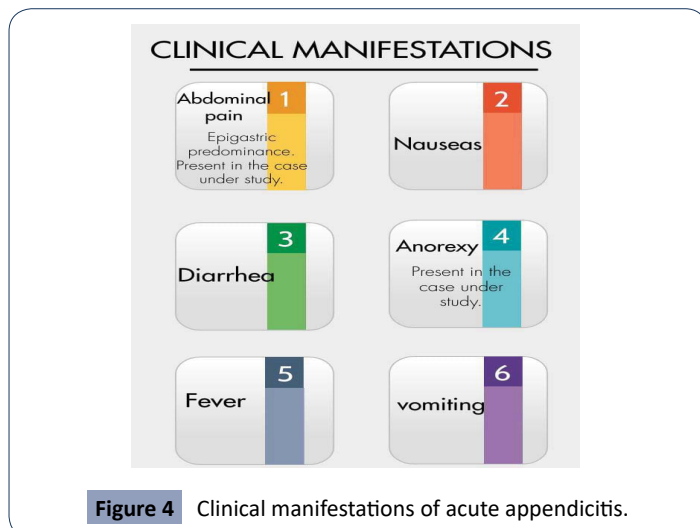
The incidence of situs inversus total's reported in the literature varies from 0.001 to 0.01% in the general population (5,6); however, the incidence of acute appendicitis with situs inversus total's is reported between 0.016 and 0.024% (6). According to reports published in the literature, acute appendicitis in situs inversus occurs between 8 and 63 years of age and is 1.5 times more frequent in men. In a study in the World Journal of Gastroenterology, it was found that the average age was 29.3 ± 16.1 years and the male-female ratio was 1.5:1 [1].

Acute appendicitis in situs inversus is a true diagnostic dilemma due to the abnormal position of the appendix. It is assumed that although the viscera are transposed, the nervous system may not show the corresponding transposition, which results in confusion of signs and symptoms [9]. Thus, in about 18.4 to 31% of patients with situs inversus and intestinal malrotation, the pain caused by acute left appendicitis has been reported in the right lower quadrant [1, 7].

In general, the diagnosis of surgical diseases in these patients is delayed due to low clinical suspicion, so these patients are usually diagnosed in advanced stages of the disease [5].

Diagnosis of acute appendicitis in situs inversus is generally Trans operative motivated by the clinical picture of acute abdomen. In some cases, the imaging diagnosis evidenced by contrast-enhanced abdominal CT is documented, and in some population groups where tomography is contraindicated, another viable option would be MRI [7].

The tomographic findings evidenced would emanate the situs



inversus of the intra-abdominal organs, in addition to the findings already described by the appendicular inflammatory process.

Conclusions

It is extremely important to know the different existing differential diagnoses of acute abdominal pain in the lower right quadrant, since many of these pathologies can cause referred pain with completely different clinical presentation and surgical findings, at the time of performing the operative approach.

Acute left appendicitis, as a rare clinical entity, should not be underestimated and as an aetiology of acute abdomen, its diagnosis and possible treatment should not be delayed as long as possible. With the aim of avoiding life-threatening complications for this small population group. The obvious shortcomings in our health system, especially in the public network. As well as the non-availability of immunological diagnostic aids and minimally invasive or laparoscopic equipment, clinical suspicion should

not be delayed or diminished, especially in this entity, and the surgical approach should be carried out according to the devices or instruments available in each service.

Imaging research and multidisciplinary follow-up, after the generally intraoperative diagnosis of situs inversus, is essential, in order to evaluate possible complications in other organs or systems.

Compliance with ethical standards

Informed consent

Informed consent from the patient in reference to the case was obtained with extensive and extensive prior information.

Interest conflict

The authors declare that they have no conflict of interest with respect to the publication of this document.

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