Situs inversus totalis. A case report

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Case Report

General Surgery

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Introduction: Situs inversus totalis (SIT) is a rare hereditary congenital malformation. It consists of a transposition of thoracic and abdominal organs, which complicates the diagnosis and management of an acute abdomen.
We present the case of a 47 year old male referred to the emergency department after a traffic accident on the public road, on admission he complains of abdominal pain in the left upper quadrant, physical examination shows hematoma in the anterior abdomen, FAST (focused abdominal sonography for trauma) in shock room negative, however continues with abdominal pain located in the left upper quadrant, computed tomography of the abdomen and chest, reports left hemothorax, as a finding SIT and cholelithiasis. Pleural probe was placed in the shock room, favorable evolution after its removal, at his discharge he was referred to general surgery for elective cholecystectomy.

Key Words: Situs inversus.

Introduction

Situs inversus totalis (SIT), a rare autosomal recessive anatomical variant with incidences ranging from 1/10,000 to 1/20,000 per live birth. In a female to male ratio 1:1.5, with no racial predilection. (1)It was first described by Aristotle in animals. In Mexico, the first reported case is attributed to the surgeon Domingo Russi, who in 1760 reported in detail the autopsy performed on the Viceroy, Marques de las Amarillas, which coincided with findings of situs inversus. (2)

Case report

We present the case of a 47 year old male referred to the emergency department after a traffic accident on the public road, on admission he complains of abdominal pain in the left upper quadrant, physical examination shows hematoma in the anterior abdomen, FAST (focused abdominal sonography for trauma) in shock room negative, however continues with abdominal pain located in the left upper quadrant, computed tomography of the abdomen and chest, reports left hemothorax, as a finding SIT and cholelithiasis. Pleural probe was placed in the shock room, favorable evolution after its removal, at his discharge he was referred to general surgery for elective cholecystectomy.

Discussion

SIT an infrequent anatomical variant where the abdominal and thoracic organs, undergo a left-to-right transposition of the same with respect to the sagital plane; this is due to an alteration during the third week of embryological development, at the stage of gastrulation in which the right-left axes of the embryo are established. It is believed that fibroblast growth factor 8, the Lefty1 gene and PITX2 are involved in this modification. (3)

Visceral transposition is a complex group of visceral and vascular abnormalities classified as follows; situs solitus is the normal arrangement of body organs, cardiac apex, aorta, bilobed lung, stomach and spleen to the left of the axial axis, and inferior vena cava, liver together with the gall bladder and trilobed lung to the right of the axial axis. Situs ambiguous which is presented in 1:10.000 to 1:20.000, refers to the modification of the normal anatomical conformation, establishing right or left isomerism, in which the subdivision is presented in situs ambiguous with polysplenia, situs ambiguous with asplenia and Ivemark syndrome. Situs ambiguous with asplenia is more prevalent in males, there is duplication of the right-sided viscera and they are located on the opposite side, typically presenting adult respiratory distress syndrome and severe cardiac anomalies. In contrast, situs ambiguous with polysplenia is more prevalent in women and the structures on the left side of the axial axis are duplicated; likewise, patients with this variation present less congenital cardiac pathology. (4)

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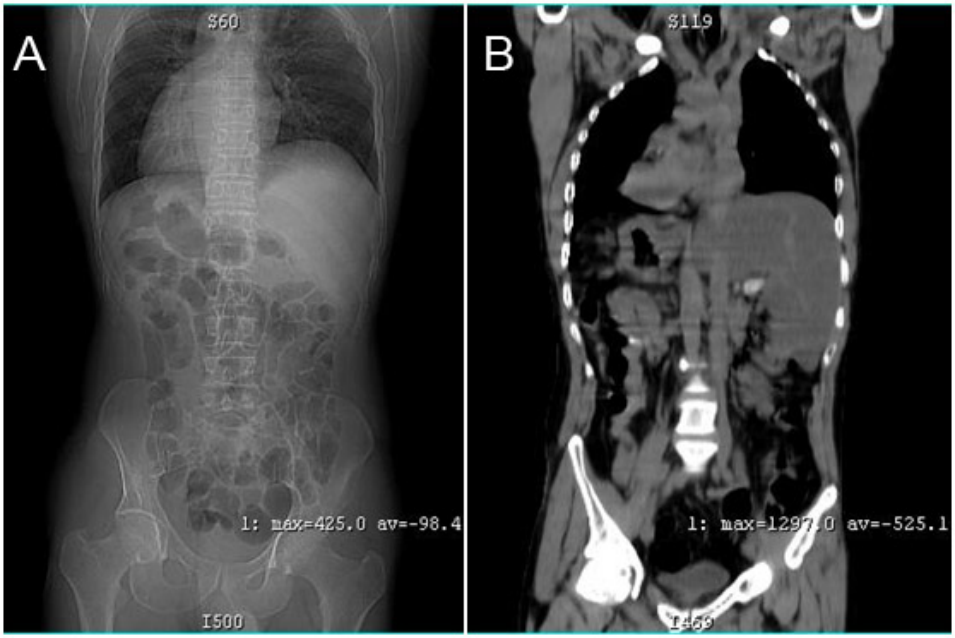

Figure 1. Computer tomography scan, transposition of the cardiac apex and liver. (a) Chest and abdomen CT showed that all situs inversus totalis, that is the heart, lung, stomach, liver, spleen is completely opposite to the normal position. (b)

Finally, the situs inversus is the mirror image of the situs solitus, which presents subdivision in SIT, which is the most usual form, characterized by the mirror location of the intra-abdominal and thoracic organs including the heart; the cardiac apex, spleen, stomach, jejunum, descending colon and aorta are located as right body structures. The right lung is bilobed, with a hypoarterial bronchus. The inferior vena cava, liver together with the gallbladder, ligament of Treitz, ileum and ascending colon are structures located in the left body region. Finally, partial situs inversus is less common, with a prevalence of 1:22,000 births, and mainly the mirror image of the intra-abdominal and thoracic organs is found, but the heart in normal arrangement. (5)

Most patients with SIT lead a completely normal life, but in approximately 20-25% of patients an associated primary ciliary dyskinesia (PCD) may also be observed. Congenital anomalies, including Kartagener's syndrome (bronchiectasis, sinusitis), polysplenia syndrome, biliary atresia (10% 20%), congenital heart disease and midgut malrotation, often coexist with situs inversus. In addition, vascular anomalies (20% 40%), aberrant hepatic arterial anatomy, are associated with this condition. (6) Recently Weicheng et al, published a retrospective cohort study where the prevalence of comorbid diseases was determined, showing 5.8% of digestive system alterations (biliary atresia 4.5% anal atresia 0.6%, congenital biliary dilatation 0.6%). (7)

According to the literature, malignant neoplasms in patients with SIT can be sporadic, with digestive system neoplasms being reported more frequently in the literature. (8)

The diagnosis of SIT goes unnoticed in approximately 45% of the cases, reaching the diagnosis incidentally during the transoperative period of an emergency surgery or as an imaging finding during the study of an unrelated pathology, for example, cholecystitis or appendicitis. (9)

The presence of symptomatic vesicular lithiasis in a patient with SIT leads not only to diagnostic doubts, but also constitutes a real therapeutic challenge, since any surgical procedure is technically more difficult in these patients.

Situs inversus is not a risk factor for cholelithiasis per se, but it carries a risk of diagnostic confusion, as the symptoms and signs will be misleading as they arise from an abnormally placed gallbladder, especially in patients in whom the diagnosis of situs inversus has not yet been made and a delay in diagnosis is expected. In a patient who is already a known case of situs inversus, there will be a high index of suspicion and the diagnosis will be easily made.

Multiple cases of cholecystectomy performed for gallbladder pathology in patients with situs inversus have been described; (10) it is documented that laparoscopic cholecystectomy in patients with situs inversus can be safely performed by adjusting the location of the ports to accommodate.

Laparoscopic cholecystectomy remains the gold standard for symptomatic cholelithiasis even in the presence of situs inversus. (11)

Conclusion

Situs inversus totalis is a rare anatomical alteration that confuses the diagnosis of surgical pathology in the emergency room. However, it will go unnoticed until a series of signs and symptoms are presented as characteristic of a disease, confusing the physician about the existing pathology. This becomes a diagnostic challenge in different scenarios.

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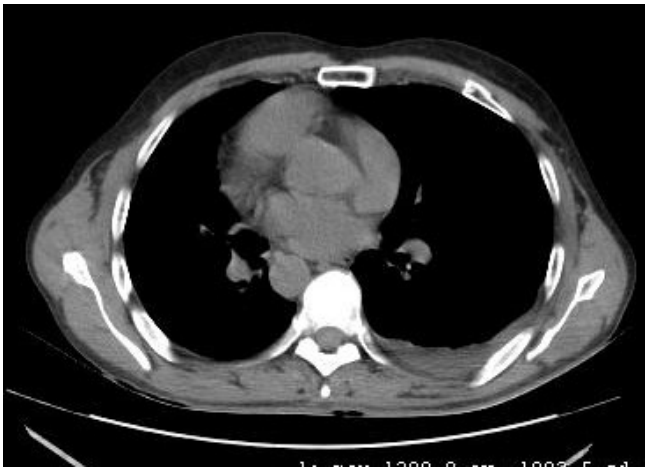

Figure 2. Computer tomography scan, hemothorax in a patient with situs inversus totalis.

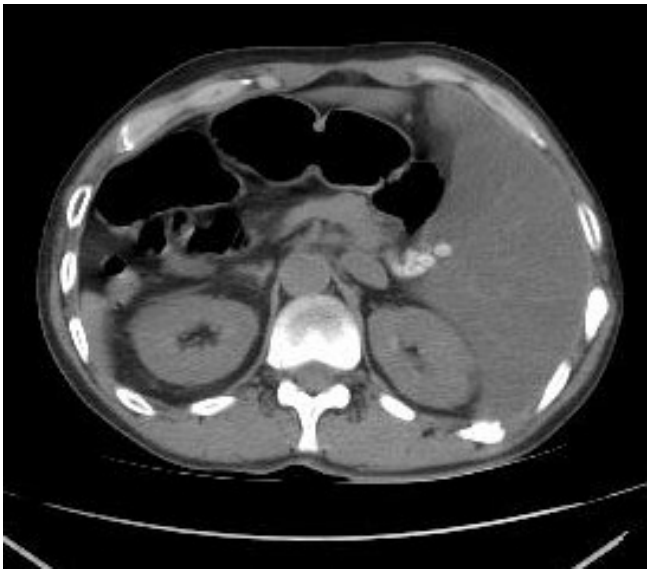

Figure 3. CT image showing complete transposition of the thoracic and abdominal viscera and gallbladder with cholelithiasis.

Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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