Situs inversus totalis and abdominal aortic aneurysm: Surgical repair of an extremely uncommon association

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1. Introduction

Situs inversus totalis (SIT) is an uncommon congenital syndrome characterized by a complete inversion of the thoracic and abdominal viscera with an estimated prevalence of live births 1/10,000 with no sex differences [1].

The cause of situs inversus totalis is due to a disruption in embryological development during gastrulation stage (third week), a period during which the axes are set craniocaudal, dorsoventral, and right-left in the embryo [2]. The exact mechanism by which the situs inversus occurs is still unknown. However, it has been proposed that it is due to a genetic autosomal recessive alteration, in the long arm of chromosome 14, which affects the genetic cascade responsible for the left-rights differentiation.

Most of the patients affected of SIT remain unknown unless cardiac symptoms are present. Only 5–10% has congenital cardiovascular malformations associated [3].

2. Presentation of the case

We report a case of a 69-year-old man, with hypertension controlled by medication, dyslipidemia, hyperuricemia and benign prostatic hyperplasia. By chance in the imaging study of his prostate disease an abdominal aortic aneurysm (AAA) and SIT were found.

The CT scan showed an infrarenal AAA of 6 × 5.5 cm (Fig. 1), extending caudally to the iliac bifurcation with no evidence of rupture.

Under general anesthesia, the patient underwent open aneurysm repair with the interposition of a straight (aorto-aortic) 20-mm Dacron® tube graft. The aneurysm was located on the right side of lumbar spine and we found the inferior mesenteric artery heading right (Fig. 2).

The surgical procedure was performed through a mini median laparotomy (Fig. 3) from the right hand side of the patient without evisceration and with intestinal scrolling to left hemiabdomen. After surgery, the patient progressed favourably without...
Fig. 1. A computed tomography (CT) scan shows SIT and AAA. The inferior vena cava is on the left side.

Fig. 2. The photograph shows the inferior mesenteric artery heading right.

Fig. 3. A surgical photograph shows a mini median laparotomy without evisceration and the graft interposed before being wrapped with the aneurysm sac.

postoperative complications and remained symptom-free at follow-up of 4 years.

3. Discussion

Coexistence of abdominal aortic aneurysm and SIT is extremely rare, so its prevalence has never been estimated, because only seven cases have been reported [4–10].

Kato et al. [4] reported a case of SIT and AAA with satisfactory results using traditional surgical approach. Occhionorelli et al. [5] disclosed a case of symptomatic AAA with SIT requiring emergency surgery by mid-line laparotomy following the usual technique without complications. Baccellieri et al. [6] described a similar case that required interposition of bi-iliac graft with good results. Kimura et al. [7] published the only case of urgent resection and prosthetic graft replacement of a ruptured AAA in a patient with SIT through a right anterolateral thoracotomy successfully performed. Ricci y Deshmukh [8] concluded that routine aneurysmectomy procedure could be performed without problems in the presence of AAA in patients with SIT.

Recently Chan et al. [9] reported the first case of exclusion of AAA using elective endovascular procedure in a patient with high surgical risk and SIT and achieved a satisfactory result.

It is very important to know the presence of this syndrome in order to plan surgery to avoid unexpected surgical complications, such as the left position of the inferior vena cava.

Despite the limited number of cases described, we can conclude that the SIT is not a surgical problem. Occhionorelli et al. [5] described the approach from the left of the patient to prevent surgical complications related to anatomical differences.

In our case the approach from the right side of the patient through a mini median laparotomy of about 15 cm length (Fig. 4) without evisceration and intestinal scrolling to the left hemiabdomen avoided technical complications, quite the contrary, since it facilitated the surgical procedure without anatomical changes will result a problem. The short length of the incision has several benefits, not only the aesthetics, but also allows an adequate surgical approach without exposing the intestine to the air with confinement within the abdominal cavity. This last aspect mentioned is very important for reducing postoperative ileus, hospital stay [11], helps in earlier resumption of oral intake and ambulation [12]. It is also reported that patients not undergoing evisceration through a minimally invasive technique need significantly less fluid administration during surgery, possibly because of less fluid loss and intestinal edema and after surgery because of an earlier return to oral intake [12].

The fact of intestinal scrolling to the left abdomen, unlike usual, is due to the anatomical arrangement of the root of the mesentery which is directed obliquely from duodenojejunal on the left side of the vertebra L2 to the ileocecal junction and right sacroiliac joint. In our case the mesentery root was on the opposite side mentioned. At present, there is no consensus on which should be the optimum approach in cases of open surgical repair of AAA with so uncommon congenital anomaly.

4. Conclusion

Approaching from the right hand side of the patient by using a mini median laparotomy without evisceration and with intestinal scrolling to left hemiabdomen, can be very useful and beneficial
to those cases of congenital anatomical abnormalities that may disturb the surgical repair procedure.

**Conflict of interest**

None.

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None.

**Author contribution**

Dr. Claudia Riera Hernández (main author): Data collection and writing.

Dr. Paulina Pérez Ramírez: Surgical procedure and data collection.

Dr. Carlos Esteban Gracia: Surgical procedure.

Dr. MA. Jiménez Olivera: Surgical procedure.

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**Consent**

We obtained the informed consent from the patient for publication of this case report and accompanying images.

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