CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 10 (2015) 216–218

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

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

Claudia Riera Hernández[a,*], P. Pérez Ramírez[a], C. Esteban Gracia[a], M.A. Jiménez Olivera[b], S. Llagostera Pujol[a]

[a] Vascular and Endovascular Surgery Department, Germans Trias i Pujol University Hospital, Crta Canyet s/n 08916, Badalona, Barcelona, Spain
[b] Vascular and Endovascular Surgery Department, Sant Jaume de Calella Hospital, Carrer Sant Jaume, 209, 08370 Calella, Barcelona, Spain

ARTICLE INFO

Article history:
Received 13 January 2015
Received in revised form 12 March 2015
Accepted 3 April 2015
Available online 8 April 2015

ABSTRACT

INTRODUCTION: Situs inversus totalis (SIT) is an uncommon congenital syndrome, which refers to a reversal mirror-image of the normal thoracoabdominal organs position. The coexistence of SIT and abdominal aortic aneurysm has been seldom previously reported.
PRESENTATION OF THE CASE: We report a case of a 69-year-old man with SIT and infrarenal abdominal aortic aneurysm (AAA) that underwent open repair with a straight graft through a minilaparatomy without evisceration.
DISCUSSION: There is no consensus on which should be the optimum approach in cases of open surgical repair of AAA due to the limited number of cases described. 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.
CONCLUSION: A minilaparotomy without evisceration and with intestinal scrolling to left hemiabdomen, can be very useful and beneficial on those cases of congenital anatomical abnormalities that may add difficulty during the surgical procedure.

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, has been proposed that 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 laparatomy (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

* Corresponding author. Tel.: +34 934978922; fax: +34 934978940.
E-mail addresses: crierahernandez@gmail.com (C. Riera Hernández), paulinaperezramirez@gmail.com (P. Pérez Ramírez), carlosestebangracia@yahoo.com (C. Esteban Gracia), ma_jimenez_olivera@hotmail.com (M.A. Jiménez Olivera), sllagostera.germanstrias@gencat.cat (S. Llagostera Pujol).

http://dx.doi.org/10.1016/j.ijscr.2015.04.007

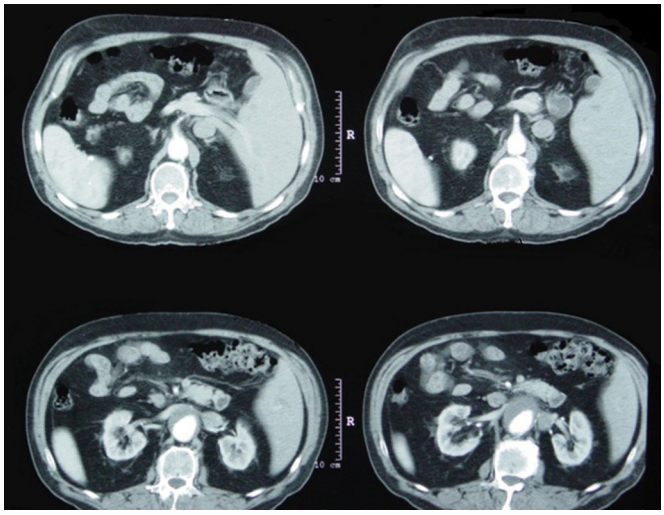

Fig. 1. A computed tomography (CT) scan shows SIT and AAA. The inferior vena cava is on the left side.

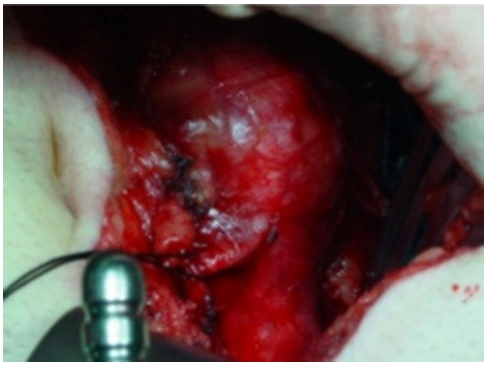

Fig. 2. The photograph shows the inferior mesenteric artery heading right.

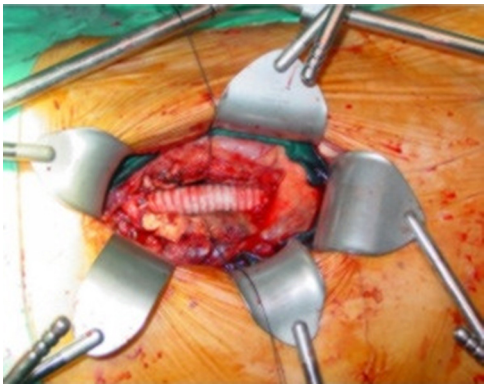

Fig. 3. A surgical photograph shows a mini median laparotomy without evisceration and the graft interposed before being wrapped with the aneurysm sac.

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.

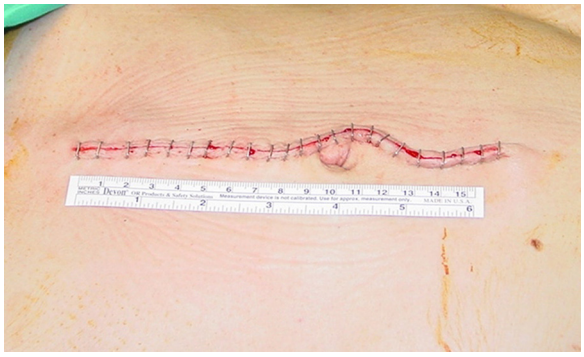

Fig. 4. A photograph of the surgical incision length.

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.

Sources of funding

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.

Dr. Llagostera Pujol: Final paper revision.

Consent

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

References

[1] K. Christian, E.M. Targarona, V. Alonso, I. Moya, G.E. Bendahan, C. Cherichetti, C. Balagué, et al., Cirugía laparoscípica y situs inversus. Revisiín de la literatura y presentaciín de un caso de sigmoidectoméa por diverticulitis, Cir. Esp. 73 (6) (2003) 381–384.

[2] T.W. Sadler, Desarrollo temprano: desde la fecundación hasta la gastrulación, in: En: Langman. Fundamentos De Embriología Médica Con Orientación Clínica, Panamericana, Buenos Aires, 2006, pp. 5–14.

[3] P.D. Maldjian, M. Saric, Approach to dextrocardia inadults: review, AJR Am. J. Roentgenol 188 (June (6 Suppl.)) (2007) S39–S49, quiz S35–8. Review.

[4] T. Kato, H. Takagi, S. Sekino, H. Manabe, Y. Matsuno, T. Umemoto, Abdominal aortic aneurysm in situs inversus totalis, Ann. Vasc. Surg. 20 (March (2)) (2006) 267–270, Epub 2006 Mar 21.

[5] S. Occhionorelli, G. Navarra, M. Santini, P. Carcoforo, A. Sartori, V. Pollinzi, F. Mascoli, Symptomatic abdominal aortic aneurysm and situs viscerum inversus. Diagnostic and therapeutic approach, J Cardiovasc. Surg. (Torino) 39 (February (1)) (1998) 31–33, PubMed PMID. 9 537 532.

[6] D. Baccellieri, F. Mirenda, T. Mandolfino, M. La Spada, F. Stilo, F. Spinelli, [Symptomatic aorto-iliac aneurysm and situs viscerum inversus: case report], Chir. Ital. 58 (January–February (1)) (2006) 113–116, Italian.

[7] N. Kimura, K. Kawahito, S. Ito, Rupture of an abdominal aortic aneurysm in a patient with a situs inversus totalis: report of a case, Surg. Today 3 (2008) 249–252, Epub 2008 Feb 29.

[8] M.A. Ricci, N. Deshmukh, Situs inversus and abdominal aortic aneurysm–a case report, Vasc. Surg. 23 (1989) 413–416.

[9] Y.C. Chan, A.C. Ting, K.X. Qing, S.W. Cheng, Successful endovascular infrarenal aneurysm repair in a patient with situs inversus totalis, Ann. Vasc. Surg. 24 (November (8)) (2010) 1134 e5-7, Epub 2010 May 15.

[10] J.R. Dupont, Isolated left-sided vena cava and abdominal aortic aneurysm, Arch. Surg. 102 (March (3)) (1971) 211–212.

[11] W.D. Turnipseed, et al., Less invasive aortic surgery: The minilaparotomy technique, Surgery 128 (4) (2000) 751–756.

[12] M. Matsumoto, et al., Minimally invasive vascular surgery for repair of infrarenal abdominal aortic aneurysm with iliac involvement, J. Vasc. Surg. 35 (4) (2002) 654–660.