

## CASE REPORT

# Torsion of an accessory spleen with situs inversus in a child

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**Abstract :** We present an unusual case of acute abdomen caused by torsion of an accessory spleen with situs inversus in a child. A three-year-old girl was admitted to our hospital with an 11-day history of right flank pain with fever. Her medical history revealed an operation of coarctation of the aorta with situs inversus at one month of age. Physical examination revealed a right flank mass and tenderness. A contrast-enhanced CT scan showed a normally enhanced small spleen in the right upper quadrant and a 7.0×6.0×3.5 cm, hypodense, marginal enhancing mass in the right midabdomen adjacent to the intestine. An emergency laparotomy was decided upon with a preoperative diagnosis as an acute abdomen. During surgery, a mass was found under the greater omentum and two accessory spleens of 1.5 cm in diameter were found surrounding the main spleen. Several loops of bowel were adherent to the mass. The loops of bowel were dissected away. A pediculated congested mass was observed as an accessory spleen emerging from the greater omentum. The mass was twisted on its vascular pedicle and strangulated. The necrotic mass was removed and the postoperative recovery was uneventful. Though torsion of an accessory spleen is extremely rare, it should be considered in the differential diagnosis of acute abdomen in childhood. *J. Med. Invest.* 59 : 220-223, February, 2012

**Keywords :** accessory spleen, torsion, situs inversus

## INTRODUCTION

Accessory spleen is a congenital anomaly characterized by ectopic splenic tissue separated from the main body of the spleen. It is a relatively common condition that appears in 10% to 30% of autopsy findings and is usually asymptomatic (1). Torsion is a possible complication and occurs exceptionally. Its clinical presentation is characterized by a nonspecific

acute onset or recurrent abdominal pain. The diagnosis is difficult, even with the modern imaging techniques (2, 3).

We report a case of acute torsion of an accessory spleen with situs inversus in a three-year-old girl presenting with acute abdomen. The literature is also reviewed

## CASE REPORT

A three-year-old girl was admitted to our hospital with an 11-day history of right flank pain with fever. Her medical history revealed an operation of coarctation of the aorta with situs inversus at

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one month of age.

Physical examination revealed a right flank mass and tenderness with voluntary guarding. Laboratory findings disclosed only a white blood cell count of  $12,060 /\text{mm}^3$  with 64.6% neutrophils and a c-reactive protein level of 3.38 mg/dl.

Plain abdominal X-ray films revealed a stomach bubble in the right upper abdomen without an ileus pattern (Fig. 1). An ultrasound examination was performed and revealed a well-defined oval mass with a central hypoechoic region, measuring 7.0 cm in



Fig. 1 : The abdominal X-ray shows a stomach bubble in the right upper abdomen and reveals situs inversus.

diameter, anterior to the right kidney (Fig. 2). A contrast-enhanced CT study showed a normally enhanced small spleen in the right upper quadrant and a  $7.0 \times 6.0 \times 3.5$  cm, hypodense, marginal enhancing mass in the right midabdomen adjacent to the intestine (Fig. 3).

An emergency laparotomy was decided upon with a preoperative diagnosis as an infectious intestinal duplication.

During surgery, a mass was found under the greater omentum and two accessory spleens of 1.5 cm in diameter were found surrounding the main spleen. Several loops of bowel were adherent to the mass. The loops of bowel were dissected away. A pediculated congested mass was observed as an accessory spleen emerging from the greater omentum. The mass was twisted anti-clockwise by  $360^\circ$  around its pedicle and strangulated. The necrotic mass was removed (Fig. 4).



Fig. 2 : The US shows a well-defined oval mass with a central hypoechoic region.

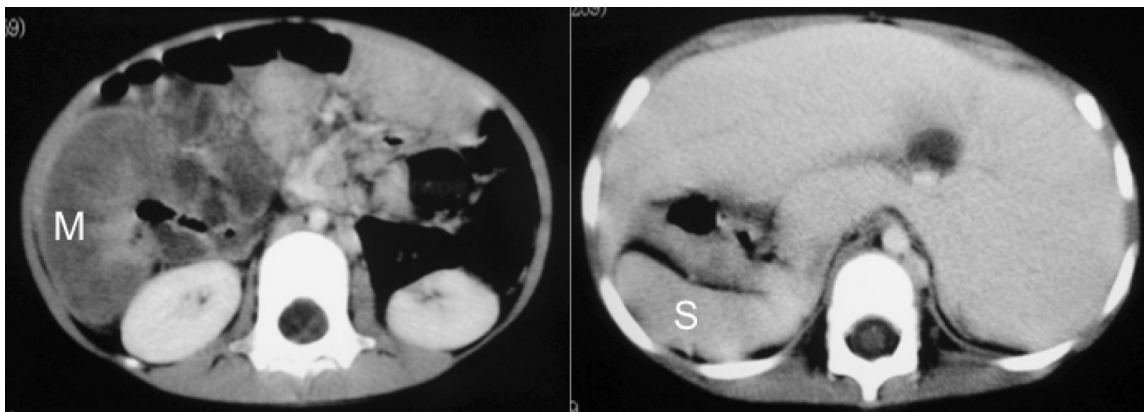


Fig. 3 : A contrast-enhanced CT study showed a normally enhanced small spleen in right upper quadrant (S) and a  $7.0 \times 6.0 \times 3.5$  cm, hypodense, marginal enhancing mass in the right midabdomen adjacent to the intestine (M).

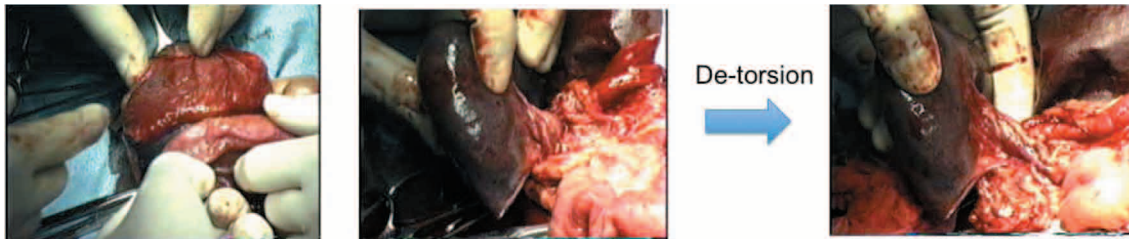


Fig. 4 : A mass adhering to several loops of bowel was found on the greater omentum. A rounded violet mass measuring 7 cm in diameter with a 360° torsion around its vascular pedicle was resected.

The patient's postoperative course was uneventful, and she was discharged in good condition on the 13th postoperative day. Histological examination of the mass was consistent with hemorrhagic and necrotic splenic tissue.

## DISCUSSION

Situs inversus is a congenital condition. It can either be partial or total. This entity is considered to have a genetic predisposition that is autosomal recessive with the defect being localised on the long arm of chromosome 14. The incidence of situs inversus has been thought to be 1 : 5,000 to 1 : 20,000. This condition may affect thoracic organs, abdominal organs or both. It is associated with a number of other conditions such as Kartagener's (bronchiectasis, sinusitis, situs inversus) and cardiac anomalies. There is no current evidence showing increased incidence of accessory spleens in patients with situs inversus.

The spleen arises during the fifth week of gestation from mesenchymal tissue in the dorsal mesogastrium between the pancreas and stomach. Incomplete fusion of these mesenchymal buds during embryogenesis can result in the formation of accessory spleens. Because the spleen is formed in the dorsal mesogastrium and then rotates to the left side, accessory spleens are always situated on the left side of the abdomen (4). Therefore, if there is situs inversus as in our case, it reverses left and right. Most are not separated very far and are vascularized by branches of the splenic artery.

The most common sites of an accessory spleen are the splenic hilum (75%), pancreatic tail (20%), splenic artery, gastrosplenic and splenocolic ligament, and gastrocolic ligament. Other rare locations are mesentery, splenorenal ligament, greater omentum, jejunal wall, presacral area, adnexal region, scrotum, and mediastinum (1-6). Accessory

spleens are rarely found in more than two locations in one person (7).

Usually, accessory spleens are asymptomatic ; torsion and infarction, rupture with bleeding, and infection with abscess are very rare complications. Torsion of an accessory spleen with resultant infarction may cause an acute abdomen (2, 6, 8, 9). In our case where the vascular pedicle of an accessory spleen was very long, there might be pedicular torsion.

Alexander *et al.* (10) reported the first adult case with torsion of an accessory spleen in 1914. A review of the literature revealed only 16 cases (including ours) in the pediatric age of torsion of accessory spleen presenting as acute abdomen (1-3, 6, 7, 11-18) (Table 1). The youngest patient in the literature was described by Gardikis *et al.* (7) in a 14-day-old female patient. There is no case with situs inversus as in our case. All the reported cases went on to

Table 1 Case reports of children with torsion of an accessory spleen

Author	Age	Sex	Location
Settle, 1940	4y	M	Gastrosplenic ligament
	8y	F	Gastrocolic ligament
Babcock <i>et al.</i> , 1974	5y	F	Not stated
Onuigbo <i>et al.</i> , 1978	9y	M	Jejunal wall
	5y	F	Not stated
Nutman <i>et al.</i> , 1982	3.5y	F	Greater omentum
Muller, 1988	15mo	F	Greater omentum
Seo <i>et al.</i> , 1994	10y	M	Greater omentum
Chateil <i>et al.</i> , 1996	15y	F	Not stated
Valls <i>et al.</i> , 1998	13y	F	Pancreatic tail
Pérez Fontán <i>et al.</i> , 2001	13y	M	Greater omentum
Gardikis <i>et al.</i> , 2005	14d	F	Greater omentum
Mendi <i>et al.</i> , 2006	12y	F	Splenic hilum
Impellizzeri <i>et al.</i> , 2009	12y	M	Mesentery
Yousef <i>et al.</i> , 2010	12y	M	Greater omentum
Present case, 2012	3y	F	Greater omentum

the excision of the strangulated accessory spleen, and in only one case described by Yousef *et al.* (18), was a laparoscopic excision performed.

Diagnostic imagings (US, CT) can provide abundant but aspecific information ; however, radiological findings are mandatory for preoperative diagnosis despite not being always available in emergency situations. The correct diagnosis is still quite difficult. In all the previous reported cases with torsion of an accessory spleen, as in our case, the diagnosis was made in the operating room.

Wacha *et al.* (19) reported the value of diagnostic laparoscopy in finding an accessory spleen. Grinbaum *et al.* (4) reported a laparoscopic excision of the strangulated accessory spleen in a young adult and found that in the face of the acute presentation and the radiologic findings, laparoscopy was the preferred action for definitive diagnosis and therapy.

In conclusion, torsion of an accessory spleen should be considered in the differential diagnosis of acute abdomen in childhood. The difficulty of distinguishing this extremely rare entity makes surgical intervention a necessity for diagnosis and treatment.

## CONFLICT INTEREST

The author and co-authors disclosure that there are no conflicts of interest associated with the present study.

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