Accessory spleen torsion: rare cause of acute abdomen in children and review of literature

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Abstract Torsion of an accessory spleen is an extremely rare condition. We describe an unusual case of acute abdomen caused by torsion of an accessory spleen in a 12-year-old boy. The patient underwent a laparotomy with splenectomy; the course was favorable. We discuss the clinical findings and values of preoperative instrumental diagnosis. The literature is also reviewed. This is the 11th case reported in the English literature. Torsion of an accessory spleen should be considered in the differential diagnosis of acute abdomen or subacute abdominal pain.

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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 in a 12-year-old boy presenting with acute abdominal pain. The preoperative diagnosis with abdominal ultrasonography (US) and computerized tomography (CT) was not specific. The literature is also reviewed.

1. Case report

A 12-year-old boy was admitted to our institution after 1 day of increasing intense abdominal pain with vomiting, diarrhea, and no urinary symptoms. The pain was constant and exacerbated by movements. At the time of admission, there was mild pyrexia (37.5°C). Physical examination revealed tenderness to palpation in the mesogastrium and right iliac fossa with moderate resistance. Rectal tenderness was also present. Laboratory findings disclosed only a white blood cell count of 13 600 mmc with 84% neutrophils and polymerase chain reaction of 12.70 mg/dL. An ultrasound examination was performed and revealed a well-defined, homogeneous hypoechochogenic oval mass, measuring 8.7 cm in diameter, in mesogastrium, anterior to the aorta and iliac bifurcation (Fig. 1). Kidneys, liver, pancreas, and spleen appeared of normal size and without any morphological alterations. No evidence of ascites was documented. At Doppler US, no flow within the mass could be recorded. At
CT scan, a homogeneous mass of 8.5 cm in the midportion of the abdomen, within the mesentery, displacing adjacent loops of bowel and in contact with anterior abdominal wall was evident. The mass after intravenous injection remained hypodense with a clear cleavage plane with adjacent structures (Fig. 2). With the suspected diagnosis of congenital malformations of the gut or complicated mesenteric cyst, the patient was operated on. At laparotomy, a rounded violet mass measuring 8.5 cm in diameter with a 760° torsion around a long vascular pedicle along the left side of the abdomen was found and resected (Fig. 3). The pedicle was attached to the root of mesentery. During abdominal revision, the appendix appeared inflamed and then resected. The patient’s postoperative course was uneventful, and he was discharged in good condition on the sixth postoperative day. Histological examination of the mass was consistent with hemorrhagic and necrotic splenic tissue.

2. Discussion

The spleen derives, around the fifth week of pregnancy, from mesenchymal tissue in the dorsal mesogastrium between the pancreas and stomach. An accessory spleen most likely originates from incomplete fusion of the mesenchymal buds. The accessory spleen may be leaded by splenic ligaments to ectopic locations. Accessory spleens are always situated on the left side of the abdomen because of the rotation of the spleen, during embryogenesis, to the left side. The most common site of an accessory spleen is the splenic hilum (75%), pancreatic tail (20%), splenic artery, gastrosplenic and splenocolic ligament, and gastrocolic ligament. Other rare locations are mesenterium, as in our case, splenorenal ligament, greater omentum, jejunal wall, presacral area, adnexal region, scrotum, and mediastinum [1,2,4-6]. Accessory spleens can be either solitary or multiple and receive their vascular supply from branches of the splenic artery.

Usually, accessory spleen is asymptomatic; torsion and infarction, rupture with bleeding, and infection with abscess are a very rare complications [2,7,8]. In these cases, the clinical presentation may be as acute abdomen or with a history of intermittent abdominal pain of uncertain origin. Intermittent torsion-detorsion may produce recurrent bouts of abdominal pain caused by short-lasting ischemia of the accessory spleen or from direct mechanical irritation of surrounding organs [5]. Torsion of the accessory spleen with

Fig. 1 The US shows a well-defined, homogeneous hypoechogenic oval mass, in mesogastrium.

Fig. 2 The CT images scan from the upper abdomen show the spleen in the usual location (S); scans show at a lower level the homogeneous hypodense mass (M).
resultant infarction and necrosis may cause an acute abdomen secondary to long vascular pedicle twisting. This entity is slightly different from splenic torsion in cases of wandering spleen [9].

In our case, we documented an accessory spleen along the mesenteric side presenting as acute abdomen. The torted mass was very large, measuring 8.7 × 8.2 cm. The diagnosis was made in the operating room and confirmed by histological examination. The review of the literature revealed only 13 cases in the pediatric age of torsion of accessory spleen presenting as acute abdomen (Table 1). A case has been recently reported in a neonate [10].

Diagnostic imagings (US, CT) can provide many but aspecific informations; however, the radiological findings are mandatory for preoperative diagnosis despite not being always available in an emergency situation. The US findings can include different heterogeneous features [1,2,11,12]. In our case, US was characterized by well-defined, homogeneous hypoechogenic oval mass in the mesogastrium. The absence of flow within the mass documented at Doppler US did not permit to distinguish between cystic or corpuscular nature.

The CT findings of a twisted infarcted accessory spleen do not differ from that of an orthotopic splenic infarct [1-3]. In our case, the CT revealed only a cystic mass. Consequently, the Doppler US and CT findings were not suggestive of a twisted mass, and the exact organ involved was not determined preoperatively. Moreover, the large size of the mass was atypical for an accessory spleen, as well as the lack of proximity to the spleen made the exact diagnosis uncertain. In all the previous reported cases with torsion of an accessory spleen, as our case, the diagnosis was made in the operating room [1-4,7,13-17].

In conclusion, torsion of an accessory spleen is extremely rare and is still a diagnostic dilemma. In presence of a patient with acute or intermittent abdominal pain and an avascular intraperitoneal mass, torsion of an accessory spleen should also be considered in the differential diagnosis of acute abdomen in children. This condition is very likely if the

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**Table 1** Review of literature

<table>
<thead>
<tr>
<th>Author</th>
<th>Cases</th>
<th>Age</th>
<th>Sex</th>
<th>Location</th>
<th>Diagnostic procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Settle, 1940 [15]</td>
<td>2</td>
<td>4 y</td>
<td>Male</td>
<td>Gastroplenic ligament</td>
<td>US</td>
</tr>
<tr>
<td>Terence et al, 1974 [16]</td>
<td>1</td>
<td>5 y</td>
<td>Female</td>
<td>Gastrocolic ligament</td>
<td>US</td>
</tr>
<tr>
<td>Nutman et al, 1983 [14]</td>
<td>1</td>
<td>3.5 y</td>
<td>Female</td>
<td>Great omentum</td>
<td>US</td>
</tr>
<tr>
<td>Muller, 1988 [13]</td>
<td>1</td>
<td>15 mo</td>
<td>Female</td>
<td>Great omentum</td>
<td>US</td>
</tr>
<tr>
<td>Mendi et al, 2006 [17]</td>
<td>1</td>
<td>12 y</td>
<td>Female</td>
<td>Splenic hilus</td>
<td>CT</td>
</tr>
<tr>
<td>Present case</td>
<td>1</td>
<td>12 y</td>
<td>Male</td>
<td>Mesenterium</td>
<td>US, CT</td>
</tr>
</tbody>
</table>

MR indicates magnetic resonance.
spleen is in normal position. However, preoperative diagnosis is only hypothetical, and prompt surgical intervention is necessary for definitive diagnosis and treatment.

References