



# Accessory spleen located in the right parietal peritoneum

## The first case report

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#### Abstract

**Rationale:** Accessory spleen is a congenital abnormality caused by failure of the splenic anlage to fuse during embryology. The presence of an accessory spleen located in the parietal peritoneum has not been reported so far, and an accessory spleen situated on the right side is extremely rare. In the present study, we describe the first case of an accessory spleen located in the right parietal peritoneum.

Patients concerns: A 65-year-old man, presented with pain in his left abdomen for 1 month.

**Diagnoses:** With ultrasonography and computed tomography, it was difficult to determine the accurate location and diagnosis, and an abdominal fibroma was preoperatively considered.

**Interventions:** By laparotomy, we found a mass connected to the right parietal peritoneum by a vascular pedicle. We resected it completely, and the gross specimen measured  $5.0 \times 3.0 \times 2.5$  cm and was a localized tumor with a capsule.

**Outcomes:** Microscopically, sinusoids were visible, as well as scattered lymphoid follicles, eosinophils, histiocytes, plasma cells, neutrophils, and red blood cells, indicative of splenic tissue. Finally, the lesion was diagnosed as an accessory spleen located in the right parietal peritoneum. Postoperatively, he recovered well and was followed up for a 31 months, during which he was well with no complication.

**Lessons:** We present the first accessory spleen located in the right parietal peritoneum. Awareness of the accessory spleen and familiarity with typical imaging findings are necessary for surgeons to make a precise preoperative diagnosis.

**Abbreviations:** CT = computed tomography, DWI = diffusion-weighted imaging; MRI = magnetic resonance imaging, US = ultrasonography.

Keywords: accessory spleen, case report, imaging modalities, laparotomy, parietal peritoneum

### 1. Introduction

An accessory spleen is a congenital anomaly consisting of normal splenic tissue, and it is caused by the failure of the splenic anlage to fuse during embryology.<sup>[1]</sup> It is seen in approximately 10% to 15% of individuals.<sup>[1,2]</sup> Occasionally, an accessory spleen occurs after splenectomy or trauma.<sup>[3,4]</sup> The lesions are almost smaller than 2 cm; 1 bigger than 4 cm is very rare.<sup>[5,6]</sup> Accessory spleens are usually solitary, as only approximately 10% of patients with one have a second site and accessory spleens with more than 2

deposits are extremely rare.<sup>[1,2]</sup> Patients with accessory spleens are normally asymptomatic, and the lesions are usually found incidentally. Preoperative diagnosis of the congenital abnormality, which usually mimics lymphadenopathy and a tumor, is very difficult.<sup>[7]</sup> Accessory spleens are almost located on the left side of human body. The splenic hilum and tail of the pancreas are the most common anatomical sites.<sup>[5,8]</sup> The splenocolic ligament, greater omentum, mesenterium, adnexal region, and scrotum are occasionally involved. However, an accessory spleen located in the

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Figure 1. Ultrasongram showing a  $4.4\times2.6\,{\rm cm},$  well-defined solid mass (arrow) in the middle upper quadrant of the right abdominal cavity.

parietal peritoneum has not been reported so far. In the present study, we describe the first accessory spleen, measuring  $5.0 \times 3.0 \times 2.5$  cm, in the right parietal peritoneum of an elderly man.

#### 2. Case report

In February 2014, a 65-year-old man was admitted to our hospital because of pain in his left abdomen for 1 month. His abdomen was flat and soft with no palpable mass. He had hypertension for 20 years. He had no history of operative treatment or trauma, and his family history revealed no significant disease. Laboratory results were normal. The ultrasonogram showed a  $4.4 \times 2.6$  cm, well-defined solid lesion in the middle upper quadrant of the right abdominal cavity (Fig. 1). An unenhanced computed tomography (CT) scan showed a well-defined homogeneous mass in the area corresponding to the right liver, abdominal wall, and colon, measuring  $4.9 \times 3.0$  cm (Fig. 2A). On the contrast-enhanced CT scan, the mass was homogeneously enhanced (Fig. 2B). Colonoscopy was performed and no mass was found in the intestinal tract. According to these imaging results, an abdominal fibroma was primarily considered.

We performed a laparotomy and found a well-circumscribed soft mass connected to the right parietal peritoneum by a vascular pedicle. We completely removed the mass, and the intraoperative frozen pathological examination suggested an accessory spleen. Macroscopically, the mass with a capsule measured  $5.0 \times 3.0 \times$ 2.5 cm and was dark red. Microscopically, sinusoids were visible, and also scattered lymphoid follicles, eosinophils, histiocytes, plasma cells, neutrophils, and red blood cells (Fig. 3A and B). Finally, the mass was diagnosed as an accessory spleen in the right parietal peritoneum.

Postoperatively, the patient recovered well and was discharged on the sixth postoperative day. He was followed up for 31 months, during which he was well with no complication.

#### 2.1. Institutional review board statement

The study was reviewed and approved by the Institutional Review Board of the First Affiliated Hospital, School of Medicine, Zhejiang University. Informed consent was obtained from the patient.



Figure 2. An unenhanced computed tomography (CT) scan showing a well-defined homogeneous mass (arrow) in the area corresponding to the right liver, abdominal wall, and colon, measuring 4.9 × 3.0 cm (A). On the contrast-enhanced CT scan, the mass (arrow) is homogeneously enhanced (B).



Figure 3. Histopathological examination showing that the mass is enclosed by a fibrous capsule (arrow) (A) (HE ×50). Sinusoids, scattered lymphoid follicles, eosinophils, histiocytes, plasma cells, neutrophils, and red blood cells are visible (B) (HE ×100). HE=hematoxylin and eosin.

#### 3. Discussion

An accessory spleen is an ectopic mass of healthy splenic tissue separate from the main body of the spleen.<sup>[1]</sup> The false fusion of some buds of splenic tissue in the dorsal mesogastrium during embryologic phase is the reason.<sup>[9]</sup> Of the general population, 10% to 15% may have an accessory spleen. Reported by a study, 364 accessory spleens were found in 3000 autopsies (12.1%).<sup>[2]</sup> When the main spleen is involved with hematologic or other systemic diseases including lymphoma, leukemia, thrombocytopenia, and hemosiderosis, ectopic spleens are also often involved.<sup>[2,10]</sup> Patients with the lesions are usually asymptomatic, and the lesions are usually found incidentally on imaging studies, and mimic lymphadenopathy and a tumor.<sup>[11]</sup> Accessory spleens are often located on the left side of the human body.<sup>[12]</sup> Nearly all the lesions are smaller than 2 cm, and 65% of them are 1 cm in diameter or less.<sup>[10]</sup> They can be located in the hilum of the spleen, splenic artery, pancreas, splenocolic ligament, greater omentum, mesenterium, adnexal region, and scrotum.<sup>[5,8]</sup> However, an accessory spleen in the parietal peritoneum has not been reported so far. In the present study, we described the first accessory spleen, measuring  $5.0 \times 3.0 \times 2.5$  cm, located in the right parietal peritoneum in a 65-year-old man.

The preoperative accurate diagnosis of an accessory spleen is difficult, as it can be mistaken for other mass-forming lesions. Imaging modalities such as ultrasonography (US), CT, and magnetic resonance imaging (MRI) have certain diagnostic value, but lack specificity.<sup>[13,14]</sup> On an ultrasonogram, a typical accessory spleen is often shown as a well-defined lesion. US with the administration of an intravenous contrast agent is helpful for the visualization of vascular hilum of an accessory spleen.<sup>[13,14]</sup> On an unenhanced CT scan, a typical accessory spleen is depicted as a well-marginated mass. On a contrastenhanced CT scan, the lesion is enhanced homogeneously as splenic parenchyma.<sup>[10,15]</sup> MRI using superparamagnetic iron oxide as a negative contrast is also helpful for the detection of an accessory spleen.<sup>[16]</sup> In both T1-weighted and T2-weighted images, accessory spleen shows reduction of the signal intensity, consistent with the observation for the main spleen.<sup>[14]</sup> Recently, MRI with diffusion-weighted imaging (DWI) has been increasingly used in abdominal imaging examination. Accessory splenic tissues show marked hyperintensity on high b-value DWI, identical to that of the spleen.<sup>[17]</sup> The combination of DWI and conventional MRI may improve the characterization and detection of heterotopic splenic tissue. Scintigraphy with Tc-99m phytate may be the most useful method for evaluating a functional accessory spleen.<sup>[18]</sup> However, scintigraphy is difficult to perform immediately after the incidental detection of a lesion. In our case, we did not consider scintigraphy even after US and CT were performed. A US-guided fine-needle aspiration biopsy is also an important tool for making a differential diagnosis.<sup>[19,20]</sup>

An accessory spleen generally does not require therapy.<sup>[1]</sup> Accurate preoperative distinction would obviate unnecessary operation. However, despite availability of multiple diagnostic tools, it is difficult to make a clinically preoperative diagnosis, and the mass usually mimics lymphadenopathy and a tumor. In the present case, the mass was preoperatively considered as an abdominal fibroma. The definitive diagnosis of an accessory spleen is determined by a histopathological examination of the surgical specimen. When the accessory spleen is involved with hematologic or other systemic disorders, such as leukemia/ lymphoma or thrombocytopenia, accessory spleens are also usually involved, and surgical treatment is necessary.<sup>[2]</sup> Operative treatment is also suggested when accessory spleens become symptomatic because of torsion, spontaneous rupture, hemorrhage, or cyst formation.<sup>[21]</sup> In our case, the definitive location and nature of the mass were both difficult to preoperatively determine. In addition, the vascular pedicle of the accessory spleen may cause abdominal symptoms due to torsion, which can lead to rupture and infarction.<sup>[22]</sup> We performed operative treatment and found that the mass was an accessory spleen located in the right parietal peritoneum by a vascular pedicle.

Macroscopically, a typical accessory spleen usually appears as a solid mass with a smooth, round, ovoid, or minimally lobulated shape. Microscopically, it reproduces the splenic pattern.<sup>[10]</sup> An accessory spleen usually has a well-defined fibrotic capsule that separates the surrounding normal tissue.<sup>[16]</sup> A polymorphous population of hematopoietic cells can be seen, including lymphocytes, eosinophils, histiocytes, plasma cells, and red blood cells, admixed with a large number of small blood vessels representing splenic sinusoids.<sup>[20]</sup> Immunohistochemical staining for CD8 is important to confirm the diagnosis, as it specifically highlights the endothelial cells of the splenic sinus.<sup>[19]</sup> When superimposed masses, such as lymphoproliferative disorder and hemosiderosis, are located in an accessory spleen, they frequently have an consistent appearance to the same masses within the main body of the spleen.<sup>[10,15]</sup>

#### 4. Conclusions

An accessory spleen in the parietal peritoneum has not been reported so far, and an accessory spleen in the right side is extremely rare. Most masses have been less than 2 cm in diameter. Herein, we described the first case of an accessory spleen located in the right parietal peritoneum, measuring  $5.0 \times 3.0 \times 2.5$  cm. We resected the mass completely, and the patient was well with no complication during the 31-month follow-up. Awareness of an accessory spleen and familiarity with typical imaging findings are necessary for surgeons to make a precise preoperative diagnosis.

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