Original Article
Diagnosis of multiple splenosis in right thorax and retroperitoneum: a case report and literature review

Yuwei Tian1, Qingfeng Jiang1, Ke Li1, Quan Shen1, Fangfang Guo2, Fangfang Fu3, Cuiyun Chen3, Huanzhou Xue1

Departments of 1Hepatobiliary Surgery, 2Pathology, 3Radiology, Zhengzhou University People’s Hospital, 7th Weiwu Road, Zhengzhou 450003, Henan, P. R. China

Received December 13, 2015; Accepted February 18, 2016; Epub July 1, 2016; Published July 15, 2016

Abstract: Ectopic spleen is an autotransplantation of splenic tissue after traumatic rupture of the spleen or a splenectomy. Clinically, since most splenosis patients have no obvious symptoms and the condition is usually detected during accidently physical examination. Splenosis is easy to be misdiagnosed as a tumor, leading to unnecessary surgical treatment. Multiple splenosis growing in the right thorax and retroperitoneum is very rare. In this study, we report on the first case of combined thoracic and retroperitoneal splenosis in a 42-year-old male who was involved in a traffic accident 35 years ago. Multiple splenosis in the right thorax and retroperitoneum were accidentally detected by ultrasound examination. Histopathological examinations of the nodules resulted in obvious red pulp and white pulp structures. The imaging results and histopathological examinations collectively supported the diagnosis of combined thoracic and retroperitoneal splenosis. The purpose of this article is to share the experience of multiple splenosis diagnosis in the right thorax and retroperitoneum. We also review the literature of retroperitoneal splenosis in order to improve the understanding of splenosis and avoid unnecessary surgery in the future.

Keywords: Ectopic spleen, splenosis, thoracic, retroperitoneal, diagnosis

Introduction
Splenosis is a unique acquired condition resulting from the autotransplantation of splenic parenchyma into unexpected locations, such as the abdomen or subcutaneous tissue [1]. The precise pathogenesis of splenosis is unknown, but it is suggested that the condition is related to mechanical trauma and splenic rupture thus releasing splenic pulp into the surrounding tissues [2]. For this reason, splenosis can occur anywhere in the body, but is most frequently found in the intraperitoneal space and may be present in as many as 65% of splenic rupture cases [3]. Clinically, since most splenosis patients have no obvious symptoms; occasionally the malady is detected during physical examination. Overall, splenosis is easy to be misdiagnosed as other tumor types, often leading to unnecessary surgical treatment. Splenosis is most commonly found in the peritoneal cavity unless the retroperitoneal reflection is breached, and right retroperitoneal splenosis is rarely reported. The majority of splenosis cases occur in males. This phenomenon is probably related to the higher incidence of risky behavior and trauma in men [4]. To our knowledge, multiple splenosis growing in the right thorax and retroperitoneum is indeed very rare. Here we report on a case of combined thoracic and retroperitoneal splenosis in a 42-year-old male who was involved in a traffic accident when he was 7 years old. This is the first report of combined thoracic and retroperitoneal splenosis found in English medical literature. The purpose of this article is to report on the experience of diagnosing multiple splenosis in the right thorax and retroperitoneum. We also review the literature of retroperitoneal splenosis in order to improve the understanding of this condition, which can help avoid unnecessary surgery in the future.

Case report
A 42-year-old male was admitted to our institution (Zhengzhou University People’s Hospital) for the complaint of upper right abdominal pain.
The patient mentioned that his spleen and left renal were dissected for the repair of a diaphragmatic hernia after a traffic accident 35 years earlier. In the local hospital, the abdominal B ultrasound and computed tomography (CT) examinations showed gallbladder stones and multiple lesions growing in the right thorax and retroperitoneum. The density of the multiple lesions was slightly lower than that of the liver in the CT plain scan. These lesions were heterogeneous in the arterial phase and homogeneous in the venous phase. Upon admission to our hospital, we performed magnetic resonance imaging (MRI) in order to confirm the diagnosis. At the time of evaluation, the patient did not have any respiratory symptoms. The patient had no family history of related diseases and his physical examination was unremarkable with the expected healed surgical scar at the left lateral abdomen. The MRI examination showed multiple lesions in the right thorax and retroperitoneum which were believed to be a neurogenic tumor. T1WI (T1 weighted imaging, T1WI) showed homogenously hypointense, while T2WI (T2 weighted imaging, T2WI) and DWI (diffusion weighted imaging, DWI) showed homogenously hyperintense. Dynamic enhancement in the early and later arterial phase showed uneven enhancement, while the venous and delayed phases were homogenous-ly enhanced (Figure 1A-G). Thoracic and retroperitoneal splenosis were suspected, considering the history of abdominal trauma and the nature of the lesions found during CT imaging. However, a malignant tumor or pleural metastasis was also considered. A blood test demonstrated: 140 g/L of hemoglobin, 4.23 × 10^{12}/L of red blood cell, 7.8 × 10^{9}/L of white blood cell, and 213 × 10^{9}/L of platelet. Intraoperative findings showed that there were multiple nodules in the right thorax and retroperitoneum of varying shapes (e.g., round, ovoid and spindle). The nodules were of different sizes, measuring 7 cm × 4 cm × 1.8 cm, 4 cm × 2.2 cm × 0.7 cm, 2.6 cm × 1.3 cm × 0.6 cm, and 1.1 cm × 0.6 cm × 0.3 cm, respectively. It was suspected that those nodules were derived from the small blood vessels near the parietal pleura and peritoneum.

The imaging results (CT and MRI) and other laboratory tests could not convince the patient and he decided to receive surgery to remove the lesions instead of minimally puncture aspiration. As required by the patient, the lesions were resected and sent for pathological examination for further confirmation. The pathological examination showed clearly that the lesions were splenosis with obvious red pulp and white pulp structures. The white pulp consisted of
Splenosis in the right thorax and retroperitoneum

Figure 2. Histopathological examination of splenosis by Hematoxylin & Eosin staining. Splenic parenchyma composed of red pulp and white pulp, and white pulp structure is similar to the splenic tissue. The images were taken under 40× objective. Red pulp and white pulp are indicated by arrows.

thick wall, small artery and lymphoid follicles, while the red pulp was comprised of a venous sinus and splenic cord, which contained a large number of macrophages (Figure 2A, 2B). The patient’s postoperative recovery was uneventful, and no signs of recurrence were detected on CT scan during a six month follow-up period. In summary, the imaging results and histopathological examinations supported the diagnosis of combined thoracic and retroperitoneal splenosis in this case. The patient was given informed consent and this study was approved by the Ethics Committee of Zhengzhou University People’s Hospital.

Discussion

Splenosis is the dissemination of splenic cells when splenic capsule integrity is breached. Splenosis is usually asymptomatic except for some unusual presentations [5]. For this reason, splenosis is usually incidentally discovered after general screening or during the workup for another health issue. In 1937, Shaw and Shaft H firstly reported six clinical cases of splenosis after splenic surgeries. The true incidence of splenosis remains unknown, with a reported incidence of up to 67%. The occurrence is higher in young male patients due to the increased ability of young splenic cells to implant as well as the increased incidence of young males who sustain splenic injury [6]. Ectopic spleens are driven by a few factors. First, the majority of patients had a traumatic rupture of the spleen or splenectomy, which caused the spleen cells to shed and plant in potential lacuna in the body. Most diffused spleen cells grew in the left abdominal cavity, followed by the omentum of the abdominal cavity, peritoneum, serosa layer of the small intestinal, retroperitoneum, diaphragm ventral side, etc. Splenic implants can be found anywhere, including: the peritoneal cavity, pelvis, retroperitoneum, mediastinum, pleural cavity, and subcutaneous tissue [7].

Thoracic splenosis is a rare condition that usually follows simultaneous splenic and diaphragmatic injury, with autotransplantation of splenic tissue into the left hemithorax [8]. A review of 38 cases of thoracic splenosis found that the diagnosis was more common in males than females, possibly reflecting the higher incidence of trauma in young males [4]. Thoracic splenosis typically presents on chest radiographs as an asymptomatic pulmonary mass, either solitary or multiple. Thoracic splenosis can mimic primary or metastatic lung cancer and diagnosis is difficult without knowledge of previous splenic injury. Combined thoracic and retroperitoneal splenosis are much less reported. As shown in Table 1, we reviewed the available literature reporting retroperitoneal splenosis in the English literature. Only 5 cases of retroperitoneal splenosis have been reported in the English language with full information. Most instances of retroperitoneal splenosis are due to traffic accidents and diagnosed using CT or MRI. The present study is the first report of combined thoracic and retroperitoneal splenosis found in the English literature.

The age of ectopic spleen after splenic rupture and splenectomy spans from 15 to 79 years with an average age of 44 years [9]. The time
Splenosis in the right thorax and retroperitoneum

Table 1. Review of the clinical characteristics of patients with retroperitoneal splenosis

<table>
<thead>
<tr>
<th>Age/sex</th>
<th>Cause of splenectomy</th>
<th>Chief complaint</th>
<th>Interval (year)</th>
<th>Size (cm)</th>
<th>Number of lesion</th>
<th>Diagnostic approach</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>55/F</td>
<td>Laparoscopic radical nephrectomy</td>
<td>Renal tumor recurrence</td>
<td>0.6</td>
<td>1.5</td>
<td>2</td>
<td>CT</td>
<td>[7]</td>
</tr>
<tr>
<td>52/M</td>
<td>Traffic accident</td>
<td>Progressive renal failure</td>
<td>36</td>
<td>5</td>
<td>1</td>
<td>MRI</td>
<td>[16]</td>
</tr>
<tr>
<td>40/M</td>
<td>Trauma splenectomy</td>
<td>Incidental finding</td>
<td>25</td>
<td>3.3</td>
<td>1</td>
<td>CT</td>
<td>[17]</td>
</tr>
<tr>
<td>54/F</td>
<td>Post-traumatic splenectomy</td>
<td>Epigastric and low back pain</td>
<td>38</td>
<td>ND</td>
<td>Multiple</td>
<td>CT</td>
<td>[5]</td>
</tr>
<tr>
<td>65/M</td>
<td>Traffic accident</td>
<td>Abdominal pain</td>
<td>20</td>
<td>3</td>
<td>1</td>
<td>CT, MRI</td>
<td>[18]</td>
</tr>
</tbody>
</table>

interval of the splenic rupture into the abdominal and pelvic was from 5 months to 32 years; its counterpart into the thorax was from 3 to 45 years [10]. Since the patient in this study had a history of a left thoracic and abdominal accident at his 7 years old, he underwent splenectomy, left nephrectomy and left diaphragmatic repair. A lapse of 35 years later, he was diagnosed with multiple splenosis in the right thorax and retroperitoneum accidentally, due to the suspicion of gallbladder stones. The male patient in this study was referred to our Hospital for the complaint of upper right abdominal pain. The clinical symptoms of splenosis depend on the specific position of the splenosis. For instance, gastroenterological splenosis may lead to gastrointestinal bleeding; multiple splenosis tubercles in abdominal cavity may result in intestinal adhesion followed by acute intestinal obstruction. Some splenic tissue growing in the thorax and lung parenchyma could cause chest pain, hemoptysis, cough, etc. However, splenic tissue growing in the liver, omentum, peritoneum, intestinal serosa, and pelvic organs, could lead to abdominal pain, intestinal obstruction, hemorrhage of the digestive tract as well as other different clinical manifestations [11, 12]. There have also been thoracic cases presenting with pleurisy and hemoptysis. Intra-abdominal cases have presented with bowel obstruction, and the compression of other neighboring organs [3]. However, there have also been a few ectopic spleens in patients with nonspecific clinical symptoms.

Ectopic spleens are easy to be misdiagnosed as other types of tumors. It is very important to distinguish splenosis from other benign or malignant tumors and CT and MRI play vital roles in diagnosing the condition. In general, MRI resolution provides a significantly improved resolution when compared to other imaging modalities. For this reason, MRI may be superior in diagnosing splenosis [3]. In this case, the lesions were hypodense on a plain CT scan, heterogeneous in the arterial phase, and homogeneous in the venous phase. In the MRI, the lesions were hypointense on T1WI, while hyperintense on T2WI and DWI. On GD-DTPA enhanced MRI, they showed uneven enhancement in the early arterial and arterial phases, but were homogeneously enhanced in the venous and delayed phases. The CT and MRI features of splenosis were similar to the primary spleen and consistent with those reported in the literature [12]. Therefore, based on the typical imaging findings, and the clinical history of previous splenic trauma or splenectomy, splenosis should be a given consideration. In addition to regular CT and MRI, specific inspection methods, such as the mononuclear phagocyte system specific MRI contrast agent SPIO [13] and percutaneous cytologic diagnosis, can also help to make a definitive diagnosis and to determine the best treatment plan. However, despite these advanced imaging techniques, a pathological diagnosis of splenosis is usually required, predominantly due to the possibility of malignancy or to preoperative diagnostic uncertainty. In the present case, the histopathological examination of splenosis showed red and white pulp structures, which was consistent with splenosis and helped confirm our diagnosis [14].

Since ectopic spleens are benign lesions, they are involved in humoral and cellular immunity, thereby compensating for the spleen’s function to a certain extent. The existences of ectopic splenic tissues are hypothesized to provide extra immunity for splenectomized patients. Researchers have strengthened this hypothesis through demonstrating an increase in IgM and IgG antibody levels in patients with splenosis [15]. Thus, ectopic splenic tissues should be carefully examined before surgery and those patients should be continuously followed instead of being given immediate treatment.
Splenosis in the right thorax and retroperitoneum

Once the diagnosis of splenosis is made during clinical practice, if the patient is asymptomatic, surgical resection is not necessary. But if the splenosis can cause acute symptoms including intestinal obstruction and gastrointestinal hemorrhage, surgical resection should be performed.

In summary, we report on the first case of combined thoracic and retroperitoneal splenosis in a male patient after trauma. The purpose of this article is to share experience in the diagnosis of multiple splenosis in the right thorax and retroperitoneum. This study could improve the understanding of splenosis in order to avoid unnecessary surgery in the future.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Huanzhou Xue, Department of Hepatobiliary Surgery, Zhengzhou University People’s Hospital, 7th Weiwu Road, Zhengzhou 450003, Henan, P. R. China. Tel: (86)-371-65580177; E-mail: xhzzzu@163.com

References