Reminder of important clinical lesson

The (re)generation of splenic tissue

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Summary

A 48-year-old man with a history of a traumatic splenic rupture followed by splenectomy at the age of 5 years was referred to the outpatient clinic with markedly elevated liver enzymes. He was diagnosed with alcoholic liver cirrhosis. Ultrasound of the upper abdomen revealed hepatomegaly and suggested a central mass in the liver. Subsequent MRI of the abdomen did not show a hepatic mass, but revealed multiple intraperitoneal and retroperitoneal ovoid structures with a maximum diameter of 3 cm. A peripheral blood smear did not reveal Howell-Jolly bodies suggesting intact splenic function. The diagnosis splenosis—that is, autotransplantation of splenic tissue after iatrogenic/traumatic rupture of the spleen—was considered and confirmed by SPECT-CT with technetium-99m (^{99m}Tc) labelled heat-denatured autologous red blood cells.

BACKGROUND

Splenosis is an acquired condition defined as autotransplantation of viable splenic tissue throughout different anatomic compartments of the body. Abdominal splenosis (AS) is estimated to occur in 65% of traumatic splenic ruptures with an average interval between trauma and splenosis of more than 10 years. The phenomenon is not well known among physicians. Usually, splenosis requires no treatment, but the diagnosis is warranted to exclude metastasised malignancy and to assess splenic function.

CASE PRESENTATION

A 48-year-old man was referred to the outpatient clinic with markedly elevated liver enzymes. His medical history included splenectomy due to a car accident at the age of 5 years old and a gastric ulcer. Furthermore, he had a habit of excessive alcohol abuse. The patient was diagnosed with alcoholic liver cirrhosis. Alpha-fetoprotein levels were marginally elevated (9 µg/litre). Ultrasound of the upper abdomen revealed hepatomegaly and suggested a central mass in the liver (not shown). MRI of the abdomen did not show a hepatic mass, but revealed multiple intraperitoneal and retroperitoneal structures with a maximum diameter of 3 cm and ovoid of shape (figure 1). A peripheral blood smear revealed no Howell-Jolly bodies (figure 2).

INVESTIGATIONS

Laboratory results: alanine transaminase 100 IU/ml (<45 IU/ml); aspartate transaminase 128 IU/ml (<40 IU/ml); γ-glutamyl transpeptidase 664 IU/ml (<60 IU/ml); alkaline phosphatase 302 IU/ml (40–120 IU/ml); bilirubin total 128 µmol/litre (<17 µmol/l); bilirubin direct 98 µmol/litre (<7 µmol/litre); partial thromboplastin time (PTT) 11.2 s (9.7–11.6 s); activated PTT 29.1 s (22–30 s); albumin 38 g/litre (35–50 g/l); glucose 5.6 mmol/litre (4.1–5.6 mmol/litre); antithrombin III 66% (80–140%); immunoglobulin A 6.5 g/litre (0.7–4.0 g/litre); α-fetoprotein 9 µg/ml (<7 µg/l).

A MRI of the abdomen showed hepatomegaly and multiple (>20) intraperitoneal and retroperitoneal ovoid structures with a maximum diameter of 3 cm (figure 1). The differential diagnosis included metastasised malignancy and, with a medical history of traumatic splenectomy, AS.

A peripheral blood smear revealed no Howell-Jolly bodies (figure 2) implying intact splenic function.

A single photon emission computed tomography (SPECT) with ^{99m}Tc-labelled heat-denatured autologous red blood cells (figure 3) showed markedly elevated uptake of heat-damaged red blood cells (cross) in multiple (>20) intraperitoneal and retroperitoneal masses depicted on low-dose CT, corresponding to lesions visualised on ultrasonography and MRI and confirming the diagnosis AS.

OUTCOME AND FOLLOW-UP

The central mass visualised by ultrasound suggested hepatocellular carcinoma. This was not confirmed by MRI and was, thus, most probably an artefact due to the inhomogeneous aspect of the liver parenchyma. However, multiple intraperitoneal and retroperitoneal ovoid structures were visualised, which could be attributed to AS (figure 1). Indeed, the peripheral blood smear revealed no Howell-Jolly bodies (figure 2) implying normal splenic function. The diagnosis was confirmed by SPECT with ^{99m}Tc-labelled heat-denatured autologous red blood cells (figure 3). SPECT did not reveal extra AS. The condition was left untreated, no further (invasive) diagnostic tests were performed and the patient was not vaccinated against encapsulated bacteria. The work-up for the alcoholic liver cirrhosis included a gastroscopy, which showed a congestive gastropathy and a mild erosive gastritis, but no oesophageal varices. Treatment with β-blockers and proton pump inhibitors was initiated. Cirrhosis was confirmed using transient elastography (Fibroscan; Artemis Medical, Kent, UK; not shown).

Figure 1 MRI of the abdomen. Representative transverse sections (A, B and C) corresponding to the sections in figure 3 are depicted.

DISCUSSION

AS is a condition in which autotransplantation of splenic tissue occurs after iatrogenic/traumatic rupture of the spleen.¹ It does not occur in individuals who undergo uncomplicated splenectomy for haemato-oncological conditions. AS differs from accessory spleens that arise during embryogenesis. In AS, depending on the amount of splenic pulpa that is spread throughout the abdominal cavity, several to hundreds of foci can be indentified. In addition, intrathoracic

Figure 2 Peripheral blood smear.

Figure 3 SPECT low-dose CT of the abdomen 30 min after (re)injection of 80 MBq ^{99m}Tc-labelled heat-denatured autologous red blood cells. Representative transverse low-dose CT slices (left panel) and corresponding SPECT fusion slices (middle panel) are shown representing different levels in the abdomen (A, B and C). Slice reference is marked on the maximum intensity image (right panel). There is markedly elevated uptake of heat-damaged red blood cells (cross), diagnostic for splenosis in multiple (20) intraperitoneal and retroperitoneal masses depicted on low-dose CT and corresponding to lesions visualised on ultrasonography and MRI (data not shown and figure 1).

and even intracerebral splenosis have been described.^{2 3} The size of the splenic lesions varies from several millimetres to approximately 3 cm, with blood supply being the limiting factor for unrestrained growth. It occurs more often

in males than in females, reflecting behavioural rather than biological differences, and it is estimated to arise in 65% of traumatic splenic ruptures although not diagnosed until 10 years after splenectomy. The borne again spleen can be functional as has been suggested by mouse⁴ and human studies.⁵ Like in our patient, splenosis is often asymptomatic and is diagnosed by coincidence. However, infarction, bleeding or obstruction can lead to a symptomatic presentation. The diagnosis is suggested by a peripheral blood smear without Howell-Jolly bodies or 'pitted cells' despite the absence of a spleen and is confirmed by imaging studies.¹⁻⁵ Several imaging techniques are available; however, SPECT with ^{99m}Tc-labelled heat-denatured autologous red blood cells has been suggested as the 'gold standard'.⁶⁻⁷ Usually AS requires no treatment, but the diagnosis is warranted to exclude metastasised malignancy and to assess splenic function.

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Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Fremont RD**, Rice TW. Splenosis: a review. *South Med J* 2007;**100**:589–93.
2. **Rickert CH**, Maasjosthusmann U, Probst-Cousin S, *et al*. A unique case of cerebral spleen. *Am J Surg Pathol* 1998;**22**:894–6.
3. **Singh P**, Munn NJ, Patel HK. Thoracic splenosis. *N Engl J Med* 1995;**333**:882.

Learning points

- Splenosis is a condition in which autotransplantation of splenic tissue occurs after iatrogenic/traumatic rupture of the spleen.
- It is estimated to occur in 65% of all cases of traumatic/iatrogenic splenic rupture.
- Diagnosis can be suggested by a peripheral blood smear (absence of Howell-Jolly bodies despite splenectomy) and is confirmed by imaging studies—for example, SPECT with ^{99m}Tc-labelled heat-denatured autologous red blood cells.
- Usually AS requires no treatment, but the diagnosis is warranted to exclude metastasised malignancy and to assess splenic function.

4. **Dickerman JD**, Horner SR, Coil JA, *et al*. The protective effect of intraperitoneal splenic autotransplants in mice exposed to an aerosolized suspension of type III *Streptococcus pneumoniae*. *Blood* 1979;**54**:354–8.
5. **Pearson HA**, Johnston D, Smith KA, *et al*. The born-again spleen. Return of splenic function after splenectomy for trauma. *N Engl J Med* 1978;**298**:1389–92.
6. **Hagan I**, Hopkins R, Lyburn I. Superior demonstration of splenosis by heat-denatured Tc-99m red blood cell scintigraphy compared with Tc-99m sulfur colloid scintigraphy. *Clin Nucl Med* 2006;**31**:463–6.
7. **Yamine JN**, Yatim A, Barbari A. Radionuclide imaging in thoracic splenosis and a review of the literature. *Clin Nucl Med* 2003;**28**:121–3.

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