CASE REPORT

Splenic peliosis: an unusual entity

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ABSTRACT. This case of splenic peliosis, describes a rare condition when it presents in the spleen, and is better understood when it occurs in the liver. However, the ultrasound and CT features have a wide differential diagnosis including more common aetiologies, of either a vascular, infective or neoplastic nature, which should be considered. Peliosis is an important condition to be aware of because rupture of the blood-filled cysts on the spleen surface can lead to haemorrhagic peritonitis and ultimately be fatal.

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A 56-year-old female shop assistant presented acutely with abdominal pain, anorexia and weight loss. Her past medical history included pneumonia, hypothyroidism and irritable bowel syndrome. She was not taking any prescribed medication.

An ultrasound of the abdomen was performed, which showed a spleen that was normally sized but that had heterogeneous echotexture, and multiple fluid-filled and echogenic areas. There was also an echo-poor area of 2 cm in the lower pole of the spleen (Figure 1). The other solid organs had normal sonographic appearances.

The patient underwent a contrast-enhanced CT scan of the abdomen and pelvis, which showed multiple round focal areas of decreased enhancement within the spleen (Figures 2 and 3). Initially, these were thought to be due to infiltration of lymphoma. There was no other evidence of nodal disease but the patient was found to have a sigmoid carcinoma and underwent curative surgery including splenectomy. She made a full post-operative recovery and started adjuvant chemotherapy. Several follow-up CT scans have not shown any recurrence.

Histopathology demonstrated a normal-sized spleen, involving numerous cystic lesions lined by fibrous tissue, and containing cholesterol crystals and fibrinoid debris with adjacent giant-cell reaction to foreign bodies. No epithelial or endothelial lining was identified. There were also dilated blood-filled vascular spaces (Figure 4). There was no evidence of lymphoma or metastatic disease. Stains for fungal infection were negative. The disease appearances were consistent with peliosis of the spleen. The sigmoid specimen confirmed a moderately differentiated adenocarcinoma with one out of 21 nodes was positive.

Discussion

Peliosis is an unusual benign disorder characterised by the presence of irregular cystic blood-filled cavities. Fewer

than 100 cases have been described in the literature. The term originates from the Greek *pelios*, meaning dusky or purple, which arose from the macroscopic appearance of the lesion. However, this condition is now defined histopathologically. Documented causal associations include the oral contraceptive pill, anabolic steroids, advanced tuberculosis and human immunodeficiency virus (HIV). Peliosis has also been associated with chronic haematological disorders (Hodgkin's disease, myeloma and aplastic anaemia), disseminated cancer, previous thorium dioxide contrast injection and certain viral infections. In all these cases, both the liver and the spleen are usually involved. When peliosis occurs in isolation in the spleen, it is usually asymptomatic and detected incidentally on imaging for other indications or at autopsy. As well as the liver and spleen, organs including lymph nodes, bone marrow, lungs, pleura, kidneys, adrenals, stomach and ileum can be affected [1]. Peliosis hepatitis has been observed in two forms. In the parenchymal form the cysts are lined by hepatocytes, whereas in the phlebotatic form, the cysts are lined by endothelial cells. However, only Yanoff and Rawson's group [2] have described this and it has not been possible to reproduce these findings.

The aetiology of peliosis hepatitis is unclear but several processes have been postulated: outflow obstruction of blood flow at a sinusoidal level, hepatocellular necrosis or direct lesions of the sinusoidal barrier [3]. Bagheri and Boyer [4] suggested that the initial process is focal cell necrosis that leads to destruction of the reticulum framework, which allows formation of a cyst from the inflow of blood from adjacent sinusoids.

In HIV patients, a *Rickettsia*-like organism, *Rochalimae henselae*, is the causative agent of peliosis hepatitis and hence erythromycin therapy improves liver function and symptoms. If left untreated, peliosis hepatitis may be fatal in these cases. Cessation of oral contraceptives or oral steroids may also cause regression of peliosis hepatitis.

Complications of hepatic peliosis include hepatocellular dysfunction, portal hypertension and life-threatening haemorrhage due to rupture of a solid organ [5]. It is important that the diagnosis is made in patients who have factors that predispose them to these complications. The complications

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Figure 1. Ultrasound image demonstrating a normal size spleen, with hypoechoic areas in the lower pole, the larger of which measures 2 cm. The white arrow indicates a hypoechoic area within the spleen.

of splenic peliosis include rupture of the haemorrhagic cysts, leading to life-threatening intraperitoneal haemorrhage. This can occur spontaneously or secondary to minimal trauma. Percutaneous aspiration, confirming the presence of black blood and excluding pus, can provide strong evidence to support the diagnosis but is hazardous. Therefore, recognition of the imaging features without the use of intervention is optimal. The definitive diagnosis and treatment, however, usually involves splenectomy.

Pathologically, peliosis can be differentiated from haemangioma by blood-filled spaces that are haphazardly scattered in the red pulp, with preferential involvement of the parafollicular areas of the spleen. Ultrasound appearances include an echogenic mass with numerous poorly defined foci of varying hypoechogenicity. The larger lesions demonstrate moderate posterior acoustic enhancement. The differential diagnosis for these sonographic findings is shown in Table 1 [6]. In the context of peliosis hepatis, contrast-enhanced ultrasound (with Levovist) demonstrates transient “fast surge” central echo enhancement with no centripetal enhancement on delayed imaging [2].

On non-contrast-enhanced CT images, the appearances are of a hypoa attenuating, multiloculated lesion with well-defined septa. On contrast-enhanced CT images, the lesion demonstrates significant enhancement with loss of defini-

Figure 2. Portal venous phase axial CT images showing areas of decreased attenuation within the spleen.

Figure 3. Portal venous phase axial CT images showing areas of decreased attenuation within the spleen.

tion of the lobules and septa. Fluid–fluid levels can also be seen, and these are thought to represent haematocrit levels that demonstrate enhancement in their dependent portions. Calcification or extra-capsular extensions tend not to be seen unless the lesions rupture, in which case subcapsular haematoma may be seen in association with intraperitoneal haemorrhage [7]. MRI findings were described by Maves et al [5] as mixed signal on T_2 weighted sequences attributed to the presence of deoxyhaemoglobin and methaemoglobin. Peliosis hepatis is generally associated with increased signal on T_2 weighted sequences and variable signal on T_1 weighted and proton-density sequences, reflecting various stages of subacute haemorrhage. None of the lesions enhances after the administration of gadopentatetate dimeglumine, but they are seen as low-signal foci within enhancing hepatic and splenic parenchyma.

The radiological differential diagnosis of peliosis includes haemangiomatosis, lymphangioma and angiosarcoma. In conclusion, splenic peliosis is a rare benign disorder seen in the context of other chronic contributing factors (HIV, haematological factors, steroids and the oral contraceptive pill). Life-threatening haemorrhage can occur from the rupture of surface lesions, either spontaneously or as a result of minor trauma. An awareness of

Figure 4. Histopathology slide from the splenectomy specimen illustrating the vascular pools within the splenic white substance.

Table 1. Classification of cystic masses of the spleen (taken from Urrutia et al, 1996) [6]

Classification	Cystic entity
Congenital	True cyst
Inflammatory	Pyogenic abscess, echinococcal cyst, fungal abscess
Vascular	Infarction, peliosis
Posttraumatic	Haematoma, false cyst
Neoplastic	
Benign	Haemangioma, lymphangioma
Malignant	Lymphoma, metastasis

this rare condition may suggest the diagnosis and lead to the definitive management of splenectomy.

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