Rare disease

Splenic peliosis: an incidental finding

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Summary

Peliosis is a rare condition of multiple blood-filled cysts within the parenchyma of a solid organ. Splenic peliosis is an extremely rare entity, although there is often a risk of spontaneous rupture. We present a case where our otherwise fairly healthy patient presented us with a diagnosis dilemma and was then found to have splenic peliosis histologically post-splenectomy. We discuss the current available literature and emphasise the importance of awareness of such a condition.

BACKGROUND

Peliosis, the Greek word for livid spot or purpura, is a rare disorder of undetermined pathogenesis characterised by the gross appearance of blood-filled cysts in solid parenchyma.¹ This condition is most commonly seen in the liver; however, very rarely, it can appear in other organs—for example, spleen, kidneys and lymph nodes. We present a case of an incidental finding of an isolated splenic peliosis in which the patient presented with symptoms suggestive of malignancy. This lesion had inconclusive radiological findings and the patient eventually underwent a diagnostic and therapeutic (retrospectively) splenectomy.

CASE PRESENTATION

A 67-year-old woman presented to general surgical clinic following a 3–4 month period of right-sided abdominal

pain. She first noticed the pain, with an episode of bloody diarrhoea, while she was on a holiday. She was diagnosed with biliary colic as confirmed by an ultrasound scan showing 1.6 cm calculus within the gallbladder. She was also treated with antibiotics for acute diverticulitis at the same instance. Despite the aforementioned treatment, she continued to have similar pain intermittently, which apparently subsided after bowel motions. There was no further episode of rectal bleeding reported but she claimed to have occasional faecal incontinence due to loose stools. She did not report any appetite loss; however, she did mention some weight loss noticeable on daily dressing. Her past medical histories include microcytic anaemia (in 2009) and total abdominal hysterectomy (in 1986). She is on medications for her hypertension and chronic back pain due to disc degeneration. On initial examination, she had marked

Figure 1 A splenic lesion suspicious of metastasis of unknown primary.

Figure 2 Blood-filled spaces with haphazard arrangement and well delimitation from the surrounding parenchyma.

tenderness at the right side of her abdomen but otherwise no other significant finding.

INVESTIGATIONS

At this point, with a suspicion of a possible intestinal malignancy, we organised a colonoscopy and a CT scan of her abdomen. CT showed a splenic lesion suspicious of metastasis (figure 1). Unfortunately, her colonoscopy was unsuccessful due to poor bowel preparation; therefore, we organised a CT colonography for her as well as an oesophagogastroduodenoscopy (OGD) in search for a gastrointestinal (GI) primary malignancy. In the meantime, her pain had become more generalised and involved the left side of her abdomen associated with appetite and weight loss. Both CT colonography and OGD were reported as normal. She then had a MRI scan that confirmed a solid lesion in her spleen of unknown origin but highly suspicious of malignancy.

DIFFERENTIAL DIAGNOSIS

A few differential diagnoses were postulated during the patient's initial presentation of symptoms such as right upper quadrant tenderness and bloody diarrhoea—that is, biliary colic, acute diverticulitis. However, based on the unusual presentation of continuing pain with GI symptoms, malignancy has to be excluded. All the symptoms were rather non-specific and could be misleading, bearing in mind that peliosis patients are usually asymptomatic. We could only presume the patient had a concurrent biliary colic. The diagnosis of peliosis was difficult even following radiological investigations. Certainly, differential diagnoses involving haematological malignancies such as myeloma and Hodgkin's disease would have to be considered based on CT findings.

TREATMENT

After discussion with the patient, diagnostic and, potentially, therapeutic open splenectomy was agreed and performed. Prophylactic vaccines for post-splenectomy infection were in place.

OUTCOME AND FOLLOW-UP

Macroscopically the splenic lesion consisted of irregular haemorrhagic cavities. Histological examination revealed blood-filled spaces with haphazard arrangement and well-delimitation from the surrounding parenchyma (figure 2). Apart from blood, the other constituents of the lesion were eosinophilic acellular material reminiscent to fibrin (figure 3A–C) and scanty lymphoid tissue. There was no evidence of lymphoma, angiomatous lesions or amyloidosis. There were no other organs found to be involved and the patient remains well to date.

DISCUSSION

'Peliosis' was first used by Wagner in 1861 to describe the gross appearance of lesions found on the cut surface of a liver affected by the condition.² It is mostly seen in organs belonging to the mononuclear phagocytic system—for example, liver (most common), spleen, bone marrow and lymph nodes—but can also appear in the lungs, kidneys and parathyroid glands.¹ Splenic peliosis may be idiopathic or associated with infections such as tuberculosis, haematological diseases such as myeloma and myelofibrosis, and with the use of androgenic steroids and erythropoietin.³ However, in our patient, no predisposing factor can be associated.

Pathogenesis of peliosis remains uncertain. Some suggest that there is more than one way of pathogenesis (eg, congenital malformation of vessels, acquired vascular disorder triggered by toxic noxae, angiogenesis on the benign end of

Figure 3 (A) Other constituents of the lesion were eosinophilic acellular material reminiscent to fibrin and scanty lymphoid tissue. (B) $\times 40$ magnification. (C) $\times 100$ magnification.

spectrum of neoplastic vascular changes).¹ Peliosis can usually be recognised macroscopically. The splenic surface may show nodular areas, but it is usually the cut surface that demonstrates the blood-filled cavities usually in the parafollicular area of the red pulp.^{1 4}

Isolated peliosis of the spleen is extremely rare occurring in less than 1% of cases at autopsy.¹ Our case is somewhat unusual as the patient was symptomatic for other diseases and it was not until an incidental finding on a CT scan that we picked up this lesion. Furthermore, due to the undistinguished nature of the lesion to malignancy, the patient had to undergo a collection of investigations and, certainly, posed a challenge for us in reaching a definitive diagnosis. In addition, more invasive diagnostic tools,^{5 6} such as imaging guided or laparoscopic biopsy, may have caused serious complications that we were attempting to prevent—for example, spontaneous haemorrhage.⁷ However, it is important to explore for other organ involvement.

Histologically, differential diagnoses considered for the lesion in our patient include angiomatous lesions (littoral cell angioma, lymphangioma, angiosarcoma, bacillary angiomatosis and sclerosing angiomatoid nodular transformation). However, all of these were excluded based on the fact that there was no vascular proliferation demonstrated within this nodule. There was no cytological atypia too. Amyloidosis was also considered but ruled out as the amorphous eosinophilic material (though reminiscent to amyloid) was negative with Congo red.

Currently, there are no guidelines describing the surgical management of patients with an incidental finding of splenic peliosis as well as the role of elective splenectomy in such patients. The diagnosis is of such a rare entity; therefore, clinicians should retain a high index of suspicion for

the presence of this condition and be aware of the potentially fatal consequences.

Learning points

- ▶ This is a rare condition, which requires a high index of suspicion when common causes of symptoms have been ruled out.
- ▶ Occasionally, one may come across a dilemma in reaching firm diagnosis despite history, clinical examination as well as radiological findings. This poses a challenge in general surgery when decision making becomes crucial as to whether to pursue an operative exploration and intervention.

Competing interests None.

Patient consent Obtained.

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