On-line Case Report

'Not just another appendicitis!' – a case report of acute abdominal pain caused by splenic rupture secondary to isolated splenic peliosis

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ABSTRACT

We present the case of a 31-year-old man admitted with acute abdominal pain who was subsequently found to have a ruptured spleen. A splenectomy was performed as an emergency and he was discharged from hospital 4 days later. Histological analysis revealed isolated splenic peliosis as the underlying condition predisposing to his splenic rupture.

Keywords: Acute abdominal pain – Ruptured spleen – Splenectomy

Case history

A previously healthy 31-year-old Polish male presented to the accident and emergency department with sudden-onset of abdominal pain. The pain followed an alcohol binge 8 h prior to his presentation and was generalised and constant in nature. There was no history of trauma related to his excessive alcohol intake. There was one episode of vomiting but no other bowel or urinary symptoms. He also complained of pain in his left shoulder.

On physical examination, the abdomen was soft and non-distended. Tenderness was elicited all over the abdomen but was more severe in the epigastric region and right loin with mild guarding. There was no rebound

tenderness or rigidity. There were no masses or organomegaly to find. He was afebrile and no abnormalities were detected in the cardiovascular and respiratory examinations. His blood pressure was 130/85 mmHg and heart rate was 83 bpm.

Laboratory tests demonstrated a raised white blood cell count of $13.7 \times 10^9/l$ (normal range, $4.5\text{--}11.0 \times 10^9/l$) and a C-reactive protein of 16 mg/l (normal range, 0–8 mg/l). Renal, liver function tests, serum amylase and clotting profile were all within normal limits. An erect chest and abdominal radiograph did not reveal any abnormalities. Abdominal ultrasound showed a large amount of abdominopelvic free fluid. The liver was normal in echogenic texture with no focal lesions, and the pancreas and kidneys were also normal. The spleen was

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Figure 1 Post-splenectomy.

described as being normal and non-enlarged. A blind-ending tubular structure was visualised to the right of the midline just above the bladder. Although not thick-walled, this was reported as representing an abnormal appendix. At this point, the differential diagnoses were perforated acute appendicitis, a perforated viscus or acute pancreatitis.

The patient was rehydrated. A decision was made to do a diagnostic laparoscopy with or without appendicectomy. Laparoscopy revealed a large amount of fresh blood and clots were seen in the peritoneal cavity, which obstructed the view. No clear source of bleeding could be identified. A midline exploratory laparotomy was performed and 1.8 l of blood was evacuated. A haemorrhagic cyst was identified at the hilum of the spleen and a total splenectomy was performed. The bowel, liver and appendix were all normal. The patient made an uneventful recovery and was discharged 4 days later.

Macroscopically (Fig. 1), the spleen was 15 × 9.5 × 5 cm and weighed 250 g. The cut surface revealed a 5 × 3 × 2.5 cm, well-circumscribed, haemorrhagic area with satellite haemorrhagic nodules. No other focal lesions or hilar lymphadenopathy was identified.

Histologically, the lesions corresponded to irregular blood-filled spaces of variable calibre (Fig. 2, left), some with partial thrombosis. Also present was normal splenic

Figure 2 Haematoxylin and eosin stained tissue section of the spleen showing the peliotic lesion. Original magnification ×20.

red and white pulp (Fig. 2, right). Perifollicular flares were seen near the lesion but the uninvolved spleen appeared normal and there was no evidence of malignancy. Although there was some fibrosis between the spaces the distinct lack of vascular wall structures (muscle and elastic) and the lack of an endothelial lining (confirmed by immunostains for CD31, CD34 and CD8) favour the diagnosis of peliosis of the spleen rather than a haemangioma or AV-malformation.

Discussion

Peliosis is a term derived from the Greek *pelios*, which means 'blue-black'. It was first used by Wagner in 1861 when referring to the colour of liver parenchyma with peliosis.¹ It is a relatively rare pathological entity characterised by the presence of multiple cyst-like, blood-filled cavities within the parenchyma of solid organs. Most commonly affecting the liver, we present a case of isolated splenic peliosis, which is an even rarer phenomenon.

The aetiology of peliosis is, as yet, unknown. A review of the literature demonstrates that splenic peliosis has been associated with chronic alcoholism,² infectious agents (hepatitis B and C,³ HIV,³ *Bartonella henselae*), haematological disorders (immune thrombocytopenia,⁴ Waldenstrom's macroglobulinaemia,⁵ chronic myelomonocytic leukemia⁶) and drugs (corticosteroids,⁷⁻¹⁰ danazol,^{4,11} erythropoietin¹²), as well as other rarer medical anomalies.

Peliosis is often asymptomatic and in the past was commonly diagnosed only on autopsy.³ Our patient had a spontaneous splenic rupture and this was found to be a relatively common mode of presentation in the literature,

with a large proportion of these cases being fatal. Fortunately, our patient was a previously healthy young man and was able to compensate for the large intravascular volume loss prior to diagnosis. Other patients with splenic peliosis have presented with pain in the left hypochondrium^{10,11,13} and splenomegaly,¹⁴ as well as with features of any of the associated medical conditions.

With the evolution and wider availability of radiological imaging, peliosis is rapidly becoming an incidental finding when investigating for other conditions.¹⁰ However, due to the rarity of splenic peliosis, many of the characteristic imaging results are deduced from peliosis hepatis. The radiological differential diagnosis of peliosis includes hemangiomatosis, lymphangioma, and angiosarcoma.¹⁵ The clinical significance of this differentiation lies in the potential of peliotic lesions to rupture and cause life-threatening intraperitoneal hemorrhage.^{2-4,6,9,16-19} Due to the risk of rupture, needle biopsy of a suspected peliotic lesion is ill-advised and further enhances the need for sensitive radiological tests. On ultrasound, peliotic lesions may be hypo-echoic or hyperechoic lesions without sharply demarcated borders.¹⁵ No such lesions were demonstrated on ultrasound imaging of our patient. Most commonly, peliosis manifests as multiple small, well-defined hypo-attenuating lesions on non-contrast enhanced computed tomography (CT). The introduction of contrast reveals slow centripetal enhancement of the lesions.¹⁵ CT scan was not performed on our patient in the acute setting; however, a subsequent abdominal CT scan revealed no peliotic lesions in other organs including the liver. The use of magnetic resonance imaging in detecting splenic peliosis has not been evaluated in recent literature.

Peliosis needs to be differentiated from other cystic lesions of the spleen. Macroscopically, these may be true congenital cysts, benign cysts (cavernous haemangioma), malignant cysts (metastases, lymphoma), inflammatory (fungal or pyogenic abscess), or vascular cysts. Histologically, peliotic lesions are vascular cysts typically filled with blood. These have been described with and without an endothelial lining.²⁰

The treatment of peliosis has mostly been confined to the medical management of its associations such as treating infection or ceasing steroid use. The definitive diagnosis and treatment of peliosis of the spleen is currently with splenectomy. However, the existing literature does not explore the surgical management of patients in whom an incidental diagnosis of splenic peliosis has been made. Splenectomy has been advocated to prevent organ rupture in conditions such as splenic sarcoidosis,²¹ but the role of elective splenectomy in splenic peliosis is not known. The introduction of

laparoscopic splenectomy has significantly reduced morbidity when compared with open surgery.²² Furthermore, splenic autotransplantation has been reported as a viable method of avoiding the problems associated with asplenia,²³ though this may not be possible in an emergency splenectomy, or where there is uncertainty of diagnosis or a suspicious appearing lesion.

Conclusions

This case promotes the consideration and early recognition of spontaneous splenic rupture as a cause of abdominal pain in patients presenting to the emergency department. It also highlights the ability of the younger, physiologically fit patient to maintain cardiovascular stability in spite of significant intravascular depletion.

On further investigation and retrospective review of the ultrasound scans initially performed, despite the size of the splenic cyst, the large amount of abdominopelvic free fluid (subsequently found to be blood) partially obscured the view of the hilum of the spleen. In addition, the scan was performed on-call by a junior radiology registrar and this may explain why the cyst was not initially identified. We would still advocate, however, an ultrasound scan as a first-line investigation of abdominal pain in a fit, young, haemodynamically stable patient such as ours.

Despite the rarity of isolated splenic peliosis, the literature review highlights the importance of its recognition as a possible cause for splenic rupture. Efforts in elucidating the radiological features of splenic peliosis will enable us to identify this commonly asymptomatic, but potentially fatal, condition more readily and possibly advocate elective splenectomy in these cases to avoid the late-stage presentation and its potential consequences.

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