A case of splenic peliosis associated with pathologic rupture
有關紫癜的病理性脾臟破裂個案

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A patient walked into the accident and emergency department with spontaneous onset of left-sided abdominal pain. Physical examination suggested the presence of an acute abdominal condition. He was admitted to the surgical ward with a provisional diagnosis of perforated peptic ulcer. Emergency laparotomy revealed haemoperitoneum caused by rupture of the spleen. Pathologic examination confirmed the rare diagnosis of splenic peliosis. Emergency physicians should be aware of the possibility of atraumatic splenic rupture as a cause of spontaneous haemoperitoneum, bearing in mind that isolated splenic peliosis (also known as peliosis lienis) is one of the causes of pathologic splenic rupture. (*Hong Kong J. Emerg. Med.* 2007;14:174-178)

一名病者因左腹自發地開始作痛而步進急症室。身體檢查顯示腹部有急性狀況，他臨時的診斷為消化性潰瘍穿孔而入住外科病房。緊急剖腹揭露因脾臟破裂引致腹腔積血。病理檢驗證實診斷為罕有的脾紫癜。急症科醫生應警覺非創傷性脾臟破裂引致自發性腹腔積血的可能性，並提醒單一的脾紫癜是病理性脾臟破裂的成因之一。

Keywords: Haemoperitoneum, lacerations, peliosis lienis, splenic rupture, spontaneous rupture

關鍵詞：腹腔積血、裂傷、脾紫癜、脾臟破裂、自發性破裂

Introduction

Splenic rupture is common after blunt abdominal trauma. Rarely, splenic rupture is not associated with trauma and is termed "spontaneous" rupture. The majority of spontaneous splenic rupture is associated with a diseased spleen. "True spontaneous rupture" in a normal spleen is an extremely rare event.¹

Peliosis is a morphological condition with blood filled spaces in solid organs, most frequently occurring in the liver. Splenic peliosis (peliosis lienis) is peliosis occurring in the spleen. Patients with this condition are often asymptomatic. However, it can be lethal if spontaneous organ rupture occurs.

Case history

A 48-year-old male construction site worker walked into our accident and emergency department in the early afternoon in April 2002 with a chief complaint of left-sided abdominal pain, bilateral earache and dizziness. In the nursing triage station, his blood pressure was 110/52 mmHg with a regular pulse rate of 109/min. He was afebrile with oxygen saturation (SpO₂) of 100% while breathing room air. He was triaged as a semi-urgent case (Category IV).

Later on, he told his attending doctor that he had left upper quadrant abdominal pain, which was exacerbated by deep breathing. There was also bilateral...
tinnitus and dizziness without vertigo. No history of preceding trauma could be elicited on direct questioning. His past health was unremarkable except for occasional dyspepsia. Positive physical signs on examination were limited to the abdomen only. There were generalised abdominal tenderness, guarding, rigidity and rebound tenderness. No abnormal abdominal pulsation was detected. The bowel sound was sluggish. Per-rectal examination was normal. His repeated blood pressure was 107/76 mmHg. An intravenous infusion line was set up and he was kept nil by mouth. His supine abdominal X-ray was unremarkable. However, there was suspicious lucency beneath the right hemidiaphragm on the erect chest X-ray. He was then admitted to the surgical ward for suspected perforated peptic ulcer (PPU).

Surgical assessment confirmed the presence of peritoneal signs. Emergency laparotomy was arranged for suspected PPU. When the peritoneal cavity was opened up, there was about 1,500 ml of blood with clots. The spleen was mildly enlarged with active oozing from two deep lacerations. The liver was mildly enlarged with a firm, deep-seated nodule 1.5 cm in diameter in segment VIII. The pancreas, bowels and great vessels were all normal. There was no intra-abdominal lymphadenopathy. Splenectomy was performed and the spleen was sent for pathologic examination, which later reported as splenic laceration and peliosis.

He probably had pre-existing thalassaemia trait with initial haemoglobin of 11.5 g/dL, MCV 65 fl, MCH 20.8 pg and MCHC 31.9 g/L. Two units of blood were transfused after the operation. Initial white cell count was 22.3 x 10^9/L and platelet count was 235 x 10^9/L. Baseline coagulation studies, liver and renal functions, and serum levels of amylase, alpha-fetoprotein, and carcino-embryonic antigen were all normal. He made an uneventful recovery and was discharged one week later.

Computed tomography (CT) scan of the abdomen was done on the day of discharge. Two tiny hypodense lesions were seen in the right and left hepatic lobes and were most likely hepatic cysts. A small nodular calcification – likely granuloma, was also seen in the right hepatic lobe. There was no interval change when the scan was repeated six months later.

During the subsequent outpatient follow-up visit, the patient recalled that the abdominal pain and sweating started after the bus in which he was travelling bumped over an uneven surface of the road.

**Discussion**

Rupture of the spleen usually occurs after blunt abdominal trauma and the diagnosis is usually straightforward. Atraumatic rupture of the spleen is rupture without any obvious history of trauma. It is uncommon and potentially fatal if left unrecognised and untreated. It is sometimes referred to as spontaneous rupture of the spleen. This term is often misleading ever since the first case was reported by Atkinson in 1874. The spleen can rupture in the following circumstances: (a) trauma to a normal spleen; (b) trauma to a diseased spleen; (c) spontaneous rupture of a normal spleen and (d) spontaneous rupture of a diseased spleen. Some authors have suggested that the atraumatic rupture of a diseased spleen be termed pathologic rupture while spontaneous rupture should be reserved for rupture of a healthy spleen without overt trauma and this has also been described as idiopathic fracture of the spleen.

The incidence of pathologic rupture of the spleen is unknown. Debnath and Malerio quoted a Medline search in which 352 cases had been reported between 1966 and 2000. The spleen is a friable and vascular organ. If the spleen is diseased or enlarged, trivial trauma may result in rupture. Table 1 shows the common causes of pathologic rupture of the spleen. Infectious mononucleosis is the commonest cause in the western world while malaria is the commonest cause of pathologic splenic rupture in the tropics. Amongst the haematological malignancies, pathologic rupture of the spleen is most commonly seen in acute leukaemia.
Peliosis was first described in the liver (peliosis hepatis) in 1916. Most cases of splenic peliosis are associated with peliosis hepatis. Isolated splenic peliosis is rare. Tada, Wakabayashi and Kishimoto found splenic peliosis in 10 out of 1,200 cases autopsied from 1977 through 1980. Eight such cases had no peliosis of the liver. They also noticed that the parafollicular areas were the most common sites of the lesions histologically, and this feature seemed to be important for the histologic differentiation of peliosis from simple dilatation of splenic sinuses resulting from passive congestion.9

In 2004, Tsokus and Puschel reported two autopsy cases of isolated splenic peliosis and there were only 33 cases reported in the literature before then.10 According to Celebreze in 1998, there were only seven documented cases of spontaneous splenic rupture that was associated with isolated splenic peliosis.11

Isolated splenic peliosis has been reported in various parts of the world including but not exclusive to North America,12-14 Europe,15,16 Japan,17,18 India19,20 and Hong Kong.21 Postulated aetiologies and clinical associations include viral and chemical exposure, pregnancy15 immune complex deposition,22 intake of danazol,23 steroid17,18,24-26 and oral contraceptive pills,27 and chronic debilitating diseases such as haematological malignancies,28,29 end-stage renal failure (on continuous ambulatory peritoneal dialysis and erythropoietin),30 cirrhosis of liver,30 post-liver transplantation,31 AIDS patients and drug addiction with chronic alcoholism.32

The clinical significance of splenic peliosis lies in two aspects, namely: (a) its potential association with spontaneous splenic rupture, often after trivial injury, with life-threatening intraperitoneal haemorrhage,33 and (b) the rarely reported resolution after discontinuation of the culprit medication.10 Once splenic peliosis is diagnosed, we should pursue to detect its presence in other organs, mainly the liver; and to establish a possible cause (e.g. steroid) or an underlying serious condition such as immunodeficiency or malignancy.27

In forensic medicine, one may overlook splenic peliosis as the cause of spontaneous splenic rupture with intra-abdominal haemorrhage, leading to the false accusation of a violent death; and we may misinterpret the macro-morphological appearance of this rare disease, in the absence of splenic rupture, as a result of blunt trauma.10 There was also a report of splenic peliosis imitating metastasis on CT scan of the abdomen.34

Concerning the management of splenic haemorrhage, for those situations caused by blunt abdominal trauma, there is a shift towards non-operative management.
if the patient remains haemodynamically stable. However, for cases of pathologic rupture of spleen, there is much debate. Guth et al reported 11 cases of successful non-operative management of patients with trauma to diseased spleen or pathologic rupture of the spleen.35 Most surgeons still prefer total splenectomy if there is any concern that splenic haemorrhage is secondary to a pathologic origin.36

Conclusion

Peliosis lienis is rare. Patients with splenic peliosis are often asymptomatic. However, life-threatening conditions with spontaneous splenic rupture do occur, often after trivial injury which may not be recalled by the patient. Patients will then present to the accident and emergency department with acute abdominal pain with or without haemorrhagic shock. Prompt diagnosis of intra-abdominal bleeding with bedside ultrasound followed by emergency laparotomy can be life saving.

References