Unusual presentation of more common disease/injury

Spontaneous splenic rupture: an unusual presentation of CML

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A 47-year-old man presented with sudden onset of severe, generalised abdominal pain and collapse. He also had a 4-month history of lethargy and weight loss. On examination he was shocked with a distended tender abdomen. He had haemoglobin of 3.8 g/dl and a white cell count of $280.3 \times 10^9/l$ with predominance of neutrophils. Arterial gases showed mixed metabolic-respiratory acidosis. A CT scan of abdomen showed active extravasation in splenic bed. In view of probable hyperviscosity syndrome, it was decided to attempt an angiographic embolisation. This was successfully carried out but 4 h later, he developed abdominal compartment syndrome and underwent a laparotomy and splenectomy. Subsequently, bone marrow aspirates were taken which showed granulocytic hyperplasia. Cytogenetic studies confirmed the presence of Philadelphia Chromosome and he was started on Imatinib. It is now 5 months since diagnosis and he has achieved complete haematological and cytogenetic response.

BACKGROUND

Spontaneous splenic rupture is a rare clinical entity with grave consequences, if unrecognised and untreated. It mostly occurs in pathologically enlarged spleens but cases of spontaneous rupture in a histologically proven normal spleen have been reported.¹

Spontaneous splenic rupture has been reported as the presenting symptom in patients with lymphoma² and acute myeloid leukaemia.³ We report the first case of a Philadelphia positive chronic myeloid leukaemia which presented with spontaneous splenic rupture as the initial manifestation of the disease.

CASE PRESENTATION

A 47-year-old man presented with sudden onset of severe, generalised abdominal pain and collapse. He had a of

4-month history of lethargy and weight loss and no history of trauma.

On clinical examination, he was hypotensive with a blood pressure of 80/40 mm Hg and a tachycardia of 130 beats/min. His respiratory rate was 32 and he was in type 1 respiratory failure with a pO_2 of 7.1. His abdomen was distended and generally tender. His level of consciousness was rapidly deteriorating.

INVESTIGATIONS

Haematological investigations revealed low haemoglobin at 3.8 g/dl. His white cell count (WCC) was $280.3 \times 10^9/l$ and platelet count was $199 \times 10^9/l$. Differential WCC showed a neutrophil count of 188 and a lymphocyte count of 20.39. There were 10% promyelocytes, 15% myelocytes, 20% metamyelocytes and 20.3% blast cells. Prothrombin time

Figure 1 Axial arterial and portal venous contrast enhanced CT image.

and activated partial thromboplastin time were both prolonged at 19.8 and 53.5 s respectively. Arterial blood gas showed mixed metabolic-respiratory acidosis. A contrast enhanced CT (figure 1) scan of the abdomen revealed a large amount of free fluid in the abdomen, poor visceral enhancement on arterial and venous phase imaging including poor visualisation of the spleen, although there was a subtle heterogeneous attenuation in the left upper quadrant, with evidence of active extravasation in the splenic bed.

TREATMENT

In view of the probable hyperviscosity syndrome compromising respiratory, cerebral and renal perfusion, the presence of a visible bleeding point on CT and in the absence of a visible laceration in the spleen it was decided to attempt angiographic embolisation to control the bleeding while correcting his multiorgan failure. After aggressive

fluid resuscitation, successful radiological embolisation of a splenic artery branch was performed (figure 2).

However, 4 h post procedure, intra-abdominal pressure had increased to 28 mm Hg sufficient to cause abdominal compartment syndrome. He was now more stable following resuscitation and underwent a laparotomy and splenectomy. At operation, a significant splenic laceration was found although this was no longer actively bleeding. The spleen weighed 1810 g and on histological examination, had infiltration of the red pulp and sinuses with cords of granulocytic precursors.

As a result of fluid resuscitation and transfusion of normal donor blood, his WCC had fallen to $100 \times 10^9/l$. A bone marrow aspirate was subsequently obtained which showed granulocytic hyperplasia with a maturation pattern. Cytogenetic studies confirmed the presence of the Philadelphia Chromosome (figure 3) and he was started on Imatinib, a tyrosine kinase inhibitor.

Figure 2 Selective splenic artery angiogram.

Figure 3 Karyotype and FISH image.

OUTCOME AND FOLLOW-UP

It is now 5 months since the diagnosis and he has achieved complete haematological and cytogenetic response to treatment.

DISCUSSION

Spontaneous splenic rupture is a rare but potentially life threatening complication. It is usually a complication of disorders that cause congestion or pathological infiltration of the spleen. Several possible mechanisms have been proposed for splenic rupture in patients with haematological disorders.⁴ Most commonly malignant cells of lymphoproliferative or myeloproliferative origin infiltrate the spleen directly. Their sheer volume exceeds the capacity of the relatively non-distensible splenic capsule, causing splenic rupture and splenic haemorrhage.

The choice of treatment for spontaneous splenic rupture is not only determined by haemodynamic stability, amount of blood products used but also by the underlying pathology. Due to a substantial risk of post-splenectomy infections, there has been a shift towards non-operative management in haemodynamically stable patients.^{5 6}

Splenic artery embolisation may be used as an adjunct to non-surgical management of splenic injury.⁷ The two primary techniques for splenic artery embolisation are proximal and distal splenic artery embolisation. Proximal splenic artery embolisation is equivalent to splenic artery ligation where the splenic artery is selectively catheterised and embolisation coils placed in it, distal to the dorsal pancreatic artery. This reduces the intrasplenic blood pressure and promotes haemostasis while splenic perfusion is maintained through collateral circulation.^{8 9} In distal embolisation, the catheter is placed as close to the site of vascular injury and embolic coils are placed subsequently. This maintains perfusion to the remainder of the spleen but there is a risk of subsequent bleeding as some vascular injuries may go unnoticed due to vasospasm.¹⁰ The choice of proximal and distal embolisation is determined by anatomical feasibility, degree of injury and to a certain extent, operator preference. One problem with proximal embolisation is that it may limit the ability to perform repeat embolisation in the presence of persistent or recurrent bleeding.

If surgery is undertaken, splenorraphy, partial or total splenectomy may be performed depending on the extent of injury. Abdominal compartment syndrome may be another indication for emergency laparotomy in patients with massive intra-abdominal or retroperitoneal haemorrhage. The pathophysiology of abdominal compartment syndrome is complex and can result in multi-organ failure if early decompression is not carried out.

Learning points

- ▶ Spontaneous splenic injury needs a high index of suspicion for diagnosis.
- ▶ Surgeons and emergency room physicians should consider this diagnosis when faced with abdominal pain and hypovolemic shock even if there is no history of trauma.
- ▶ Early diagnosis, aggressive resuscitation with appropriate intervention can lead to successful outcome in these patients.

Competing interests None.

Patient consent Obtained.

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