

Asymptomatic spontaneous splenic rupture – a hair-raising experience in the operating room

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Citation

K Jeevan, S Akshay. *Asymptomatic spontaneous splenic rupture – a hair-raising experience in the operating room*. The Internet Journal of Surgery. 2008 Volume 20 Number 1.

Abstract

Background Spontaneous splenic rupture (SSR) is a rare surgical complication of hematological malignancies like hairy-cell leukemia and asymptomatic SSR is even rarer. SSR is defined as rupture of spleen in absence of any preceding trauma. **Material and methods** We present a case report of a 35-year-old male patient who was diagnosed with hairy-cell leukemia with massive splenomegaly. The patient was referred to us by the hemato-oncologist for splenectomy since the patient had hypersplenism and complained of dragging sensations in the abdomen due to an enormous spleen measuring 18 centimeters below the left costal margin. The patient was hemodynamically stable except for low hemoglobin and was prepared for elective splenectomy. Laparotomy was performed and there were around 3.5 liters of hemoperitoneum, with the splenic capsule densely adherent to the parietal peritoneum. Splenectomy was completed and the patient had an uneventful postoperative recovery. The diagnosis was confirmed by histopathological examination and marker study of the spleen. **Results** Splenic rupture presents as an abdominal emergency. The patient may become exsanguinated in a short interval of time. These are usually patients with splenomegaly presenting in the emergency department with complaints of sudden-onset pain in the left upper quadrant of the abdomen with symptoms of hypotension like dizziness. On examination, the patient is usually in hemodynamic compromise with tachycardia and hypotension. Abdominal examination may reveal signs of peritonitis. A falling hematocrit may be seen on serial hemograms. Radiological investigations (ultrasonography and CT scan of the abdomen) showing hemoperitoneum are helpful in clinching the diagnosis of splenic rupture in patients with splenomegaly presenting with abdominal pain and hypotension. The management entails an adequate resuscitation and an emergency splenectomy. More cases of asymptomatic splenic rupture have to be studied before outlining a definite management protocol, especially the indications of surgery in these patients. **Conclusion** The aim of this case report is to draw attention of the oncologists and surgeons towards this rare asymptomatic complication of hematological malignancies like hairy-cell leukemia which may come as a surprise at the time of laparotomy and envisage a diagnostic work-up for patients with splenomegaly.

BACKGROUND

SSR is a very rare complication^[1] of hairy-cell leukemia. Symptomatic SSR is an abdominal catastrophe with patients presenting with hemodynamic instability^[2]. This requires urgent diagnosis and emergency surgery. The majority of cases of splenic rupture are those of diseased spleen and are called 'pathological rupture of spleen'. 'True spontaneous rupture of spleen', which is very rare, is the rupture of normal spleen. Infections are the most common cause of splenic rupture^[3,4,5,6,7,8].

CASE REPORT

We present a case of a 35-year-old male patient who was referred to us by the hemato-oncologist as a diagnosed case of hairy-cell leukemia. The patient had developed features of hypersplenism and had a persistent dragging sensation in the

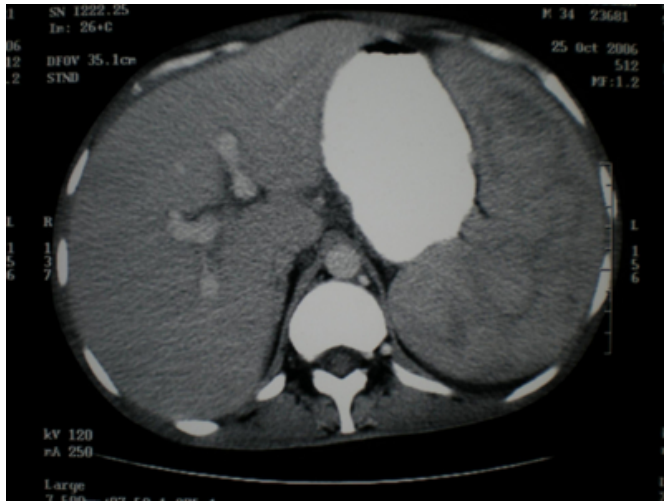
abdomen. These were consequence of a massive splenomegaly.

On general physical examination there was marked pallor and cervical lymphadenopathy. The heart and the lungs were normal. On per abdomen examination there was no tenderness, guarding or rigidity. The spleen was palpable 18cm below the left subcostal margin.

In the preoperative work-up of the patient the hemoglobin was 5.4g%, total leukocyte count was $0.46 \times 10^3/\text{ml}$ and the platelet count was $16 \times 10^3/\text{ml}$. A previously done CT scan of the abdomen and thorax revealed hepatosplenomegaly with generalized lymphadenopathy (figure 1).

Figure 1

Figure 1: CT scan showing hepatosplenomegaly

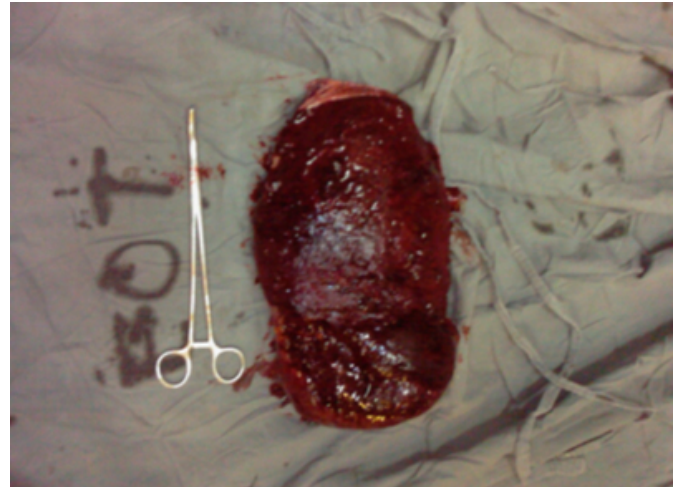


Since the patient was asymptomatic and hemodynamically stable, no further radiological work-up was done. Splenectomy was planned and polyvalent pneumococcal vaccine, H.influenza type B vaccine and meningococcal serogroup C vaccine were administered as a prophylactic measure against overwhelming post-splenectomy infections.

The abdomen was opened through a midline incision. There were around 3.5 liters of hemoperitoneum. The spleen was ruptured at the diaphragmatic surface and the capsule of the spleen was densely adherent to the parietal peritoneum. The splenectomy was successfully undertaken. The oozing capsule was taken down from the peritoneum and hemostasis was ensured. The delivered specimen showed complete loss of capsule at the diaphragmatic surface of the spleen (figure 2 and 3).

Figure 2

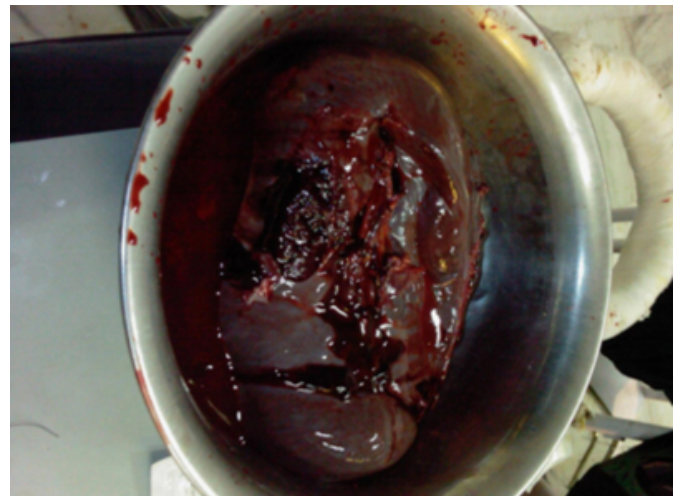
Figure 2: Splenectomy specimen: the diaphragmatic surface of the spleen shows loss of capsule



This was extending into the hilum of the spleen which was also ruptured.

Figure 3

Figure 3: Splenectomy specimen showing the hilum



The patient had an uneventful postoperative hospital stay of 5 days, during which the patient was kept on parental antibiotics. A routine hemogram was repeated at the time of the discharge; hemoglobin was 8.9g% and total leukocyte count was $1.75 \times 10^3/\text{ml}$. The diagnosis of hairy-cell leukemia was confirmed by histopathological examination of the specimen and tumor cells expressing CD 20 and hairy-cell leukemia antigen, DBA44.

SSR as a complication of hematological malignancies is reported in the literature as an abdominal emergency but in the case of our patient it was an asymptomatic rupture of spleen which was accidentally diagnosed at the time of

laparotomy.

DISCUSSION

True spontaneous rupture of spleen was described by Peskin and Orloff^[9] in 1958. They gave the following criteria for its diagnosis: 1. No history of trauma either prior to operation or retrospectively after operation, 2. No evidence of disease that can affect the spleen, 3. No evidence of perisplenic adhesions or scarring of the spleen, suggestive of trauma or previous rupture, 4. The spleen should be normal on gross and histologic examination apart from findings of hemorrhage and rupture. In 1991, Crate and Payne added a fifth criterion: “Full virological studies of acute phase and convalescent sera should show no significant rise in viral antibody titers suggesting recent viral infection of types associated with splenic involvement.”^[10] It is a rare surgical entity with very few cases reported in literature^[10,11]. Rupture of a diseased spleen is known as pathological splenic rupture. Pathological rupture of spleen is rare but is more common than its true variety.

Causes of pathological rupture include (a) infections^[3,4,5,6,7,8] like malaria, infectious mononucleosis, cytomegalovirus, typhoid, HIV, acute or chronic pancreatitis^[12,13,14]; (b) Malignancies, especially hematological malignancies^[15,16,17,18] like hairy-cell leukemia^[19] and other leukemias, rarely hepatocellular carcinoma^[20], angiosarcoma^[21], malignant histiocytosis^[22]; (c) metabolic disorders like Gaucher disease; (d) congenital anomalies like splenic cyst and hamartomas; rare causes include amyloidosis^[23], systemic lupus erythematosus^[24], pregnancy^[25] and rheumatic disease^[26]. A comprehensive study of cases of splenic rupture was done by Gorg et al.^[28] in 2003.

Among the above mentioned etiologies, hemato-oncological malignancies are often incriminated as a cause of pathological rupture of spleen. Giagounidis et al.^[27] studied 136 cases of splenic rupture in hematological malignancies; 34% were cases of non-Hodgkin lymphoma, acute myelogenous leukemia (34%), chronic myelogenous leukemia (18%) and lymphoblastic acute leukemia. It was found to be 3 times more frequent in men than women.

Pathogenesis^[27] of rupture includes congestion of the parenchyma by blast cells and intrasplenic hemorrhage caused by coagulation abnormalities and splenic infarction. Splenic ruptures usually occur in enlarged spleens, the patient we report had a massive splenomegaly.

Splenic rupture, traumatic or spontaneous (true or

pathological), commonly presents as an abdominal emergency. Patients usually present in the accident and emergency department with sudden onset pain in the left upper abdomen. Examination reveals hypotension and tachycardia and abdominal tenderness and guarding. The Kehr sign may or may not be present. Hemogram usually reveals falling hematocrit. CT scan may reveal hemoperitoneum and splenic parenchymal infiltration^[32].

Treatment includes intensive resuscitation and prompt diagnosis, both clinical and by using imaging techniques (ultrasonography or CT scan), followed by emergency splenectomy. Immunization to prevent opportunistic post-splenectomy infection should be undertaken^[30].

CONCLUSION

The cases of SSR reported in the literature presented with abdominal emergency. Our patient had an asymptomatic rupture which throws light over a different aspect of complication of hematological malignancies. The aim of this case report is to draw attention of the oncologists and surgeons towards this rare asymptomatic complication which may come as a surprise at the time of laparotomy. Whether routine serial radiological investigations in these patients will influence the diagnosis and management of this silent event requires more cases to be studied.

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