Case Report–Spontaneous Splenic Rupture–A Rare Complication of Haematological Malignancies

Ritu Galhotra · Samarjit Kaur · Kamini Gupta · Vishal Kalia

Received: 8 June 2010 / Accepted: 21 March 2011 / Published online: 12 April 2011
© Indian Association of Surgical Oncology 2011

Introduction

Spontaneous splenic rupture is an abdominal catastrophe with patient presenting with haemodynamic instability. This requires urgent diagnosis and emergency surgery. The majority of the cases of splenic rupture are those of diseased spleen and are called pathological rupture of spleen [1]. True spontaneous rupture which is very rare, is the rupture of normal spleen. Haemato-oncological malignancies are often incriminated as a cause of pathological rupture of spleen [1]. Splenomegaly is a very common presentation of chronic myeloid leukaemia. However the progression of splenomegaly in chronic myeloid leukaemia is usually slow and insidious that many patients may not be aware of it until spleen has become huge in size [2].

Pathogenesis 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 [1].

Here we present a case of 40 year male who was a known case of chronic myeloid leukaemia who presented in emergency with left upper quadrant pain and abdominal distention. Patient was haemodynamically unstable. After doing ultrasound and computed tomography diagnosis of spontaneous splenic rupture was made. Patient was operated and splenectomy was done. Patient was stable post operatively.

Case Report

We present a case of 40 year old male who was diagnosed as chronic myeloid leukaemia 6 months back on basis of peripheral blood film and bone marrow. His spleen was enlarged in size. Patient did not receive any treatment for this.

He presented in the emergency with left upper quadrant pain and progressively increasing abdominal distention. On examination patient was mildly febrile and pale. He was having cold and clumsy skin. His pulse was 136/min and BP was 94/60. Heart and lungs were normal. No past history of trauma was there.

Perabdominal Examination

Abdomen was distended with left upper quadrant tenderness guarding and rigidity were present. Spleen was enlarged in size. Investigations showed low Hb of 7.4 gm with increased TLC of 1,24,000 with neutrophils 84% and PCV of 21.6%. After the examination provisional diagnosis of acute pancreatitis was made. Ultrasonography and computed tomography was done.

Ultrasound showed marked splenomegaly and large perisplenic and intra splenic haematoma (Fig. 1)

Computed Tomography

Computed tomography was performed on a multi detector dual source CT Scanner by Siemens (128 slice) using standard protocols.

Computed tomography revealed a large intraperitoneal hematoma in the left upper quadrant extending from left subphrenic space till lumbar region. Spleen was not separately visualized. Splenic artery was not opacified

R. Galhotra · S. Kaur · K. Gupta · V. Kalia
Department of Radiodiagnosis,
Dayanand Medical College & Hospital,
Ludhiana 141001, India

R. Galhotra (✉)
96 LAL BAGH, Near RAjguru Nagar, Post Office Threake,
Ludhiana, Punjab, India
e-mail: ritugalhotra96@gmail.com

Fig. 1 Large intraperitoneal hematoma is seen in the left upper quadrant extending from left subphrenic space till lumbar region. Spleen was not separately visualized. Spleen artery was not opacified throughout its whole extent. Significant ascites is present

throughout its whole extent. Significant ascites is present (Figs. 2, 3, 4).

On the basis of computed tomography findings, diagnosis of spontaneous splenic rupture was made. Patient was immediately taken up for surgery. Splenectomy with complete haemostasis was done.

Intra-Operative Findings

Enlarged spleen with large intra and perisplenic haematoma was seen. Significant ascites was there.

Fig. 2 Untrasound showing enlarged spleen with intra and perisplenic haematoma significant ascites is present

Histopathological Examination

Histopathology report showed massive splenomegaly with multiple intrasplenic infarcts and intra and perisplenic haematoma.

Post Operative Findings

The patient was stable post operatively. His repeat haemoglobin was 9.4 gm with TLC of 64,000. He was discharged after 10 days and called for follow up for the treatment of chronic myeloid leukaemia (chemotherapy).

Discussion

True spontaneous rupture of spleen was described by Peskin and Orloff 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 histological examination apart from findings of haemorrhage and rupture. In 1991, Crate and Payne added a fifth criteria—full virological studies of acute phase and convalescent sera should show no significant rise in viral antibody titres suggesting recent viral infection of types associated with splenic involvement [1].

It is a rare surgical entity with very few cases reported in literature [6, 7]. Rupture of a diseased spleen is known as pathological splenic rupture. Pathological

Fig. 3 CT sagittal image showing large intraperitoneal hematoma in the left upper quadrant extending from left subphrenic space till lumbar region. Spleen was not separately visualized. Significant ascites is present

rupture of spleen is rare but is more common than its true variety [1].

Spontaneous splenic rupture is a life threatening event which can be an initial presentation of an unpredictable complication during disease course in chronic myeloid leukaemia patient. Total 11 chronic myeloid leukaemia

Fig. 4 CT axial image showing large intraperitoneal hematoma in the left upper quadrant extending from left subphrenic space till lumbar region. Spleen was not separately visualized

cases with spontaneous splenic rupture have been reported in English literature. Of them 10 were male and the median age 49 years. Spontaneous splenic rupture as the initial presentation occurred in four of them. Seven of them could only be diagnosed at post mortem examination [2].

Causes of pathological rupture include (a) infections like malaria, infection mononucleosis, cytomegalovirus, typhoid, HIV or chronic pancreatitis, (b) Malignancies especially haematological malignancies like hairy cell leukaemia and other leukaemia, rarely hepatocellular carcinoma, angiosarcoma, malignant histiocytosis, (c) metabolic disorder like Gaucher disease (d) congenital anomalies like splenic cyst and hamartomas rare causes include amyloidosis, systemic lupus erythematosus, pregnancy and rheumatoid disease [1]. A comprehensive study of cases of splenic rupture done by Gorg et al. in 2003 [8].

Among the above mentioned etiological haematological malignancies are often incriminated as a cause of pathological rupture of spleen [1].

One or more of the following mechanism may be incriminated as the cause of spontaneous splenic rupture in Leukaemia. (i) The mechanical effect of leukaemic infiltration in the spleen especially if the capsule is involved. (ii) Splenic infarction with consequent subcapsular hemorrhage and subsequent rupture of splenic capsule. (iii) Defect in blood coagulation [2].

In our Patient Probably the Leukemia Infiltration and Splenic Infarction both were the Cause of Splenic Rupture

In a review of literature, Giagounidis et al. been able to identify 136 cases of pathological rupture since 1861. 34% have occurred in acute leukaemia, 34% in non Hodgkin's lymphoma and 18% in chronic myelogenous leukaemia. They find male: female ratio of 3:1 with considerable differences for the specific diseases encountered [5].

Pathological rupture of spleen has happened almost exclusively in adults and ruptured spleen are generally moderate to severely enlarged [5].

Most authors continue to advocate splenectomy as the treatment of choice in spontaneous splenic rupture. They argue that there is no strong criteria that non operative management is safer short term or beneficial in long term [4].

Giagounidis et al. also reported that the mortality rate of spontaneous splenic rupture in haematological malignancies was 37% in operated case and 93% in non operated cases [2].

Splenectomy exposes the patient to the dangers of diminished bone marrow reserve and post operative complications [3].

Post splenectomy sepsis is a potential lethal complication of splenectomy with a mortality of 50 to 80% [4].

So immunization to prevent opportunistic post splenectomy infection should be undertaken [1].

Conclusion

To conclude spontaneous splenic rupture is a rare complication of haematological malignancies. The diagnosis should be considered in a patient who presents with left upper quadrant pain, hypotension and tachycardia. Ultrasound and contrast enhanced computed tomography are the diagnostic modalities of choice. Splenectomy is the treatment of choice with immunization to prevent opportunistic post splenectomy infection.

References

1. Jeevan K, Akshay S (2009) Asymptomatic spontaneous splenic rupture—A hair rising experience in the operating room. *The Internet Journal of Surgery* 20:1
2. Ho Ching-Liang, Yeu-Chinchen, Chao Tsue-Yi (1996) Spontaneous splenic rupture in chronic myeloid leukaemia: A case report and review of literature. *J Chinese Oncol Soc* 12(1):1–6
3. Mentzer SJ, Starnes HF, Canellos G, Osteen RT (1987) Splenic enlargement and hyperfunction as indication for splenectomy in chronic luvaemia. *Ann Surg* 205:13–17
4. Asgaria MM, Begos DG (1997) Spontaneous splenic rupture in infections mononucleosis: A review. *Yale Journal of Biology and Medicine* 70:175–182
5. Giagounidis AAN, Burk M, Meckeustoch G, Koch AJ (1996) Pathological rupture of spleen in hematologic malignancies: two additional cases. *Ann Hematol* 73:297–302
6. Crate ID, Payne MJ (1991) Is the diagnosis of spontaneous rupture of a normal spleen valid? *J R Army Med Corps* 137:50–51
7. Tu AS, Tran MHT, Larsen CR (1997) Spontaneous splenic rupture of five cases and a review of the literature. *Emerg Radiol* 4:415–418(S)
8. Gorg C, Colle K, Gorg K, Zugmainer G (2003) Spontaneous rupture of the spleen. Ultrasound patterns diagnosis and follow up. *Brit J Radiol* 76:704–711(S)