

**CASE REPORT**

Spontaneous Splenic Rupture - A Rare Complication of Haematological Malignancies

Ritu Galhotra, Paramdeep Singh, Samarjit Kaur, Kamini Gupta

Abstract

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 along with immunization to prevent opportunistic post splenectomy infection.

Key Words

Spontaneous Splenic Rupture, Haematological Malignancies

Introduction

Spontaneous splenic rupture is an abdominal catastrophe with patient presenting with hemodynamic 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 of spleen is very rare. 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 hemodynamically 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 six 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.

Abdomen was distended. Left upper quadrant tenderness, guarding and rigidly 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 hematoma. Computed tomography was performed on a multi-detector 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 extending till lumbar region. Spleen was not separately visualized. Splenic artery was not opacified

From the Department of Radiodiagnosis, Dayanand Medical College & Hospital, Ludhiana - 141001

Correspondence to :Dr. Paramdeep Singh, Department of Radiodiagnosis, Dayanand Medical College & Hospital, Ludhiana - 141001



Fig 1-3. Large Intraperitoneal Hematoma is seen in the Left Upper Quadrant Extending from left Subphrenic Space till Lumbar Region. Spleen is not Separately Visualized. Spleen Artery is not Opacified Throughout its Whole extent. Significant ascites is present.

throughout its whole extent. Significant ascites is present (Fig. 1-3). 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. Histopathology report showed massive splenomegaly with multiple intrasplenic infarcts and intra and perisplenic haematoma.

The patient was stable post operatively. His repeat hemoglobin 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 than 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 titers suggesting recent viral infection which can be associated with splenic involvement (1). It is a rare surgical entity

with very few cases reported in literature (3, 4). 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 (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 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, infectious mononucleosis, cytomegalovirus, typhoid, HIV or chronic pancreatitis, (b) Malignancies especially haematological malignancies like hairy cell leukaemia and other leukaemias, rarely hepatocellular carcinoma, angiosarcoma, malignant histiocytosis, (c) metabolic disorders like Gaucher disease (d) congenital anomalies like splenic cyst and hamartomas. Rare causes include amyloidosis, systemic lupus erythematosos, pregnancy and rheumatoid disease(1). A comprehensive study of cases of splenic rupture done by Gorg *et al* in 2003(5).

Among the above mentioned etiological haemato-oncological 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 leukemic infiltration and splenic infarction both were the cause of splenic rupture.

In a review of literature, Giagounidis *et al* identified 136 cases of pathological rupture since 1861. 34% occurred in acute leukaemia, 34% in non Hodgkin's lymphoma and 18% in chronic myelogenous leukaemia. They found male: female ratio of 3:1 with considerable differences for the specific diseases encountered (6).

Pathological rupture of spleen has happened almost exclusively in adults and ruptured spleen are generally moderate to severely enlarged (6). Most authors continue to advocate splenectomy as the treatment of choice in spontaneous splenic rupture. They argue that there is no strong criteria that now operative management is safer short term or beneficial in long term (7).

Giagounidis *et al.* also reported that the mortality rate of spontaneous splenic rupture in hematological malignancies was 37% in operated case and 93% in non operated cases (6). Splenectomy exposes the patient to the dangers of diminished bone marrow reserve and post operative complications (8). Post splenectomy sepsis is a potential lethal complication of splenectomy with a mortality of 50 to 80% (9). So immunization to prevent opportunistic post splenectomy infection should be undertaken (10).

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 along with immunization to prevent opportunistic post splenectomy infection.

References

1. Jeevan K and Akshay S. Asymptomatic spontaneous splenic rupture - A hair rising experience in the operating room. *The Internet Journal of Surgery*, 2009; 20: 1.
2. Ching-Liang Ho, Yeu-Chinchen, Tsue-Yi Chao. Spontaneous splenic rupture in chronic myeloid leukaemia: A case report and review of literature. *J Chinese Oncol Soc* 1996; 12(1): 1-6.
3. Crate ID, Payne MJ. Is the diagnosis of spontaneous rupture of a normal spleen valid? *Journal of the Royal Army Medical Corps* 1991; 137: 50-51.
4. Tu AS, Tran MHT, Larsen CR. Spontaneous splenic rupture of five cases and a review of the literature. *Emerg Radiol* 1997; 4: 415-18(S).
5. Gorg C, Colle K, Gorg K, Zugmainer G. Spontaneous rupture of the spleen. Ultrasound patterns diagnosis and follow up. *Brit J Radiol* 2003; 76: 704-11(S).
6. Giagounidis AAN, Burk M, Meckeustoch G and Koch AJ. Pathological rupture of spleen in hematologic malignancies: two additional cases. *Ann Hematol* 1996; 73: 297-302.
7. Maryan M Asgaria Dennis G Begos. Spontaneous splenic rupture in infections mononucleosis: A review. *Yale Journal of Biology and Medicine* 1997; 70: 175-82.
8. Mentzer SJ, Starnes HF, Canellos G, Osteen RT. Splenic enlargement and hyperfunction as indication for splenectomy in chronic leukaemia. *Ann Surg* 1987; 205: 13-17.
9. Edgard Badine, Elie Richa, Dany El-Sayal, Terenig Terjanian. A case report of splenic rupture due to marginal zone lymphoma. *Ann Nalel* 2007; 86: 845-46.
10. Au H, Mac YK, Luk SH, Cheung CC. Chronic myeloid leukaemia presenting as traumatic peritoneum and duodenal injury. *Hong Kong J Emerg Med* 2007; 7: 236-37.