

Case Report-III

Spontaneous Splenic Rupture- A Rare Initial Manifestation of Acute Lymphoblastic Leukemia

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ABSTRACT

Pathologic splenic rupture is a rare and life threatening complication of acute leukemia. It is even more uncommon as initial manifestation and only a few cases have been reported in the literature. One such case is being reported to emphasize the importance of this potentially fatal complication.

INTRODUCTION

Pathologic rupture of spleen is a serious complication appearing with relative low frequency in infections or parasitic disease, but is rare in haematological malignancies.¹ It was first reported in 1861 by Rokitansky in cases of leukemia.² It has an incidence of 0.72% but acute leukemia manifesting primarily as splenic rupture is extremely rare.³ Here, we report one such case.

CASE

A 25-year old female presented in a state of shock. She had history of severe pain abdomen. There was no history of trauma or history of any systemic symptoms including fever, bleeding diathesis or bone pains. On examination, she was drowsy and pale. Her

pulse was very feeble and blood pressure was unrecordable. She had generalized lymphadenopathy. There was distension of abdomen. She was diagnosed as acute abdomen and immediate exploratory laparotomy was performed. No haematological or radiological investigations could be done before emergency surgery. During surgery she was found to have gross hepatosplenomegaly with rupture of spleen. She was managed with splenectomy, IV fluids and blood transfusions.

Later her investigations revealed Hb-7.9 gm/dl, WBC-90,000 / cu mm, with 67% blasts. The platelet count 45,000 / cu mm and her coagulation parameters were normal. Bone marrow aspiration showed a hypercellular marrow with predominant blast cells. The blast cells were small with high nuclear cytoplasmic ratio, scant cytoplasm, condensed chromatin and indistinct nucleolus. The peroxidase staining was negative while PAS staining showed fine block positivity in occasional cells. Acute lymphoblastic leukemia, FAB type L1 was diagnosed on the basis of these features. Her kidney and liver function tests as well as blood sugar levels were within normal limits.

DISCUSSION

Spontaneous rupture of the spleen is a serious complication appearing frequently in infections and parasitic diseases like infectious mononucleosis and malaria but is rare in

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haematological malignancies. It has been reported that between 0.72% and 35% of spontaneous splenic rupture can be associated with leukemia.^{3,4}

The first case of spontaneous rupture of the spleen in a patient with leukemia was reported in 1861 by Rokitansky.⁵ One hundred and thirty-six cases were reviewed by Hernandez et al.¹ In this review, 3% of the cases were of acute leukemia, 34% had non-Hodgkin's lymphoma and 18% had chronic myeloid leukemia. There is a male to female ratio of 3:1. Other pathogenetic factors are splenic infiltration, splenic infarcts, coagulation disorders leading to intrasplenic and subcapsular bleeds and subsequently capsular rupture.⁸ Male patients, aged above 20 years, gross splenomegaly and those who have recently received cytoreductive chemotherapy seem to be at a higher risk of rupture.² An acute increase in portal venous pressure from a valsalva like manoeuvre or a sudden compression of pathological spleen from diaphragmatic or abdominal wall compression are possible reasons for splenic rupture.⁹ More recently other cases of spontaneous splenic rupture have also been reported.¹²⁻¹⁹ Advances in imaging techniques may have lead to greater identification of splenic rupture in recent years.¹⁹

Another interesting observation is that although ALL is common in age group less than 18 years, splenic rupture is reported mostly in patients aged 21-59 years.^{6,9} This is attributed to age related anatomical defects in the spleen and the fact that a difference in cell types associated with the poorer prognosis in adults may predispose them to splenic rupture.⁷ There are some reports of spontaneous splenic rupture in children as well.^{13,19}

Most of the clinical symptoms of splenic rupture are the result of intra-abdominal

haemorrhage: hypotension (54%), tachycardia (54%), abdominal tenderness (68%), nausea and vomiting (25%). Interesting but rare is an Kehr's sign (19%) i.e. left hypochondral pain radiating to left shoulder and coughing, both of which are probably the result of diaphragmatic irritation.² Abdominal ultrasound is a non-invasive diagnostic aid and can be accompanied by needle aspiration under ultrasound guidance.¹⁰ CAT scan is equally effective but should not be performed in unstable patients.¹¹ Splenectomy is generally considered the optimal management for rupture in haematological malignancies because of underlying problem.⁸

To conclude, any patient with acute leukemia, complaining of left upper abdominal pain, which may radiate to left shoulder (Kehr's sign), should be observed closely and further diagnostic investigations (abdominal ultrasound or CAT scan) should be initiated to rule out rupture of the spleen.

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