Case Report
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Non-Traumatic Spontaneous Splenic Rupture: A Rare Initial Presentation of Chronic Myeloid Leukaemia

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Abstract
Chronic myeloid leukaemia (CML) is a myeloproliferative disorder, usually asymptomatic and diagnosed incidentally with an elevated WBC counts on a routine laboratory tests. Its presentation in emergency as hem peritoneum is extremely rare. Here we report a rare case of CML presented to us as spontaneous splenic rupture. A 20-year female presented with generalised abdominal pain for past 2 days with no history of trauma. On examination, patient was in shock and abdomen was distended with generalised guarding and rigidity. Ultrasound abdomen revealed enlarged liver and spleen with large fluid collection in the pelvis suspecting haemorrhage.

After adequate resuscitation, patient was taken for exploratory laparotomy. Intraoperatively there was a massively enlarged spleen with bleeding actively from its superior pole. Splenectomy was done. Post operatively, patient was diagnosed as chronic myeloid leukaemia by peripheral smear and bone marrow study in view constantly elevated white blood counts. Spontaneous splenic rupture is a rare but potentially life-threatening entity. It usually occurs in pathologically enlarged spleen where rupture occurs because of infiltration of spleen by malignant cells. High index of suspicion is required to clinch the diagnosis in non-traumatic patients.

Keywords: Spontaneous Splenic Rupture; Chronic Myeloid Leukaemia; Non-Traumatic Hemoperitoneum

Abbreviations: CML: Chronic Myeloid Leukaemia; SSR: Spontaneous Splenic Rupture

Introduction
Chronic myeloid leukaemia (CML) is a myeloproliferative disorder which results from genetic change in the pluripotential hematopoietic stem cells [1]. It is usually asymptomatic and diagnosed incidentally with an elevated WBC counts on a routine laboratory test. Though splenomegaly is a common manifestation in CML, spontaneous splenic rupture is a rare occurrence [2]. Its initial presentation in emergency as hemoperitoneum is extremely rare. Here we report a rare case of CML presented to us as spontaneous splenic rupture (SSR).

Case Report
A 20-year female presented to emergency department with complaints of generalised abdominal pain for past 2 days. There was no history of trauma and other significant history. Her menstrual history was normal. On examination, patient was in shock with tachycardia and feeble pulse. Abdomen was distended with generalised guarding and rigidity. Patient was initially resuscitated with i.v. fluids. After adequate resuscitation, ultrasound abdomen was done which revealed enlarged liver and spleen with large fluid collection with internal echoes in the pelvis suspecting haemorrhage. Abdominal tapping also confirmed haemorrhagic fluid. After adequate resuscitation, patient was taken for exploratory laparotomy in view of clinical and radiological features suggestive of hemoperitoneum.

Abdomen was opened by midline laparotomy incision. Intraoperatively, there was a gross hemoperitoneum of around 3 litres with massively enlarged spleen with bleeding actively from its superior pole. Splenectomy was done with through peritoneal lavage. On gross examination, the spleen measures 22×16×5 cm weighing 2 kg. Post operatively patient received blood products transfusion in view of low hemogram levels. Post-operative period was uneventful except constantly elevated WBC counts 1,10,000 per cu.mm. Peripheral smear was done that showed raised total counts with predominant population of...
myelocytes, metamyelocytes and mature neutrophils suspecting
Chronic Myeloid Leukaemia (CML).

**Figure 1:** Splenectomy specimen. Enlarged spleen of 22×16×5 cm weighing 2kg.

**Figure 2:** Histopathological specimen of spleen showed infiltration of red pulp and white pulp by hematopoietic precursor of myeloid lineage with various stages of maturation.

On microscopic examination of section from spleen showed infiltration of red pulp and Figure 1,2 white pulp by hematopoietic precursor of myeloid lineage with various stages of maturation. Bone marrow study also revealed hypercellular bone marrow chiefly comprising myeloid cells in all stages of maturation, consistent with chronic phase of CML. Polymerase chain reaction done for BCR-ABL gene (Philadelphia chromosome) was found to positive for CML. The diagnosis of chronic myeloid leukaemia was made and the patient was put on imatinib (tyrosine kinase inhibitor). Patient laboratory parameters were improved after initiation of imatinib. It is now 8 months since the diagnosis; patient is doing well and does not shift to acceleration or blast crisis phase and responding well to imatinib therapy.

### Discussion

Non-traumatic hemoperitoneum is the presence of blood in the peritoneal cavity which is not associated with trauma, but occurring due to idiopathic cause or due to any spontaneous rupture of known or unknown pathology [3]. Trauma is the most common cause of splenic rupture, while non-traumatic Spontaneous splenic rupture is a rare but potentially life-threatening entity. It usually occurs in pathologically enlarged spleen, but can also occur in histologically proven normal spleen. Spontaneous splenic rupture is usually due to neoplastic, infectious, haematological, inflammatory, metabolic, iatrogenic or idiopathic. Neoplasia and infectious causes accounts for more than half of the cases [4].

The diagnosis of atraumatic idiopathic splenic rupture can be made with the Orloff and Peskin criteria, when the following four criteria are met [4]:

1. No antecedent trauma history.
2. No evidence of disease of organs other than the spleen that can cause rupture.
3. No peri splenic adhesions or scarring consistent with past trauma.
4. Normal spleen on gross and histological examination.

Spontaneous splenic rupture of neoplastic origin is usually due to leukaemia and lymphoma. The incidence of splenic rupture in leukaemia is about 0.7 % [1]. Here we reported a rare case of spontaneous splenic rupture as the first presentation in CML. Splenomegaly is a very common presentation in CML and has an insidious slow progression. However spontaneous rupture of spleen is a very rare presentation of CML. The mechanical effect of splenic distension secondary to infiltration of leukemic cells attributes to the major cause of splenic rupture [5]. The sheer volume of the malignant cells exceeds the capacity of relatively non-distensible splenic capsule causing splenic rupture and haemorrhage. Other possible explanations for splenic rupture are splenic infarct by leukemic cells and defects in the blood coagulation [5].

Management for spontaneous splenic rupture is determined by hemodynamic stability and underlying pathology. Due to risk of post splenic infections, there has been as shift of treatment modality towards non-operative management in a clinically stable patient [6]. Splenic artery embolization can also be employed for clinically stable patients. If the patient is clinically unstable and requires more number of blood transfusions to maintain vitals, they are definitive candidates for surgery, keeping in mind that spontaneous splenic rupture is a dreaded but easily treatable condition if diagnosed earlier. If surgery is undertaken, spleen oraphy, partial or total splenectomy can be performed depending upon the extent of splenic injury. Although there are no definitive guidelines for treatment of SSR, few
studies suggest that total splenectomy is recommended for SSR associated with neoplasm and conservative management can be attempted in SSR associated with infectious diseases [7,8].

**Conclusion**

Spontaneous splenic rupture (SSR) is a very rare, lethal but potentially treatable condition. The diagnosis of SSR must be considered in all patients with haematological malignancies who experience sudden abdominal pain, hypotension and shock, even though they deny a history of trauma. High index of clinical suspicion is required to diagnose this condition early in high risk patients. Even though neoplasia and infections predominate the list, others possible causes should also be evaluated in cases of clinical dilemma.

**References**