

Splenic Rupture in Chronic Myeloid Leukemia: A Rare Hematological Emergency

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Abstract: *Chronic myeloid leukemia (CML) is a clonal myeloproliferative neoplasm commonly presenting with splenomegaly, constitutional symptoms, and laboratory evidence of leukocytosis. Spontaneous splenic rupture is a rare but potentially fatal initial presentation. We report the case of a 40-year-old male with no prior comorbidities who presented with acute abdominal pain, distention, and vomiting. Clinical examination revealed massive splenomegaly, hypotension, pallor, and absent bowel sounds. Laboratory investigations demonstrated marked leukocytosis (total leukocyte count: 251,300/ μ L), anemia, and peripheral smear findings consistent with a left shift. Contrast-enhanced CT abdomen revealed massive splenomegaly with parenchymal disruption, large perisplenic hematoma, and hemoperitoneum. Despite fluid resuscitation and transfusion, he remained hemodynamically unstable and underwent emergency splenectomy. Histopathology confirmed leukemic infiltration of the spleen, and bone marrow studies with PCR identified BCR-ABL positivity, confirming CML. Postoperatively, the patient received appropriate vaccinations and was commenced on imatinib, with good clinical response on follow-up. This case underscores the need to consider atraumatic splenic rupture in patients with hematological malignancies presenting with acute abdomen, even without a history of trauma. Early diagnosis, prompt surgical intervention, and targeted therapy for the underlying hematologic disorder are critical to improving outcomes.*

Keywords: Chronic myeloid leukemia, spontaneous splenic rupture, splenectomy, imatinib, hematological malignancy.

1. Introduction

Chronic myeloid leukemia (CML) is a myeloproliferative neoplasm commonly presenting with organomegaly and constitutional symptoms. Spontaneous (atraumatic) splenic rupture (ASR) is a rare yet life-threatening initial presentation. This case underscores the importance of early recognition and comprehensive management strategies.

2. Case Presentation

A 40-year-old male with no known comorbidities presented to the emergency department with abdominal pain and distention for one day, associated with four episodes of bilious vomiting. On examination, he was conscious and oriented. Vital signs revealed a pulse rate of 124/min, blood pressure of 90/60 mmHg, and respiratory rate of 38/min. He was clinically pale, with no icterus or lymphadenopathy.

Abdominal examination showed a distended abdomen with tenderness in the left hypochondrium. The spleen was palpable 10 cm below the left costal margin. There was no guarding or rigidity, and bowel sounds were absent. Decreased bilateral basal breath sounds were noted, while other systemic examinations were unremarkable.

Laboratory investigations revealed hemoglobin of 6.4 g/dL, total leukocyte count of 2,51,300/ μ L, and platelet count of 1,60,000/ μ L. Peripheral smear showed a left shift of granulocytes with the presence of promyelocytes, myelocytes, and metamyelocytes.

Differential leukocyte count was N56, L12, M0, E2, B10, PM3, My18, MMy9. Renal function tests were deranged (blood urea 53 mg/dL, creatinine 1.7 mg/dL). Coagulation

profile: INR 1.3, APTT 26.4 seconds.

Plain abdominal radiograph demonstrated medial displacement of the gastric shadow with rightward displacement of bowel loops. Contrast-enhanced CT (CECT) of the abdomen revealed splenomegaly measuring ~20.2 cm (craniocaudal), with parenchymal disruption along the anterior aspect. A large perisplenic hematoma measuring ~21 × 10.0 × 21.4 cm with moderate hemoperitoneum was noted. Bilateral pleural effusion with minimal atelectasis was present. The splenic artery arose directly from the abdominal aorta, with no evidence of aneurysm or contrast extravasation.

The patient was resuscitated with intravenous fluids and transfused with 3 units of packed red blood cells (PRBC) and 3 units of fresh frozen plasma (FFP). Despite initial resuscitation, he showed minimal clinical improvement, warranting surgical intervention. He was taken up for emergency splenectomy through a midline laparotomy, which revealed approximately 2 liters of clots distributed in all quadrants, predominantly in the perisplenic region.

Intraoperatively, massive splenomegaly with rupture at the superior pole and a large subcapsular hematoma were noted. The spleen measured 30 × 20 × 6.5 cm.

Postoperative recovery was uneventful, and the patient was discharged in stable condition on the 7th postoperative day with prophylactic antibiotics. He received pneumococcal vaccination on postoperative day (POD) 3, followed by meningococcal and *Haemophilus influenzae* type B vaccines on POD 14 after discharge.

Histopathological examination of the specimen revealed

infiltration by large atypical polymorphic cells. Bone marrow aspiration showed hypercellular marrow with a predominance of myelocytes. PCR for *BCR-ABL* demonstrated a significant increase in transcript copies, confirming the diagnosis of chronic myeloid leukemia

(CML). The patient was started on tablet Imatinib 400 mg once daily from postoperative day 6. He has been responding well to therapy and is currently on regular hematology-oncology follow-up.

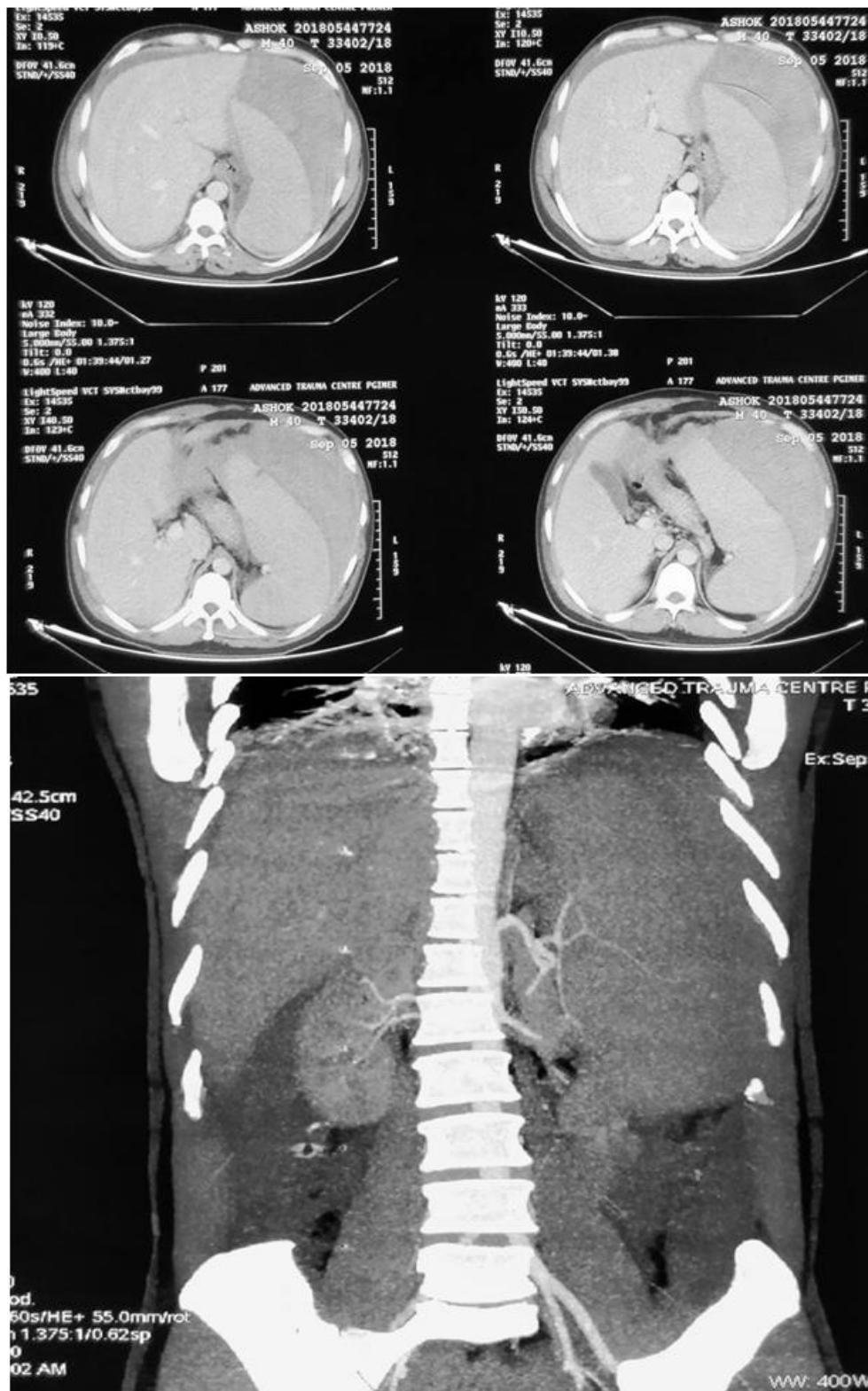
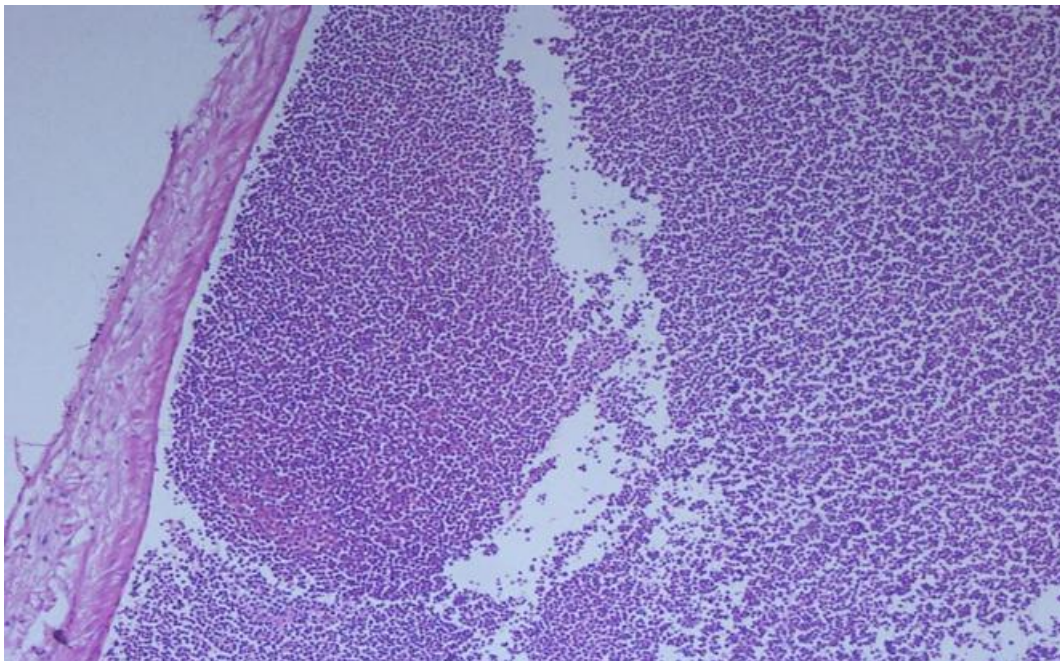
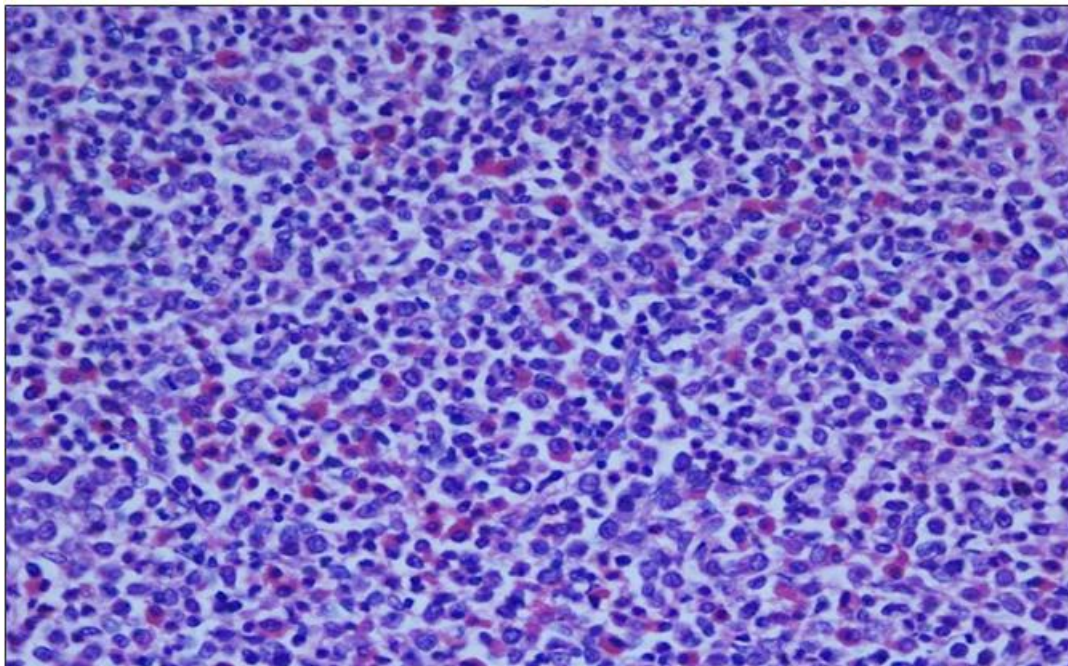


Figure 1 (a) & (b): CT scan report showing massive splenomegaly and haemorrhage



Intraoperative picture showing rupture of spleen in the superior pole





Histopathology of splenectomy specimen showing: replacement of splenic parenchyma by atypical polymorphic cells. Focal capsular breach and intra parenchymal hematoma is noted.

3. Discussion

845 patients. Among these, 152 cases (16.4%) were attributed to neoplastic hematological disorders. Of these, non-Hodgkin's lymphoma was the most frequent cause (55/152 cases), followed by myeloproliferative disorders including CML, which accounted for 24 cases¹.

Clinically, patients with atraumatic splenic rupture typically present with abdominal pain (most often in the left upper quadrant), abdominal distension, nausea, vomiting, light-headedness, or near-syncope. Kehr's sign—referred pain to the left shoulder—is reported in 80–90% of cases³. On physical examination, findings may include hypotension, pallor, left upper quadrant tenderness (with or without guarding), and occasionally a palpable tender mass in the left upper quadrant, described as Balance's sign.

Imaging plays a critical role in diagnosis. Plain abdominal radiographs may demonstrate medial displacement of the gastric bubble or elevation of the left hemidiaphragm depending on splenic size and the degree of hemoperitoneum. Ultrasound can estimate the volume of hemoperitoneum and provide a rough assessment of splenic size and site of rupture.

Contrast-enhanced CT (CECT) of the abdomen is considered

the gold standard investigation, with sensitivity and specificity exceeding 95%⁴. It allows accurate assessment of splenic size, grade and site of injury, volume of hemoperitoneum, and any associated intra-abdominal pathology. The grading of spontaneous splenic rupture follows the American Association for the Surgery of Trauma (AAST) classification used for traumatic splenic injuries.

Management depends primarily on the patient's hemodynamic status, the extent of splenic injury, amount of hemoperitoneum, and blood transfusion requirements. Surgical intervention is required in the majority of cases, with approximately 85% undergoing emergency splenectomy; organ-preserving approaches such as splenorrhaphy or partial splenectomy are rarely performed^{1,5}. Conservative management has been attempted successfully in nearly 15% of cases in select centers^{1,6,7}, and splenic artery embolization has also been reported as a therapeutic option^{1,8}. Following stabilization, it is essential to evaluate and treat the underlying pathology. Post-splenectomy vaccination and appropriate antibiotic prophylaxis remain crucial to prevent overwhelming post-splenectomy infections.

A search of the English-language literature since 2011 reveals at least **five additional well- documented cases** of CML presenting with spontaneous splenic rupture. These include:

Table 1

Year	Author(s)	Age/Sex	Presentation & Findings	Management
2011	Jafferbhoy et al. [9]	47M	Shock, active extravasation on CT	Embolization → Splenectomy
2019	García-Monje et al. [10]	26 M	Known CML, infarct & rupture	Splenectomy
2021	Vemulakonda et al [11]	48 M	Grade III laceration, LUQ pain	Emergency splenectomy
2023	Alharthy et al. [12]	68F	CML + COVID-19, rupture	Splenectomy + TKI
2025	Hassan et al. [13]	42 M	LUQ pain, rupture diagnosed on CT	Conservative management

LUQ pain, rupture diagnosed on CT
Conservative management

When combined with the earlier cases in Renzulli's review, the cumulative documented cases of CML-associated spontaneous splenic rupture in the literature amount to **approximately 30**.

Our case adds to this rare pool and is notable for the large splenic hematoma (~21 × 10 × 21.4 cm), massive splenomegaly, and presentation in the absence of prior CML diagnosis.

4. Conclusion

ASR should be considered in patients with hematological malignancies presenting with acute abdomen, even without trauma. Rapid diagnosis, decisive surgical management, and integrated postoperative care—including targeted therapy—are key to favorable outcomes.

Statements and Declarations

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