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Spontaneous splenic rupture – An uncommon complication of chronic myelomonocytic leukemia

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1. Summary

Chronic Myelomonocytic Leukemia (CMML) is a blood malignancy that presents with splenomegaly in up to 40% of the patients; however, a pathological splenic rupture is very rare. We describe a 50-year-old male, who presented to hematology clinic twice during 5-year period, asymptomatic with thrombocytopenia and monocytosis but was non-compliant to any diagnostic workup. He finally presented to us with hypovolemic shock secondary to pathological splenic rupture and subsequent blood loss. Patient underwent emergency laparoscopic splenectomy and stabilized. During the stay at hospital, his complete workup revealed the diagnosis of Chronic Myelomonocytic Leukemia. Although splenomegaly is common in CMML, it rarely result in its rupture and so this complication is often overlooked. This case is reviewed in the light of handful cases described in medical literature previously.

2. Background

Splenic rupture is a rare but a potentially life-threatening complication of chronic myelomonocytic leukemia (CMML). Here, we described sudden rupture of spleen with subsequent hypotensive shock in a 50-year-male having monocytosis for 4 years, finally diagnosed as CMML. The patient was stabilized after an emergency exploratory laparotomy and splenectomy. This case will help physicians avoid missing a rare but fatal complication of CMML, the pathological splenic rupture. In the scenario of a patient with known CMML or with persistent monocytosis presenting with hypovolemic shock accompanied with drop in hemoglobin, a physician should be aware of the possibility of splenic rupture. Purpose of writing the case report is to highlight the importance of early diagnosis and subsequent treatment of a rare but fatal complication of CMML.

3. Case presentation

In 2015, a 50-year- male, presented to hematology outpatient clinic for routine occupational health check. His clinical history was unremarkable. However, examination showed an enlarged spleen of 2 cm below the left costal margin. Initial laboratory investigations showed hemoglobin (Hb) 14 g/dL, total leukocyte count (TLC) 6.2 × 10^9/L with an absolute neutrophilic count 1.8 × 10^9/L, absolute monocytic count 2.3 × 10^9/L and a platelet count 87 × 10^9/L. Further investigations showed HbsAg, Anti HCV and HIV workup as negative. Bone marrow biopsy was advised but the patient was lost to follow-up. He visited outpatient clinic again in 2019 after four years with concerns of his low platelet counts on last visit. He was asymptomatic and systemic examination was unremarkable except the persistence of spleen 2 cm below the left costal margin. His recent laboratory investigations showed hemoglobin 13.4 g/dL, TLC 12.6 × 10^9 /L, with absolute neutrophil count 3.1 × 10^9/L, absolute monocyte count 5.5 × 10^9/L and platelets 81 × 10^9/L. He was re-advised for bone marrow biopsy however, he was non-compliant, but presented again after three months with complains of dizziness, loose stools and severe abdominal pain for last two days. On examination patient was afebrile, anemic, and drowsy but GCS of 15/15, hypotensive with a mean arterial pressure of 50 mm/Hg and heart rate of 140 bpm. Abdomen was tender on palpation in left hypochondriac region.

4. Investigations

Initial laboratory investigations showed Hb 6.2 g/dl, TLC 32.4 × 10^9 /L with 32 % neutrophils, 42% monocytes and platelets 49 × 10^9 /L. Peripheral film showed monocytosis and leucoerythroblastic picture with occasional blast cells.

Blood chemistry showed serum lactate 4.7 mg/dl and CRP 3.3 mmol/L. Ultrasound abdomen demonstrated mild splenomegaly (13cm) with normal parenchyma and mild to moderate ascites. Hematology consult was given for abnormal CBC findings and he was shifted to special care unit for receiving fluid resuscitation and red cells transfusion. A repeat hemoglobin showed a significant drop of 2 gm/dl. Patient was still tachycardic and drowsy with persistence of abdominal pain. With a suspicion of intra-abdominal bleed, general surgery was

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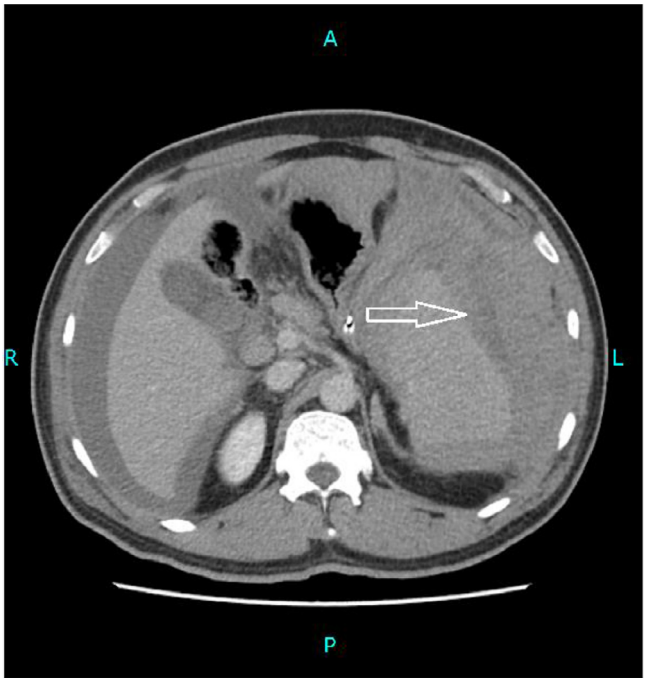

Fig. 1. CT scan abdomen and pelvis with contrast (coronal view) showing hematoma in the perisplenic and sub diaphragmatic region as shown by the arrow.

consulted and an urgent contrast CT scan abdomen /pelvis was performed on their advice. The scan showed a large hematoma measuring 196 × 126 mm in the perisplenic and sub-diaphragmatic region extending into the gastro-hepatic and gastro-splenic regions, compressing the stomach medially. (Fig. 1).

These findings were consistent with ruptured spleen. Splenectomy was performed and histopathological examination showed splenic tissue with areas of hemorrhage and congestive splenomegaly. Bone marrow was done for the first time, which was consistent with the diagnosis of CMML. Bone marrow aspirate was a hypercellular specimen showing increase in granulocytic precursors. Prominent monocytic precursors also noted. Dysplastic monocytes and erthyroid precurosors seen. (Fig. 2)

Bone trephine showed cellularity of around 85-90%. Cellular areas showed diffuse infiltration with monocytoid cells. Normal hematopoiesis was suppressed.

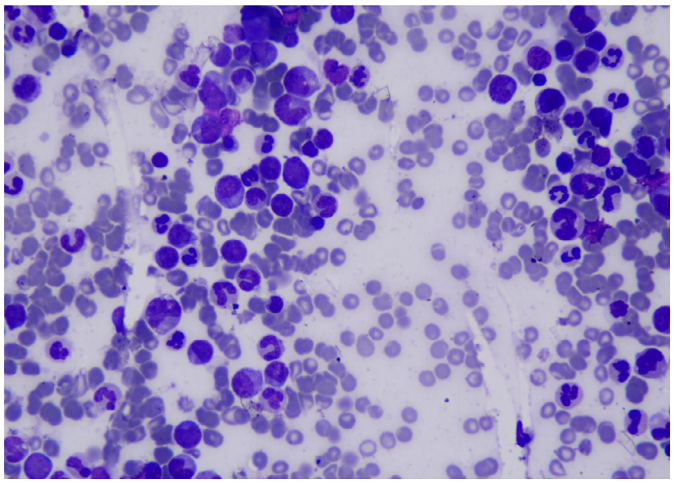

Fig. 2. Bone marrow showing cellular aspirate; dysplastic myeloid, erythroid and monocytic precursors seen. 2 % blast cells seen.

BCR-ABL, PDGFRA rearrangement and JAK2 mutation were ruled out. Bone marrow cytogenetics were normal.

5. Treatment

Patient was taken immediately for exploratory laparotomy and splenectomy. Per operative findings showed a ruptured spleen with multiple large clots. Post operatively patient was shifted to intensive care unit for monitoring. After 72 hours, he was shifted back to ward.

During post-operative period, his TLC rose to 80 × 10^9/L with neutrophilia and monocytosis probably secondary to splenectomy. However, the blood counts normalized gradually in next 7 days following treatment with hydroxyurea. In following few days, patient improved and bone marrow examination and other relevant tests were done as mentioned in investigations. The final diagnosis was CMML stage 0, according to W.H.O. 2016 classification, requiring no immediate hematological intervention and hence patient was discharged for later follow-up as outpatient.

6. Outcome and follow-up

After discharge patient was followed up in clinic. His total leucocyte counts improved. However he developed thrombocytopenia (Platelets 22 × 10^9/L) for which he was started on Azacitidine at 75mg/m2, 7 days cycle in day care. His thrombocytopenia did not improve even after two cycles of Azacitidine, for which thrombopoietin receptor agonist Eltrombopag was started and further Azacitidine was not given beyond two cycles. His last cbc shows Hb 13.0 g/dl, TLC 10.7 × 10^9 /L with 6.6 % neutrophils, 48% monocytes and platelets 42 × 10^9 /L. He has been under blood count surveillance with a plan of allogeneic stem cell transplantation when the patient consents.

7. Discussion

Here, we described a middle-aged patient with CMML who had spontaneous splenic rupture but had a successful outcome due to timely diagnosis and surgical intervention. It is well known that a normal but otherwise healthy spleen does not spontaneously rupture unless challenged with significant trauma. Generally speaking, splenic rupture is rarely observed in hematologic disorders. However, a clinical hematologist should be aware of this rare complication as it carries a significant risk of mortality. Literature review showed spontaneous splenic rupture in malignant (acute lymphocytic leukemia, acute myelogenous leukemia, Hodgkin's / Non-Hogkin's lymphoma, myeloproliferative disorders, myelodysplastic syndromes) as well as benign hematological disorders (histiocytosis and idiopathic thrombocytopenic purpura) [1]. Several mechanisms have been postulated for splenic rupture in these disorders [2]. Most frequent pathology is the direct infiltration of spleen by the malignant cells. This exceeds the capacity of the relatively non-distensible splenic capsule leading to capsular rupture and splenic hemorrhage. It can also be caused by splenic hemorrhage secondary to thrombocytopenia as seen in myelodysplastic syndromes [3]. Finally, splenic infarcts [4], which are not uncommon in hematological disorders may cause architectural changes and altered vascular resistance. These three mechanisms may act in synergy in causing splenic rupture in hematological malignancies [3].

Splenomegaly is commonly found in the myeloproliferative disorders and is seen in 17– 39% of CMML [5–7]. However, despite an enlarged spleen, a patient with CMML rarely suffers from its rupture. Our literature search indicated only six such clinical reports during 1983-2007 [3,8–11]. Similar to this report, Goddard et al described a patient who presented with pallor, hypotension and monocytosis. CT scan showed splenic rupture for which the patient underwent an uneventful emergency splenectomy [3]. Similarly, Diebold J reported a 62 year old man having untreated CMML for 8 years before undergoing spontaneous splenic rupture. He survived the event following

splenectomy. Grossly and microscopically, the spleen showed peliosis [8]. All patients with CMML who had spontaneous splenic rupture underwent emergency splenectomy after early recognition of underlying cause. Some hematologists have considered the role of prophylactic splenectomy to avoid splenic rupture. However, splenectomy is not a risk free procedure, as 3 of 12 patients with CMML who underwent such prophylaxis succumb to postoperative bleeding or wound infections [9]. This signified that splenectomy might not be uniformly helpful to all patients in avoiding rupture.

Clinical hematologists and emergency room physicians should have a high index of suspicion for splenic rupture when a patient with diagnosed CMML or with reported monocytosis presents with abdominal pain and hypotension.

Learning points

- Spontaneous rupture of spleen in CMML is rare but carries significant morbidity and mortality.
- In CMML patients hypovolemic shock accompanied with abdominal pain should raise high suspicion of splenic rupture.
- Most frequent pathology of spontaneous splenic rupture seems to be the infiltration of spleen with malignant cells.
- Prophylactic splenectomy to avoid spontaneous splenic rupture may not be helpful as post-operative bleeding and infections pose a high mortality risk.

Declaration of Competing Interests

The authors declare that they have no conflict of interest.

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