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Spleen Rupture as the First Presentation of Chronic Phase Chronic Myeloid Leukemia. Case Report

Waseem F Al Tameemi[1] *DM, FICMS, CABMS, FIBMS*, **Maan MA Hamid**[2] *CABS, FIBMS*

[1]Dept. of Medicine, College of Medicine, Al-Nahrain University, [2]Al-Kadhimyai Teaching Hospital.

Abstract Spleen rupture is a rare life threatening emergency that mostly occurs in pathologically enlarged spleens. Causes may include infections, congenital cyst, metabolic, degenerative and malignancy (leukemia). Several possible mechanisms have been proposed for spleen rupture in patients with hematological disorders. Most commonly, malignant cells of lymphoproliferative or myeloproliferative origin infiltrate the spleen directly. Their sheer volume exceeds the capacity of the relatively non-distensible splenic capsule, causing splenic rupture and splenic hemorrhage.

In this report we present a 34 years old male patient presented with an acute abdomen after 3 days from sustaining blunt hit over the abdomen. After immediate resuscitation, exploratory laporotomy had done that revealed old altered blood in the peritoneal cavity and large spleen with non-bleeding laceration in addition to hepatomegaly and dilated mesenteric veins. Evacuation of the clotted blood was done with splenectomy.

Key word Massive splenomegaly, chronic myeloid leukemia, spleen rupture

Background

Chronic myeloid leukaemia (CML) is a BCR-ABL1+ve myeloproliferative neoplasm results from an acquired genetic change in a pluripotential haemopoietic stem cell [(1,2)].

It results from translocation of ABL1 on chromosome 9 to the region of the BCR gene on chromosome 22 (Philadelphia chromosom) [(1,2)].

It occurs in about 1.0 – 1.5 per 100 000 of the population per annum, with a median age of onset is 50 – 60 years [(1)].

Most cases of CML are diagnosed in the chronic phase [(1,2)] as the majority of patients presented with symptoms, usually attributable to splenomegaly, haemorrhage or anaemia, however asymptomatic patients are reported .The patient may have severe pain or discomfort in the splenic area, often associated with splenic infarction [(1)].

The peripheral blood smear shows leukocytosis; owing to neutrophils in different stages of maturation [(1-3)]. Blasts are usually less than 2% of the leucocytes, but absolute basophilia is invariably present [(1,2)]. Bone marrow specimens show increased cellularity owing to granulocytic proliferation with a maturation pattern similar to that in the blood, and blasts account for fewer than 5% of the nucleated bone marrow cells [(1-3)].

We report a case of a Philadelphia positive chronic myeloid leukemia which presented

with spontaneous splenic rupture as the initial manifestation of the disease.

Case presentation

A 34 years old male patient presented to emergency ward complaining of an upper abdominal pain following simple hit by blunt object 3 days before.

It starts as suddenly as sharp pain at the upper abdomen radiated to left shoulder that embarrassed his breathing. Initially it managed conservatively with pain killer. On the 3th post-trauma day, the patient developed an increasing agonizing abdominal pain and consulted the emergency department at Al-Kadhimiya teaching hospital.

His story dated back to the last 6 months where he bothered by abdominal distention mainly after meal associated heaviness like pain over the left upper quadrant, in addition to intermittent low grade fever and unexplained bone aches. There was no jaundice. He reported weight loss of 6 kilogram but he didn't seek any medical advice.

Past medical history was unrevealing and no special drug history and not smoker.

The emergency surgical team received the patient as an acute abdomen. Initially the pulse is bounding with rate of 116/minute, blood pressure 100/60 mmHg, and respiratory rate 25 cycle /minute, shallow and thoracic in character in fully conscious anxious patient. Abdomen was tender with board like rigidity and negative bowel sounds. Immediate resuscitation offered including intravenous lines with administration of normal saline and meanwhile abdominal US was done for him which revealed the presence of large amount of clotted blood (thick fluid) with huge splenomegaly and moderate hepatomegaly. A decision for surgical operation was made. Two pints of blood prepared and consent was taken.

Exploratory laparotomy was made through a midline incision, old altered blood found in the peritoneal cavity, large spleen with non-bleeding laceration (Figure 1) and hepatomegaly with dilated mesenteric veins. Evacuation of the clotted blood was done with splenectomy (Figure 2) and liver biopsy.

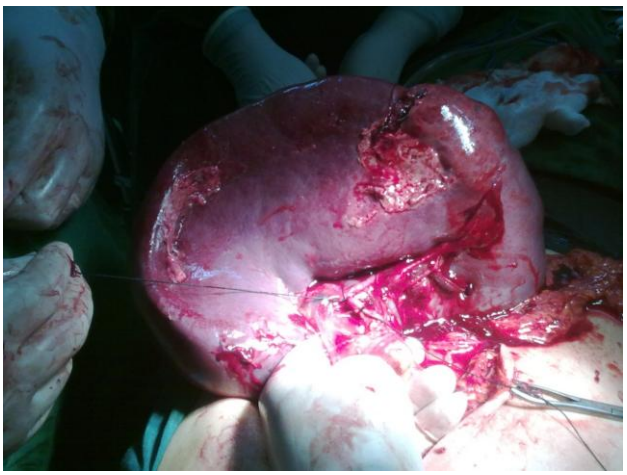

Figure 1: peritoneal cavity with altered blood and laceration in splenomegaly

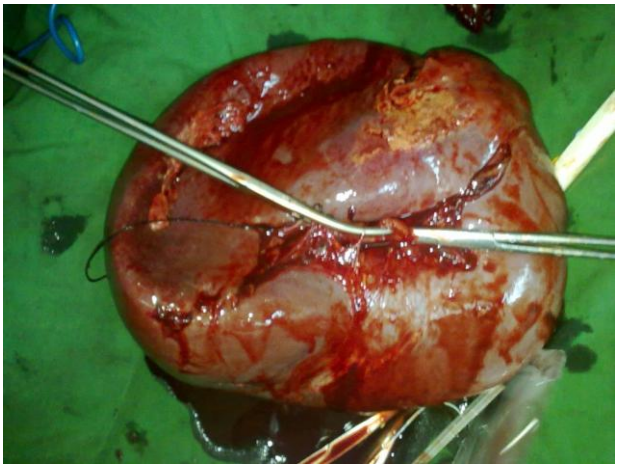

Figure 2: Splenctomy

The patient had a smooth postoperative recovery without major complications. Post operatively; re-evaluation investigations: Hb 8.3 g/dl, WBC 139 x 10^9/ml (differential count; N: 44%, L: 3%, M: 3%. E: 2%, B: 3% with presence of immature granulocyte as metamyelocyte 4%, myelocyte 37%, promyelocyte 1% and blast 3%) and platelet of 1723 x10^3/ml

Blood film confirms leucocytosis with basophilia, left shift and double peaks in granulocyte series, with marked thrombocytosis.

Bone marrow study including aspiration and biopsy revealed hyper cellular marrow with increase in M: E ratio. Erythropoiesis, graulopoiesis (in all stage of maturation) and megakaryopoiesies were hyperactive and blast cells formed only 3% with no increasing in fibrosis.

Peripheral blood sample in heparinised tube were aspirated for FISH study to look for Philadelphia chromosome (BCR-ABL proto-oncogen) and it revealed that t (9; 22) BCR-ABL fusion proto-oncogen presented in 93% metaphases.

Histological examination of the spleen and liver biopsy was that of chronic mylocytic leukaemia with liver infiltration.

Other laboratory assays reveal elevated serum uric acid 480 micromol/l, with normal liver function test and normal renal function. Post operative ultrasound was normal apart from splenectomy. Cardiac assessment was normal by ECG and echocardiography.

Initial treatment given in addition to antibiotics includes hydroxyurea 2000 mg /day with aspirin 100 mg /day and allopurinol 300 mg /day maintained for 10 days until preparation of imatinib mesylate therapy which had been used in dose of 400 mg /day. Initial re-assessment 3 weeks later reveled normalization of CBC as Hb. 13 g/dl, WBC 10 000/ml (differential count: N: 65%, L: 30%, M: 1%, B: 2% and E: 2% with no immature cells. Platelet of 800 000/ml.

Discussion

Splenic rupture is a rare clinical entity with grave consequences, if unrecognized and untreated. It mostly occurs in pathologically enlarged spleens but cases of spontaneous rupture in a histologically proven normal spleen have been reported [3].

Spontaneous splenic rupture has been reported as the presenting symptom in patients with CLL [4], lymphoma [5] and acute myeloid leukaemia [6].

The incidence of splenic rupture in leukemia is about 0.72% [2]. Although the mortality rate is high, prompt diagnosis and appropriate surgery can save the life of the patient [3-11].

Rupture of the spleen is usually associated with trauma [3-11]. Causes of pathological rupture [3-11] of the spleen have been reported as follows: [1] infections [i.e., viral (infectious mononucleosis), parasitic (malaria), and bacterial (abscess)], [2] congenital (i.e., cyst), [3] metabolic (i.e., Gaucher's disease), [4] degenerative (i.e., amyloidosis), [5] malignancy (i.e., leukemia).

Several possible mechanisms have been proposed for splenic rupture in patients with haematological disorders. Most commonly malignant cells of lymphoproliferative or myeloproliferative origin infiltrate the spleen directly. Their sheer volume exceeds the capacity of the relatively non-distensible splenic capsule, causing splenic rupture and splenic haemorrhage [7].

Three mechanisms of rupture of the spleen in leukemia [8-11] were described as follows: [1] mechanical effect of distension secondary to leukemic infiltration of the spleen, especially the capsule; [2] splenic infarct with capsular hemorrhage and subsequent rupture; [3] defects in blood coagulation.

Rupture probably results from a combination of these mechanisms rather than from any single mechanism

The choice of treatment for spontaneous splenic rupture is not only determined by haemodynamic stability, amount of blood products used but also by the underlying pathology [8].

Splenic artery embolisation may be used as an adjunct to non-surgical management of splenic injury [9].

There has been a shift towards non-operative management in haemodynamically stable patients to reduce risk of post splenectomy infection [10].

When surgery is undertaken, spleenoraphy, partial or total splenectomy may be performed depending on the extent of injury. Abdominal compartment syndrome may be another indication for emergency laparotomy in

patients with massive intra-abdominal or retroperitoneal haemorrhage [8-11].

There was evidence of a coagulation defect in this CML patient; platelet and von Willebrand's factor dysfunction which could presumably have played a role in the pathogenesis of splenic rupture [11].

References

1. Goldman JM, Mughal TI. Chronic myeloid leukemia. In Hoffbrand AV, Catovsky D, Tuddenham EGD and Green AR: Post Graduate Hematology, 6th ed. Wiley-Blackwell, 2011; p. 27, 483.
2. Vardiman JW. Chronic Myelogenous Leukemia, BCR-ABL1+. *Am J Clin Pathol* 2009; 132: 250-260.
3. Paulvannan S, Pye JK. Spontaneous rupture of a normal spleen. *Int J Clin Pract* 2003; 57: 245-246.
4. Wilson KS. Spontaneous splenic rupture and pseudo-hyperkalaemia in chronic lymphatic leukaemia. *Postgraduate Med J* 1976; 52: 470-472.
5. Gennai A, Basili G, Lorenzetti L, et al. Spontaneous rupture of the spleen in non-Hodgkin lymphoma: a case report. *Chir Ital* 2008; 60: 739-744.
6. Tan A, Ziari M, Salman H, et al. Spontaneous rupture of the spleen in the presentation of acute myeloid leukemia. *J Clin Oncol* 2007; 25: 5519-5520.
7. Bauer TW, Haskins GE, Armitage JO. Splenic rupture in patients with hematologic malignancies. *Cancer* 1981; 48: 2729-2733.
8. Jafferbhoy S, Chantry A, Atkey N, et al. Spontaneous splenic rupture: an unusual presentation of CML. *BMJ Case Reports* 2011; doi:10.1136/bcr.02.2011.3879
9. Sclafani SJ. The role of angiographic hemostasis in salvage of the injured spleen. *Radiology* 1981; 141: 645-650.
10. Nix JA, Costanza M, Daley BJ, et al. Outcome of the current management of splenic injuries. *J Trauma* 2001; 50: 835-842.
11. Canady MR, Welling RE, Strobel SL. Splenic rupture in leukemia. *J Surg Oncol* 1989; 41(3):194-197.

Correspondence: Dr. Waseem F Al Tameemi
drwaseem72@hotmail.com
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