

Unusual Presentation of Chronic Myeloid Leukemia as Retroperitoneal Hematoma: A Case Report

Yasir Bashir¹, Fahim Manzoor², Shuaeb Bhat², Nusrat Bashir^{2*}, Shabeer Ahmad³, Sajad Geelani², Javid Rasool²

¹Department of Critical Care Medicine, Sher -I -Kashmir Institute of Medical Sciences, Soura, Srinagar, Jammu and Kashmir, India

²Department of Haematology, Sher -I -Kashmir Institute of Medical Sciences, Soura, Srinagar, Jammu and Kashmir, India

³Department of General Surgery, Sher -I -Kashmir Institute of Medical Sciences, Soura, Srinagar, Jammu and Kashmir, India

*Corresponding author: bashimusrat@gmail.com

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Abstract Chronic myeloid leukemia(CML) usually remains asymptomatic and has varied clinical presentation. The disease usually has a chronic course till it goes into blast crisis when it develops fever, other constitutional symptoms and bleeding manifestations as seen in acute leukemia. Here, we present a case that came to our emergency department as a case of acute abdomen in the form of massive retroperitoneal hematoma and subsequently proved to be a case of Philadelphia positive CML with blast crisis.

Keywords: Chronic myeloid leukemia, retroperitoneal hematoma

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1. Introduction

Chronic myeloid leukemia is clonal expansion of a hematopoietic stem cell, possessing a reciprocal translocation between chromosomes 9 and 22. [1] The age adjusted incidence rate in the United States is approximately 2.0 per 100,000 persons for men and approximately 1.1 per 100,000 persons for women. [2] Acquisition of the BCR-ABL fusion gene as a result of the t (9;22) (q 34; q 11.2) in a single multipotential hematopoietic cell results in the CMLstem cell, necessary for the initiation and maintenance of the chronic phase of CML. [3,4] Retroperitoneal hematomas are usually seen in patients with history of trauma [5] and those who are on anticoagulants with incidence of 0.6 to 6.6%. [6,7,8,9] As management of idiopathic retroperitoneal hematoma is usually conservative, early diagnosis can save the patient an unnecessary exploration. This improves morbidity and mortality in this group of patients [10].

2. Case Report

A 35 year old male presented to the casualty department with history of vague abdominal discomfort and giddiness. On examination the patient had pulse rate of 130 per minute and blood pressure of 90/60 mm of Hg. General physical examination revealed pallor, sternal tenderness and massive splenomegaly. Investigations revealed that he had hemoglobin of 5.6 grams per deciliter, total leukocyte count of 320,000 per cubic millimeter; platelets were 70, 000 per cubic millimeter and differential

leukocyte count showed Neutrophils 50%, Lymphocytes 13%, Basophils 4%, Myelocytes 23%, Promyelocytes 5% and Blasts 5%. Initial diagnosis of CML with possible sepsis was made. On routine ultrasound examination of the abdomen, suspicion of retroperitoneal hematoma was made and was later confirmed by computed tomography (CT scan) {Figure 1, Figure 2}. Coagulation profile of the patient revealed Prothrombin time of 20 seconds and activated partial thromboplastin time of 38 seconds. Patient was resuscitated with bloodproducts and was started on allopurinol, hydration and imatinib after blood and bone marrow samples for BCR-ABL were taken. Bone marrow examination showed 26% blasts {Figure 3} and BCR-ABL was 100%. Patient stabilized and went into chronic phase with imatinib but the improvement lasted for a short time and the patient was advised to go for allogenic stem cell transplant.

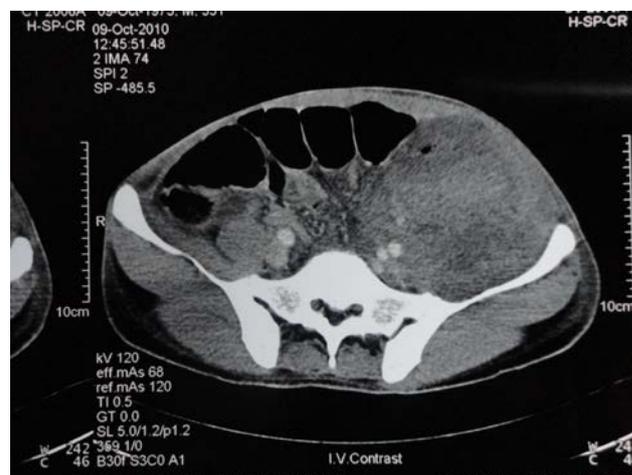


Figure 1. CT scan abdomen showing retroperitoneal hematoma

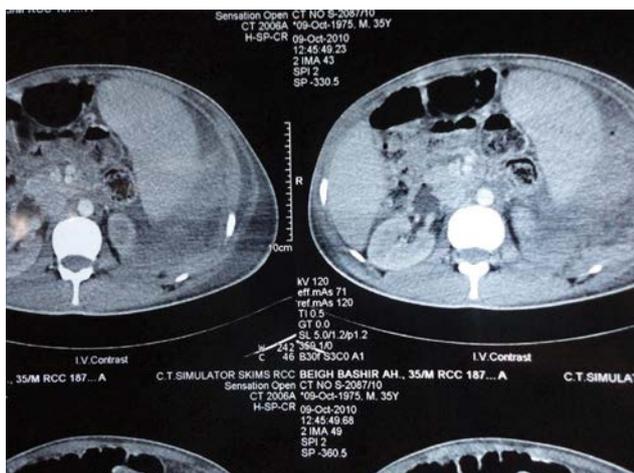


Figure 2. CT scan abdomen showing retroperitoneal hematoma

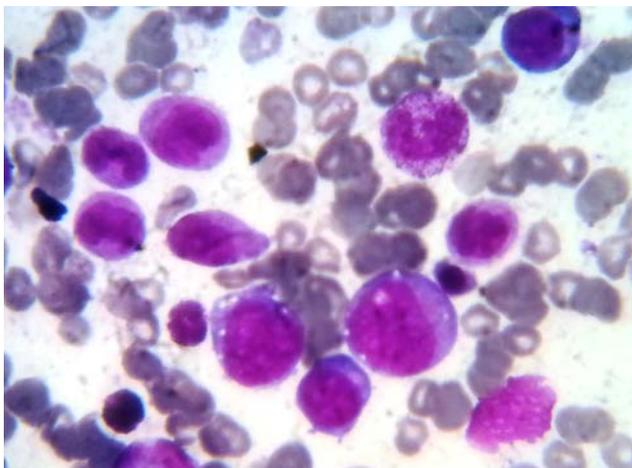


Figure 3. Bone Marrow Aspirate showing CML in Blast Phase

3. Discussion

The predilection for bleeding into the retroperitoneal space has not been fully explained but a unique weakness of the vascular and connective tissue has been suggested. [9] There are several well-recognized causes of retroperitoneal hematoma, including ruptured aortic aneurysm, traumatic vascular injury, retroperitoneal neoplasms, and coagulopathy. Acute leukemias usually present with features of cytopenias and these symptoms are less commonly seen in patients with chronic myeloproliferative disorders. Etiologies as well as the precise mechanisms leading

to spontaneous retroperitoneal hematoma (SRH) are unclear in most of the reported cases. Tumors, particularly renal cell carcinoma and angiomyolipoma, are the most common cause of SRH, occurring in 57–73% of cases. [11] The overall prevalence of SRH as a complication of tumors, however, is low. In renal cell carcinoma, it occurs in only 0.3–1.4% of cases [12], although the incidence is much higher in angiomyolipoma, occurring in 13–100% of cases, depending on tumor size. Aneurysms of the visceral circulation as a cause are rare, accounting for 0.1–10.4% in autopsy statistics. The exact mechanism of rupture of branches of splanchnic vessels is unknown, but likely represents weakness of the tunica media,

predisposing rupture in the face of abrupt increases in pressure. Pathology specimens regularly exhibit disruption of elastic lamellae. Spontaneous hemorrhage can be seen with inflammatory erosive processes which explain the association with necrotizing arteritis in polyarteritis nodosa and rheumatoid arthritis. Diagnosis via CT scan is the principal method of diagnosis but it is practical only in hemodynamically stable patients. It helps in establishing the site, size, and likely underlying causes. [13] CT angiography of vessels has proven useful as a screening tool using small amounts of contrast to elucidate sites of active bleeding. Computed tomography can miss segmental arterial mediolysis as a cause of spontaneous retroperitoneal hemorrhage [14].

4. Conclusions

We present an unusual case of Chronic myeloid leukemia who presented with retroperitoneal hematoma. Our patient did not have any clinical features suggestive of several well-recognized causes of retroperitoneal hematoma such as ruptured aortic aneurysm, traumatic vascular injury, retroperitoneal neoplasms. Neither did the patient have any clinical features suggestive of vasculitis. The CT scan of the abdomen did not show any major vessel breach. The patient was in blast phase of CML and had developed thrombocytopenia with mildly deranged coagulation profile, which could explain the development of retroperitoneal hematoma. The patient eventually responded well to tyrosine kinase inhibitors.

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