

Case Report

Spontaneous scapular region hematoma extending upto anterior chest wall in a patient of chronic myeloid leukaemia: an unusual site presentation

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ABSTRACT

Spontaneous haemorrhage and thrombosis should always raise the suspicion of CML as one of the differential as it is associated with qualitative platelet dysfunction. We here report a rare case of CML with spontaneous hematoma at an unusual site. A 26 years male presented with complaints of sudden onset non traumatic spontaneous swelling over the left scapular region and progressively extending up-to the left anterior chest wall which was associated with dull aching moderate intensity pain. Swelling was warm, soft in consistency and tender. He also had hepato-splenomegaly. General blood picture showed marked leucocytosis with myeloid hyperplasia. CT scan thorax reported a large hypodense lesion in the scapular and anterior chest wall region which on diagnostic aspiration revealed blood thereby confirming the diagnosis of spontaneous hematoma with CML. Imatinib therapy was started and hematoma was managed conservatively with antibiotics. The swelling resolved spontaneously without any surgical intervention.

Keywords: Chronic myeloid leukaemia, Philadelphia chromosome, Hematoma, Platelet dysfunction, Imatinib, Chest wall swelling

INTRODUCTION

Chronic myeloid leukaemia (CML) is a liquid tumour that occurs due to reciprocal translocation between chromosome 9 and 22 (Philadelphia chromosome) leading to the birth of a fusion gene bcr-ABL that up-regulates tyrosine kinase activity and inhibits apoptosis. It is characterised by exuberant leucocytosis (myeloid series hyperplasia), erythroid hyperplasia and thrombocytosis. Usually, it is more common in older age males over 65.¹

Bleeding diathesis developing in CML is due mostly to qualitative platelet dysfunction resulting in thrombotic and haemorrhagic manifestations at various sites; joints, mucus membranes, conjunctiva and subdural space.²⁻⁴

We here describe a case of spontaneous scapular region hematoma extending upto the anterior chest wall, a very unusual site for hematoma formation, in a diagnosed case of CML. Very few similar cases of spontaneous hematoma in CML have been reported till date.⁵⁻⁷

CASE REPORT

A 26 years old male, presented to emergency department with complaints of sudden onset swelling over the left scapular region and progressively extending upto left anterior chest wall. The swelling was associated with dull aching moderate intensity pain. There was no history of trauma, fever, cough, breathlessness, haemoptysis, any drug intake and bleeding from other site. He had similar

episode of spontaneous swelling on same site 6 months back for which he visited some local practitioner where aspiration of swelling was done accounting for 300-400 ml blood leading to resolution of swelling. His family history was insignificant. He had no addiction to alcohol, tobacco or any other drug. There was no history of concurrent medical illnesses.



Figure 1 (A) (B): Swelling over the left scapular region extending up-to the left anterior chest wall.

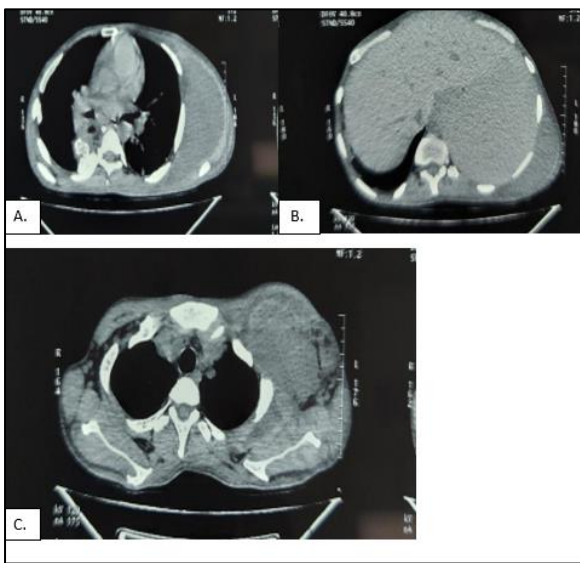


Figure 2(A, B, C): Large hypodense lesion in chest wall diffusely on left side extending from infraclavicular region, in prescapular region to lower chest wall and in abdominal wall in left hypochondrium.

On examination, vital signs were within normal limits with blood pressure 126/78 mmHg, pulse rate 108/minute, afebrile and well oriented to time place and person. General examination revealed mild pallor. Local examination revealed that the swelling was warm, soft in consistency, tender and extending from left scapular region to left anterior chest wall of approximately 15×10 cm in the scapular region and 10×10 cm on anterior chest. He also had moderate splenomegaly; 8 cm below left costal margin and hepatomegaly; 3 cm below right

costal margin. He had no petechiae, purpura, ecchymosis, skin discoloration or any swelling over other parts of his body. Other system examinations were within normal limit.

Diagnosis

Chronic myeloid leukaemia (CML) with spontaneous soft tissue hematoma.

Table 1: Investigations.

Haemoglobin	10.6g%
TLC	120100/cumm
Platelets	4.2lac/cumm
MCV/MCH	89/86
d-dimer	3.47
Fibrinogen	189mg%
Prothrombin time/ INR	14.3 seconds/1.06
General blood picture	R.B.C.-normocytic, normochromic; W.B.C.s- tlc >4 lac per mm ³ Myeloid Left shift (blast :1, promyelocyte:8, myelocyte :25, metamyelocyte:13)
BCR/ABL1: ABL1 ratio	Few megakaryocyte present) 46.302%
USG whole abdomen	Gross hepato-splenomegaly with dilated portal venous system

Treatment and follow up

Patient was started on tablet Imatinib 400 mg O.D. and hydroxyurea. He did not undergo active surgical intervention for the hematoma. The swelling spontaneously regressed in size with decrease in associated pain. Patient was discharged on the same treatment and was advised follow up after which he did not come.

DISCUSSION

Myeloproliferative disorders are associated with thromboembolic and haemorrhagic complications very often due to the underlying disease process itself. Baseline mutation in CML is the cause for abnormal platelet structure and function. Clonal expansion of megakaryocytes harbouring the 9:22 translocation have defective aggregation and can also develop acquired Glanzman thrombaesthesia phenotype.⁸ Autoantibody against the platelets membrane proteins; gp 2b3a, defective PI3 kinase pathway, alpha2b-beta 3 downregulation is also considered an etiology.^{9,10} Jain has reported a case of CML with acquired von willebrand disease presenting as subdural hematoma.^{11,12} Apart from the platelet abnormalities, studies have suggested low red

blood cell counts and extramedullary hematopoiesis also as the contributing factors.^{13,14}

Imatinib therapy serves as the cornerstone for the CML treatment leading to the disease control without cure. Being a tyrosine kinase inhibitor (TKI) it directly antagonizes the bcr-ABL gene product and produces a good hematologic and cytogenetic response. In our case also the hematoma resolved spontaneously after imatinib therapy commencement and thereby proving its effect on the platelet dysfunction too. Rothe et al have also reported the reversal of platelet dysfunction after imatinib therapy.¹⁵

Contrary to this fact, several studies have shown mixed effect of tyrosine kinase inhibitors on platelet dysfunction in CML. Imatinib induced bleeding diathesis is well reported in literature.^{16,17} Akay et al has shown that platelet dysfunction in CML is multifactorial and imatinib inhibits the ristocetin induced platelet aggregation that may cause bleeding diathesis.¹⁸ However Sener et al in their study could only conclude that TKI induced platelet dysfunction was mainly in vitro with hardly any significant in vivo bleeding events.¹⁹ Hence in a nutshell TKI therapy is not absolutely contraindicated in CML with hemorrhagic events and should be considered as the first line treatment. In our case also the existing hematoma did not increase in size and no new bleeding complication happened after imatinib commencement. Surgical approach in such cases is selected only in presence of complications like expanding hematoma, superimposed infection, abscess or mass effect at vital locations.

CONCLUSION

Spontaneous hematoma formation is usually a finding in known bleeding disorders like haemophilia. Hence investigative work up in such cases is mainly restricted to searching for clotting factor deficiencies and platelet dysfunction. Therefore, it is important to step out of this tubular vision and start with the basic routine blood tests like a complete blood count and general blood picture which can give a very important clue about the blood cells and thereby changing the entire management. Spontaneous bleeding occurs in chronic myeloid leukaemia due to qualitative platelet dysfunction from genetic mutation, so targeted therapy is the mainstay. Surgical evacuation should be instituted only in case of a life threatening emergency.

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