



SPONTANEOUS CHEST WALL HEMATOMA- AN UNCOMMON PRESENTATION OF CHRONIC MYELOID LEUKEMIA

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ABSTRACT

Introduction & Background: Spontaneous hematomas are most commonly caused by bleeding and clotting disorders followed by anticoagulant and antiplatelet drugs. Spontaneous hematoma secondary to malignancy is usually uncommon. CML is the hematological malignancy known for variability in presentation. Spontaneous hematomas are unusual presentation of CML with only few cases reported. **Case History:** A 63 years old male, presented with complaint of sudden painful, boggy swelling over the left chest wall few hours following trivial trauma. No history of drug intake, joint swelling following trauma or similar episodes in past. No history of similar illness in family members. On examination he was pale, the vital signs were within normal limits. The swelling was warm, firm, tender and overlying skin was reddish. He had a huge, firm, non-tender splenomegaly. He had no petechiae, purpura, ecchymosis or other swelling over his body. Other system examinations were within normal limits. **Investigations:** Hemoglobin- 7.6 g%, TLC- 474,300/mm³ with immature Neutrophil precursors, suggestive of CML. Liver and renal function tests were normal. PT/INR and APTT were not significantly raised. Serum LDH was 859 and serum Uric Acid was 7. Chest X-ray showed homogenous opacity in left lung field. USG of chest wall was suggestive of intramuscular hematoma extending upto lower scapular region. USG abdomen revealed gross splenomegaly (22 cm). Bone marrow examination was suggestive of CML in chronic phase. Karyotyping revealed Ph⁺ chromosome in 100 % cells. Chromosomal study showed BCR-ABL gene positive CML (ISNCN- 52.45 %). **Management:** Chest wall hematoma resolved spontaneously with conservative management within 3 weeks. Patient was started on Hydroxyurea for cytoreduction and later on Imatinib was started. **Conclusion & Discussion:** Few cases of spontaneous hematoma with CML have been reported till now. However, the exact mechanism of spontaneous hematoma in CML is not known. Further research is required to understand the exact mechanism and its management.

KEYWORDS: CML, Hematoma, BCR-ABL.

INTRODUCTION

Spontaneous hematomas are most commonly caused by bleeding and clotting disorders, followed by anti-coagulant and anti-platelet drugs.^[1,2] Spontaneous hematoma is only rarely associated with neoplasm, aneurysm and arteriovenous malformations.^[2] CML is the commonest haematological malignancy in clinical practice; and is known for variable presentation.^[3] Spontaneous hematomas are not a usual presenting symptom of CML.^[1,2,4] Patients of CML presenting as spontaneous hematomas in chronic phase are unusual and only few cases have been reported till now. We are reporting a case of spontaneous chest wall hematoma as

an uncommon presentation of Chronic Myeloid Leukemia.

CASE REPORT

A 63 years old male, presented to the Medicine OPD with complaint of sudden onset swelling over the left chest wall extending upto lower scapular region and flanks. The swelling was painful and boggy to feel (Fig. 1, Fig. 2 & Fig. 3). He noticed the swelling appeared spontaneously following pain which was dull aching and localised over the area while he was weeding his garden. There was no history of trauma, fever, any drug intake, bleeding from any other site or similar spontaneous

swelling. There was no history of similar illness among the family members.



Fig. 1.



Fig. 2.



Fig. 3.

On examination, he was found to be pale. Vital signs were within normal limits with blood pressure of 130/80 mmHg. Local examination revealed that the swelling was warm, firm in consistency, tender to touch and overlying skin was reddish. On abdominal examination he had a huge, firm, non-tender splenomegaly (8cm below the left costal margin along its long axis). He had no petechiae, purpura, ecchymosis or other swelling over the other parts of his body. Other system examinations were within normal limits. On routine investigations, his Haemoglobin- 7.6g%; TLC- 4,74,300/mm³ [differential count- N24L03E01M0Baso03Blasts01ProMy03My17Mm22Stab forms26]; few nucleated RBCs (03/100 WBCs); Platelets- 3,09,000/mm³. Blood picture was suggestive of CML in chronic phase. Liver and Renal function tests were normal. PT/INR (18secs./1.51secs.) and APTT (43secs.) were not significantly raised. BT (2mins.15secs.) and CT (4mins.45secs.) were normal. Serum LDH was 859 IU/L and Serum Uric Acid was 7 mg/dl. Chest x ray revealed homogenous opacity in the left lung field. USG of chest wall revealed hypoechoic, well defined intramuscular hematoma in left chest wall extending upto lower scapular region (219x53.8 mm²). USG abdomen revealed gross splenomegaly (22 cm).

Bone marrow was hypercellular with M:E ratio of 9:1. Myeloid series were predominantly neutrophils, myelocytes and metamyelocytes. Erythroid series were normoblastic; suggestive of CML in chronic phase. Karyotyping revealed Ph⁺ chromosome in 100% cells. (Fig. 4) Molecular study showed BCR-ABL gene positive CML (ISNCN- 52.45 %).

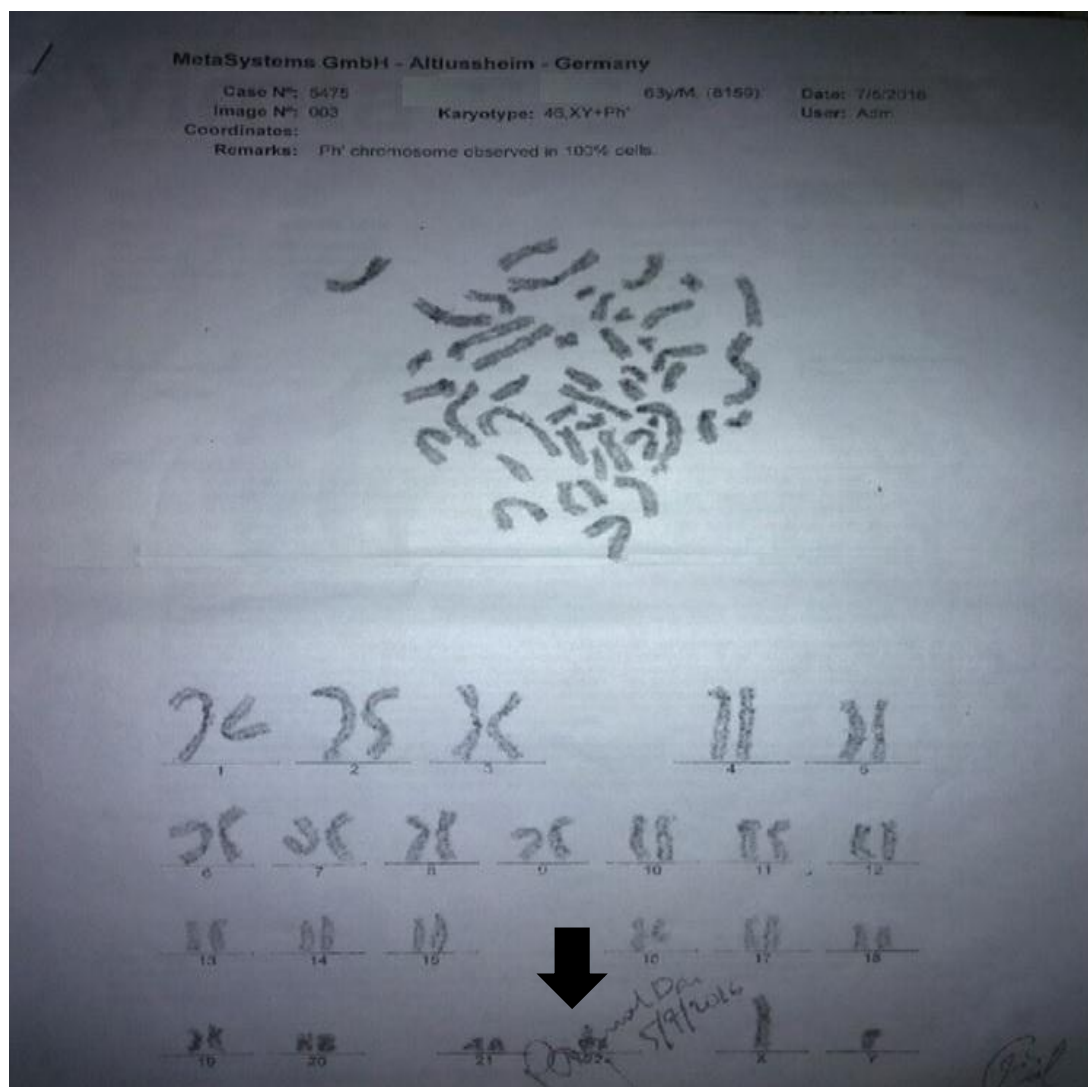


Fig. 4.

Chest wall hematoma resolved spontaneously with conservative management within 3 weeks. He was transfused with 4 units of PRBCs. Patient was started on Hydroxyurea for cytoreduction and later on shifted to Imatinib (400 mg). He did not develop further hematomas or bleeding and is doing well in regular follow-ups.

DISCUSSION

Chronic myeloid leukemia is a clonal hematopoietic malignant disorder, with an acquired genetic defect in the pluripotential stem cell population.^[4] It is characterized by the presence of Philadelphia chromosome (Ph), which is formed by reciprocal translocation between long arm of chromosomes 9 and chromosomes 22 {t(9;22)(q34;q11)} leading to formation of Philadelphia chromosome with BCR-ABL 1 fusion gene. This gene is responsible for the formation of 210 KDa chimeric protein with enhanced tyrosine kinase activity than normal which prevents apoptosis.^[5] The incidence of CML is reported as between 1-2 cases/100,000/year, without major geographical differences. The mean age at diagnosis is about 60-65 years.^[6]

In up to 50% of asymptomatic cases the diagnosis is made incidentally from a routine blood count. The common presenting symptoms are weight loss, lassitude, anorexia, night sweats, pallor, dyspnoea, palpitation, abdominal discomfort due to splenic enlargement and gout.^[7] Cutaneous and mucous membrane bleeding are common in the form of bruising, purpurae, petechiae, ecchymosis, epistaxis, menorrhagia or haemorrhages from other sites because of abnormal platelet function. Patient may present with infections due to dysfunctional neutrophils and because of leukocytosis there may be features of hyper viscosity syndrome e.g. veno-occlusive disease, cerebrovascular accident, myocardial infarction, priapism, visual disturbances and pulmonary infarction.^[1,8] In the extensive review of literature, Kuo YC et al reported a case of CML who presented with mediastinal hematoma and hemothorax^[9], Bauduceau O et al reported a case of spontaneous hematoma of the iliac psoas muscle in CML.^[10] There was no mention of any precipitating event prior to development of hematoma. Peter George et al reported a case of CML with spontaneous chest wall hematoma following trivial strain prior to the development of hematoma.^[11] In our

patient hematoma developed following dull aching local pain while he was weeding his garden but there was no history of any obvious trauma or strain. Manoj Lakhota et al. recently reported a case of spontaneous soft tissue hematoma involving left thigh treated successfully with hydroxyurea and imatinib.^[7] The mechanism of spontaneous hematoma in CML is still not known. Specific platelet defects including abnormal platelet morphology, acquired storage pool disease, platelet membrane abnormalities, and abnormal arachidonic acid metabolism were thought to be the reason.^[11] It is believed that in CML the platelet dysfunction originated from a clonal expansion of dysfunctional megakaryocytes. These are possibly derived from the identical stem cell from which the CML blasts had originated.^[11] One of the most characteristic platelet defects in myeloproliferative disorders is a reduced responsiveness to epinephrine, while the response to ADP usually is intact. This was found to be due to a reduction in α -adrenergic receptors on the platelet membrane. The cause of the decreased surface expression of α -adrenergic receptors is unknown, though.^[12] Molecularly, BCR-ABL rearrangements have clearly been demonstrated in megakaryocytic cell lines as well as in megakaryocytes from the vast majority of CML patients.^[13,14,15,16]

Thus the treatment targeting BCR-ABL would be equally effective in reducing the CML blasts and dysfunctional megakaryocytes. This is validated by observation that using tyrosine kinase inhibitors in patients with CML would improve the platelet dysfunction.^[12,17] Therapeutic approaches in Ph⁺ CML to reduce the number of CML blasts previously had proven effective in reducing the number of BCR-ABL⁺ megakaryocytes.^[14,16]

CONCLUSION

Spontaneous chest wall hematoma although not rare, is an uncommon presentation in CML. Only few cases of have been reported till now. Details of the mechanism causing spontaneous hematoma is not known but platelet dysfunction is considered as the cause of spontaneous hematoma formation. However, further research is required to establish the definite pathophysiology behind spontaneous hematoma formation. Standard treatment with tyrosine kinase inhibitors in patients with CML leads to spontaneous resolution of hematoma and improves the quality of life. Study to look for platelet function improvement following tyrosine kinase inhibitor therapy should also be considered.

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