

# Spontaneous chest wall haematoma in chronic myeloid leukaemia

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Chronic myeloid leukaemia can have different presenting signs. Here, we present a case of nontraumatic spontaneous chest wall haematoma in an 84-year-old man under investigation for myeloproliferative disorder before his admission. He was admitted with a 2-day history of sudden onset posterior chest wall pain and progressive bruises. Computed tomography showed a chest wall haematoma, while blood tests found a mild coagulation abnormality. He required fresh frozen plasma and red blood cell transfusions as part of his management, but his symptoms improved after starting hydroxycarbamide.

## Keywords:

cml, FFP, haematoma, hydroxycarbamide, management, platelets

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## Introduction

Chronic myeloid leukaemia (CML) is a myeloproliferative disorder associated with an acquired oncogene rearrangement (BCR-ABL) encoding for tyrosine phosphokinase. This happens as a result of the reciprocal translocation between chromosome 9 and 22 [1]. It accounts for 6–15% of adult leukaemia cases, with a variable incidence from 0.6 to 2.0 cases per 100 000 inhabitants, and is increasing with age [2].

The initial presentation of CML can have various clinical manifestations. Most patients present with leucocytosis in a routine blood test, fever, sweats, bone pain, lymphadenopathy, weight loss, splenomegaly, and fatigue. It can also have some unusual presentations including bleeding, thrombosis, priapism, leukaemic infiltration, peptic ulceration, and spinal cord compression [1].

Here, we present a case of nontraumatic spontaneous chest wall haematoma in a patient who was under investigation for the incidental finding of leucocytosis just before his admission.

## Case report

An 84-year-old man with a background history of hypertension and renal impairment presented to the medical admission unit with a 2-day history of sudden onset, sharp right side posterior chest wall pain. It started suddenly while he was going to his bed without any prior history of trauma. He developed redness and bruises the following day. He was referred by his General Physician to see the haematologist 2 months before his admission to further investigate an incidental finding of leucocytosis on a routine blood test.

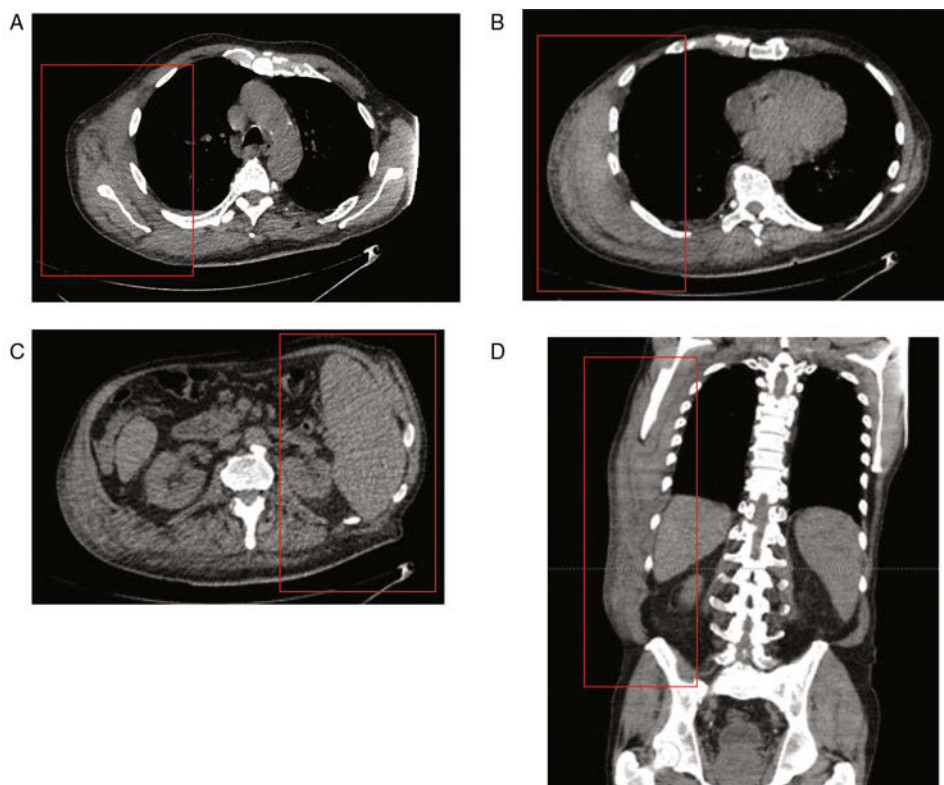
He was haemodynamically stable on admission, with normal blood pressure of 130/60 mmHg. General physical examination revealed bruising and haematoma in the right upper dorsal hemithorax expanding to the lower back and right flank and splenomegaly on deep palpation of his abdomen. His blood results were as follows: WCC  $90.1 \times 10^9/l$ , haemoglobin (Hb) 123 g/l, platelet  $228 \times 10^9/l$ , neutrophils  $73.1 \times 10^9/l$ , MCV 62 fL, reticulocyte count  $176 \times 10^9/l$ , Na 138 mmol/l, K 5.1 mmol/l, urea 18.7 mmol/l, creatinine 377  $\mu\text{mol/l}$ , CRP 18 mg/l, PT 13.4 s, APTT 37 s, fibroblast 2.1 g/l, TCT 26.3 s, reptilase time 15 s, positive plasma D-dimer, bilirubin 10  $\mu\text{mol/l}$ , ALP 146 U/l, ALT 30 U/l, albumin 45 g/l.

His computed tomography scan confirmed splenomegaly and evidence of extensive chest wall swelling over the right upper thorax deep to the lower pole of the scapula extending laterally and inferiorly; consistent with a chest wall haematoma (Fig. 1).

He was managed with joint inputs between the surgical and haematology team. He was managed initially with two units transfusion of Fresh frozen plasma during the first day of his admission. However, his PT and APTT increased to 15.5 and 38.3 s, respectively, and had further 3 units of FFP. His Hb dropped to 92 g/l on the third day and his coagulation factors showed mild coagulation abnormalities (mild deficiency in factor XI, XII, and VII measuring 36.6, 33.9, and 33.4 IU/dl,

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Fig. 1



(a–c) Transverse sections at multiple levels indicating the extent of the haematoma and the splenomegaly. (d) Coronal section further emphasizing the dimensional area involved with the spontaneous haematoma.

respectively). He was given four more units of FFP, two units of red blood cells (RBC) and was started on tranexamic acid. Nevertheless, his Hb further dropped to 75 g/l, despite the transfusion and the absence of any other focal point of bleeding. He was then started on hydroxycarbamide causing his symptoms, as well as his haematoma, to start settling. He was discharged after few more days with subsequent follow-up with the haematologist.

## Discussion

There are different possible factors to explain spontaneous bleeding in our case such as CML, hypertension and renal impairment. Although the patient had a mild deficiency of some coagulation factors, it should not have caused significant spontaneous bleeding with the picture presented on his admission.

Bleeding in association with CML can occur in about 21.3% of cases and most often manifests as excessive haemorrhage from the nasal cavity, bleeding duodenal ulcer, blood per rectum and menorrhagia [3]. It is unusual to present with spontaneous deep tissue or visceral bleeding. Spontaneous nontraumatic bleeding as the first presentation of CML has only been limited to sporadic cases reported in the literature [4–6].

The exact mechanism and the pathology driving spontaneous haematoma in myeloproliferative disorder patients is not clear in most of the reported cases. Different theories have been suggested, through either affecting the morphology and the function of platelets or granulocyte infiltration of organs. The aetiology in our case was probably the result of multiple contributing elements such as the low RBC count, coagulation abnormalities, hypertension, renal impairment and CML itself. Vasculitis was also considered in the pathological process; however, screening was normal in this case.

The previously mentioned factors are discussed below.

### Platelets' dysfunction

Myeloproliferative disorders are associated with complex changes in platelet function. The induced abnormality might be a result of defective platelet production by abnormal megakaryocyte clones. It causes a decrease in membrane glycoproteins (Ib and IIb/IIIa complex), which affect platelet adhesion, aggregation response and subsequent formation of primary clots. It is also linked with a deficiency of platelet granules and altered arachidonic acid metabolism by leucocytes and platelets [7,8].

### Excess granulocyte/infiltration

Spontaneous bleeding in some cases has been related to extramedullary haematopoiesis and granulocyte infiltration of organs. A case of spontaneous haematuria reported in the literature was attributed to the extramedullary haematopoietic process and infiltration of the patient's kidney. He developed a clot in the right collecting system with mild-to-moderate right hydronephrosis to the level of the bladder. He was first managed with continuous bladder irrigation, transfusion, clot evacuation and double-J ureteral stenting. However, he was readmitted with recurrent gross haematuria, which required right nephroureterectomy [9].

### Uraemia

Longstanding uraemia causes impairment in different stages of platelet haemostasis, including adhesion, secretion and aggregation. It causes a functional defect in the interaction of vWf with glycoprotein IIb-IIIa. It affects platelet adhesion to the endothelium by increasing the level of prostacyclin and nitric oxide. In addition, it can cause a defect in platelet secretion through altering arachidonic acid metabolism. The mechanism of uraemic effect and haemostasis is beyond the scope of this case report [10].

### Low red blood cells

Red blood cell count may contribute to the haemostatic plug formation. RBC concentration plays a role in promoting platelets' surface interaction by affecting the flow of platelets in the periphery of the blood vessels. It increases the radial movement of platelets and thus its interaction with the sub-endothelial tissue, due to its relative smaller size compared with RBC. In another study, bleeding time was shown to be prolonged in anaemic patients, independent of their platelet counts, and was shortened by elevating haematocrit [11,12].

### Coagulation defect

The patient had a transient deficiency in factor VII as well as mild persistent deficiency in factors XI (47.9) and XII (31.1), but this should not cause spontaneous bleeding to the degree presented in our case.

Acquired transient deficiency of factor VII can be caused by multiple reasons, and severe bleeding due to acquired isolated factor VII deficiency needs to be managed through a multidisciplinary approach, through intermittent intravenous recombinant

activated factor VII, platelets transfusion, FFP, vitamin K, tranexamic acid and escalating to surgical evacuation of haematoma and decompression of mass effect.

### Summary

Myeloproliferative disorders should be considered in patients who present with spontaneous bleeding, and further studies are required to investigate the functional capacity of platelets in such cases. This might offer insights into the pathogenesis of nontraumatic spontaneous bleeds.

Looking back at the time of presentation, platelet activities could have been assessed using a platelet function assay to give us more understanding of their function at a time when the platelet count was normal. Spontaneous bleeding in myeloproliferative disorders is relevant across most specialities, and the need for haematology input for timely treatment is vital to achieving optimal management.

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### Conflicts of interest

There are no conflicts of interest.

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