A case of polycythaemia vera presenting with intramuscular chest wall haematoma

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Summary: A 40 year old male, previously well, presented with a posterior chest wall haematoma. Computerized tomography and ultrasound showed this to be intramuscular. Haematological indices were consistent with a diagnosis of polycythaemia vera. The haematoma and indices responded to hydroxyurea and venesection. This rare presentation of polycythaemia vera in a young person is described and the haemorrhagic complications of polycythaemia vera discussed.

Introduction

Intramuscular haemorrhage is common in coagulation factor deficiencies but rare in myeloproliferative disorders and particularly so in young patients. Polycythaemia may present with its major complications of either thrombosis or haemorrhage. The bleeding usually presents as ecchymosis, gastrointestinal haemorrhage or epistaxis. In a review of the literature only two cases of intramuscular haemorrhage in polycythaemia vera (PV) have been described and these occurred in patients over the age of 80. We report a 40 year old man with a posterior chest wall intramuscular haematoma as the presenting feature of PV. The haemorrhagic complications of PV are discussed.

Case report

A 40 year old man was admitted as an emergency in September 1990 having developed a large swelling on the right posterior chest wall overnight. He was previously well and there was no obvious history of trauma. His pulse and blood pressure were normal and there was no other evidence of haemorrhage. There was no clinical cardiac or respiratory abnormality. Abdominal examination revealed mild splenomegaly. There was a 26 x 22 x 5 cm tense fluctuant swelling on the right posterior chest wall. There was good range of movement at the neck and shoulder.

Ultrasound and computed tomographic (CT) scan (Figure 1) confirmed the presence of haematoma in the muscle planes. Admission haematological indices showed haemoglobin 22 g/dl, white cell count 14.2 x 10^9/l (neutrophilia), platelets 662 x 10^9/l and haematocrit 0.666. Prothrombin time, activated partial thromboplastin time, factor VIII, fibrinogen, bleeding time, neutrophil alkaline phosphatase, serum B12, platelet aggregation studies and arterial blood gases were normal. Sternal marrow showed an increase in all three cell lines. Red cell mass was 43 ml/kg, plasma volume 42 ml/kg and serum urate 640 μmol/l.

Within 24 hours the haematoma had increased in size and the haemoglobin transiently fell to 14.6 g/dl, white cell count 6.4 x 10^9/l and platelets 769 x 10^9/l due to 'auto-venesection'. The haematoma resolved spontaneously over the next few days with a mild associated rise in serum bilirubin. The patient was treated with hydroxyurea and allopurinol with intermittent venesection. He remained well throughout and 6 months later indices showed haemoglobin 15.2 g/dl, white cell count 7.2 x 10^9/l and platelets 334 x 10^9/l.

Figure 1 CT scan of transverse section of thorax showing intramuscular haematoma.
Discussion

It is reported that less than 5% of cases of PV occur under the age of 40 and that the complication of haemorrhage as the presenting feature occurs in only 1.7% of such young patients. The bleeding is usually mucocutaneous and intramuscular haemorrhage in this age group has not been previously reported as a feature.

The cause of the haemorrhagic tendency in PV is not clear. Patients with myeloproliferative disorders can have normal coagulation but those with thrombocythaemia may have an increased bleeding tendency. Various platelet abnormalities including decreased adhesiveness, defective aggregation and reduced serotonin uptake have been described in association with thrombocythaemia.

References