Bilateral renal vein thrombosis due to inapparent polycythaemia

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Abstract
Thromboses at unusual sites are characteristic of polycythaemia. We present a patient of bilateral renal vein thromboses due to polycythaemia that was inapparent. The diagnosis was confirmed by trilineage hyperplasia in bone marrow and JAK 2 V617F mutation in blood.

Keywords: inapparent polycythaemia; renal vein thrombosis

Background
Thromboses at unusual sites are characteristic of polycythaemia. They include occlusion of the splenic, portal, hepatic and mesenteric veins. There were neither reports of renal vein thrombosis (RVT) due to polycythaemia in adults nor had the aetiology of RVT in adults [1] included polycythaemia as one of the causes.

Case report
A 28-year-old gentleman, non-smoker, teetotaller, non-diabetic and non-hypertensive, presented with the complaints of dysuria, haematuria of one episode and swelling of the left lower limb over 5 days. There was no history of fever, loin pain, oliguria, headache, weakness, pruritus, dizziness, visual disturbances, joint pains, chest pain or breathlessness. Systemic examination was unremarkable. The investigations were in the Table 1.

Discussion
Often patients with splanchnic vein thrombosis, as in this patient, have normal haemoglobin and haematocrit levels. This phenomenon is regarded as ‘inapparent polycythaemia vera’ (IPV) [2]. The concept of IPV is observed in two circumstances. The first one is in patients where increased plasma mass masks an increased red cell mass. This is observed in patients of portal hypertension secondary to suprarehepatic or portal vein thrombosis [3, 4]. In the series of Valla et al [3], among the four patients with portal vein thrombosis and increased plasma volume, the maximum haemoglobin observed was 140 g/L. Gastrointestinal bleeding and/or an increase in portal vein hypertension often explain the apparent normal result of the blood count.

The second one is in the event of a standard iron deficiency during polycythaemia vera (PV) and is partly explained by the high frequency of gastric ulcers and gastritis estimated at 20–30% [5]. Resulting iron deficiency can be accompanied by hypochromia and microcytosis and occasionally a substantial decrease in haemoglobin levels. In this situation, PV is not always easy to diagnose, especially in the absence of splenomegaly and leucocytosis. Thrombocytosis may be present but secondary to iron deficiency. Iron supplementation and treatment of the local cause of bleeding enable the diagnosis to be reestablished, for the increase in haematocrit and haemoglobin levels is abnormal and rapid. [5]

The real frequency of IPV is not yet established; but in one series, its frequency was 18 of 103 patients (17%) [5]. The red cell mass measurement to diagnose IPV is required...
in patients of isolated and unexplained thrombocytosis, hyperleucocytosis, splenomegaly or splanchanic vein thrombosis. [5] But with the advent of the molecular tests for the JAK2V617F mutation, the diagnosis of IPV has been dramatically simplified [6]. Because JAK2V617F is myeloid neoplasm specific and not found in other causes of polycythemia, it has lent itself to being a sensitive diagnostic marker for PV. However, in the context of myeloid neoplasms, JAK2V617F is not specific for PV and is found in ~50% of patients with essential thrombocythemia (ET), primary myelofibrosis (PMF) or ‘refractory anaemia with ringed sideroblasts associated with marked thrombocytosis’ and at a lesser frequency in other myeloid neoplasms but not in lymphoid tumours. Therefore, mutation screening for JAK2V617F cannot be used to distinguish one myeloproliferative neoplasm from another, but it does complement histology in the diagnosis of both ET and PMF by excluding the possibility of reactive thrombocytosis or myelofibrosis [6].

Conflict of interest statement. None declared.

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