A case of Budd-Chiari Syndrome presenting in a lady with newly diagnosed Churg-Strauss Syndrome

SIR, We present a case of Budd-Chiari Syndrome presenting in a lady with Churg-Strauss Syndrome (CSS). A 39-yr-old asthmatic lady presented in 2001 with a 6 week history of rash on her lower limbs. She was pyrexial and her peripheral blood film showed marked eosinophilia (56.9% of total white cell count). She was nauseated and had evidence of a lower limb peripheral neuropathy. On admission, ESR was 4 mm/h; coagulation screen, ANA and ANCA were normal. C3 was normal and C4 was low at 0.04 (normal range 0.19–0.45 g/l). Liver function tests were abnormal with raised transaminases. Skin biopsy confirmed an acute leukocytoclastic vasculitis within the dermis and a bone marrow aspirate showed increased eosinophil production with discreet areas of bone marrow necrosis. A diagnosis of CSS was made; she was commenced on high dose oral corticosteroids and intravenous immunoglobulin to reverse the thrombocytopenia. Her clinical condition improved rapidly and her peripheral eosinophil count returned to normal within 2 days.

Four days following admission, she developed abdominal discomfort and swelling, confirmed by ultrasound to be ascites. A diagnostic paracentesis revealed a low eosinophil count in the ascitic fluid and no organisms. A Doppler ultrasound showed absent flow in several hepatic veins due to thrombosis and a diagnosis of Budd-Chiari syndrome was made. She was commenced on heparin and warfarin resulting in a dramatic improvement in symptoms and repeated ultrasound showed resolution of both thrombus and ascites. Anticoagulation was continued for 6 months after which there was no recurrence of thrombus or ascites. Following discharge she received two intravenous pulses of cyclophosphamide (stopped after development of a macular-papular rash), and subsequently oral azathioprine. Prednisolone dose was gradually reduced and she was discharged from clinic after 1 year. She was re-referred in 2004 after developing joint pains, tiredness and further vasculitic rash. ESR was 81 mm/h and eosinophil count 10.5%. Her symptoms responded to 10 mg of prednisolone, which was reduced and stopped over a period of 4 months. She is currently well and has had no further relapses.

Hepatic vein obstruction (Budd-Chiari syndrome) has been reported in association with idiopathic hypereosinophilic syndrome [1], but this is the first reported case in the context of CSS. This association may have been a coincidence, however thrombosis has been documented in CSS previously [2, 3]. Reported cases have been associated with either normal or raised eosinophil counts at the time of thrombus detection. Ames et al. [2] reported three patients with CSS who developed thrombosis; subclavian vein thrombosis, deep vein thrombosis and middle cerebral artery territory ischaemic stroke. In all three cases, eosinophilia was present at time of thrombosis and the patients all had raised levels of fibrinogen. Garcia et al. [3] reported three cases of venous thromboembolism (deep vein thrombosis and pulmonary embolus). Two of these patients had had normal eosinophil counts at presentation, and the third had a blood count with 19% eosinophils.

Thrombosis has also previously been reported in patients with hypereosinophilic syndrome [1], and in a patient with venulitis [4]. It has been hypothesized that there may be a role for eosinophil granule proteins, such as Major Basic Protein (MBP) in the pathogenesis of thrombosis in conditions in which eosinophilia is a feature [5]. Wang et al. [6] hypothesize that the HOSCN, produced by eosinophils, may lead to a pro-thrombotic and pro-inflammatory state occurring in cases of thrombosis in conditions associated with raised eosinophils counts. These authors found that HOSCN induces tissue factor activity in human umbilical vein endothelial cells and the pro-inflammatory pathway p65/p50 NF-κB [6]. Young et al. [4] report a case of Budd-Chiari syndrome in which liver biopsy revealed multiple non-caseating granulomas, and a necrotizing granulomatous venulitis involving the hepatic vein, and a venogram showed hepatic venous obstruction. No systemic cause for thrombosis was found, and the patient had normal eosinophil count and coagulation. This patient improved with oral prednisolone 40 mg/day. Ames et al. [2] state that increased release of von Willebrand factor (vWF) from endothelial cells in vasculitis may contribute to thrombosis. Although not measured in our patient, raised levels of vWF may have been a contributing factor.

In conclusion, as far as we are aware, this is the first reported case of Budd-Chiari Syndrome presenting in association with CSS. Thrombosis has been previously documented, and the underlying mechanism may be related to raised eosinophil count or underlying vasculitis.

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The rehabilitation model rules in RA until biomedicine transforms tomorrow’s rheumatologist into a real thaumaturgus

SIR, In their leading article, Bertinotti et al. [1] discuss the new assessment tools for clinical trials and individual patient evaluation in rheumatoid arthritis (RA). They observe that the doctors’ and patients’ points of view often do not match, with doctors focussing on measuring disease activity, which is of little interest to their patients who only want to improve their state of health. They discuss the problems associated with trying to unify the efficacy/outcome data with patient’s need and, in particular, his/her quality of life.

The authors then go on to discuss the OMERACT core set, the ACR20 and the Disease Activity Score (DAS), which are made up of both subjective and objective measures. For example, the DAS is comprised of two objective measures (number of swollen joints and ESR or CRP) for the doctors and two objective measures (number of tender joints and patient’s assessment of global health...