Splenic infarction in a patient with Wegener’s granulomatosis

Sir. A 44-yr-old White female presented with a 4 week history of epigastric abdominal pain radiating to the left hypochondrium. She denied other gastrointestinal symptoms. Subsequently, she developed pain in her proximal interphalangeal joints (PIPJ), knees, ankles and left elbow. She complained of red eyes and of nasal stuffiness associated with a bloody discharge. She had lost 1 stone in weight over the 4 weeks. Past history was unremarkable. She smoked 15 cigarettes per day.

On examination, she looked unwell and was pyrexial with a temperature of 37.5°C. She had bilateral painless conjunctivitis, her nasal mucosa was pale and she was tender over the maxillary sinus. Movements in both knees, ankles and right elbow were restricted and were associated with some soft-tissue swelling. Her PIPJ were swollen and tender bilaterally. There was an ulcer on the dorsum of the right foot which had developed over the past 3 days. She had some mild tenderness and fullness over the left hypochondrium, but the abdomen was not distended and there was no friction rub. She was tachycardic with a rate of 140 beats/min, but cardiovascular and also respiratory examination was otherwise unremarkable.

She had a normocytic anaemia with a haemoglobin of 10.1 g/dl. The white count was $13.3 \times 10^9/\text{l}$ (granulocytes 87.6%, lymphocytes 8.5%, monocytes 3.9%). There was no evidence to suggest an eosinophilia. The platelet count was $804 \times 10^9/\text{l}$, an ESR was 109 mm/h and a CRP was 142 mg/l. Urea, electrolytes and creatinine and serum C3 and C4 complement levels were normal. However, a C-ANCA (anti-proteinase 3 antibody) was raised at 44 U (normal range = 0.0–2.0 U). A chest X-ray was unremarkable. A CT scan revealed mucosal thickening within the right maxillary antrum. Echocardiogram and abdominal ultrasound were normal, but
Fig. 1. CT scan of the abdomen obtained following oral and i.v. contrast. There is a hypodense area occupying 80% of the spleen due to infarct.

A diagnosis of splenic infarct associated with Wegener’s granulomatosis was made and she was treated with 1 g of methylprednisolone i.v. for 3 days, 15 mg/kg of cyclophosphamide i.v. as a bolus and 400 mg/kg of Sandoglobulin i.v. for 5 days. Over the next month, she clinically improved and this was reflected in the ESR and CRP. She was discharged with the above regime to be repeated on a monthly basis.

Patients presenting with splenic infarction younger than 40 yr of age often have associated haematological disorders, whilst older patients often have embolic disease. It has been reported in other cases of vasculitis such as polyarteritis nodosa [1] and in association with anti-cardiolipin antibodies [2]. Although clinically apparent splenic infarction is rare in Wegener’s granulomatosis, autopsy reports have suggested that splenic involvement is common [3, 4]. It often presents as a sudden onset of pain in the left upper abdomen, associated with a pain on respiration or, as in our case, local pain on palpation. There may be a friction rub. Haematological abnormalities include an anaemia, leucocytosis and a thrombocytosis, due to loss of splenic function.

Complications, especially abscess and haemorrhage, occur in ~7–20% of patients. Enhanced CT is the most sensitive and reliable cross-sectional technique for diagnosis and classically reveals wedge-shaped, peripheral hypodense lesions [5]. The diagnosis is important as other causes of left upper quadrant pain, e.g. splenic rupture or abscess, perforation of a hollow viscus or ruptured aortic aneurysm, need surgical intervention. In view of this, splenic infarction should be investigated as a cause in a patient with Wegener’s granulomatosis presenting with abdominal pain, especially if associated with abnormal haematology.

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