Return of the spleens

Introduction

This article reports an exceptional cause of multiple intra-abdominal masses that linked an unusual form of interstitial lung disease with a tendency to recurrent respiratory infection.

Discussion

The presence of hypogammaglobulinaemia and splenomegaly (albeit in multiple and unusual anatomical sites), combined with a characteristic form of unusual interstitial lung disease (Bierry et al, 2009), allowed a firm diagnosis of common variable immunodeficiency. This is one of the commonest causes of acquired immunodeficiency, characterized by poor antibody production and an inability to mount a specific antibody response. There is frequently lymphadenopathy as a result of disordered and hyperplastic follicle formation.

Despite the history of splenectomy the absence of Howell–Jolly bodies, target cells and acanthocytes on the blood film suggested normal splenic function and suggested that the intra-abdominal masses might be enlarged splenic remnants. Heat-damaged red cell scanning works on the principle that heat-damaged erythrocytes are less deformable than untreated cells and transit only slowly through the splenic sinusoid (mean transit time 10–15 minutes compared to 40 seconds) where they are subsequently entrapped by erythrophagocytosis some hours later. As only splenic tissue takes up red cells in this way it renders this scan more specific than colloid scans for small nests of functioning tissue (Peters, 2006). Splenosis (the implantation of ruptured splenic fragments onto vascularized surfaces) was confirmed. These splenic fragments can function normally if of sufficient volume (approximately 20–30 cm^3) and render the host immunologically competent.

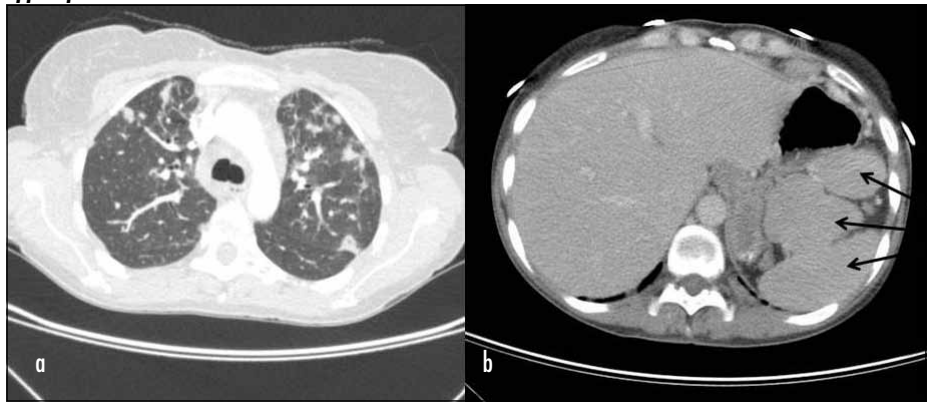
Splenomegaly, particularly in the presence of lymphoid hyperplasia elsewhere, is common in cases of common variable immunodeficiency (Oskenhendler et al, 2008) and is thought to cause the multiple splenic fragments to enlarge. Typically there is follicular hyperplasia of the white pulp, prominent germinal centres and sometimes granulomatous inflammation. Splenosis is commonly misdiagnosed as malignancy and the presence of hypogammaglobulinaemia with multiple intra-abdominal masses raised the possibility of lymphoma.

This case is particularly instructive, for not only did a simple blood film examination lead to suspicion of functioning splenic

Dr Umesh Bangalorelakshminarayana *is Registrar in the Medical Chest Unit, Castle Hill Hospital, Cottingham, East Yorkshire,* **Dr Russell Patmore** *is Consultant Haematologist in the Department of Haematology and* **Dr Philip Dore** *is Consultant Immunologist in the Department of Immunology, Hull Royal Infirmary, Hull,* **Dr Radhakrishnan Jayan** *is Consultant Radiologist in the Department of Nuclear Medicine, Royal Liverpool University Hospital, Liverpool, and* **Dr Michael Greenstone** *is Consultant Physician in the Medical Chest Unit, Castle Hill Hospital, Cottingham, East Yorkshire HU16 5JQ*

Correspondence to: Dr M Greenstone (greenstone.mike@googlemail.com)

Figure 1. a. High resolution computed tomography scan of thorax showing bilateral diffuse upper lobe abnormalities including peripheral nodules, peribronchial consolidation and ground glass opacification. b. Computed tomography scan of abdomen showing multiple intra-abdominal masses (arrows) in the left upper quadrant.



Case Report

A 42-year-old woman presented with a 7-year history of recurrent chest infections and a previous history of splenectomy for traumatic rupture at the age of 21 years.

The initial investigation when she was 35 years of age showed normal immunoglobulins, subclasses and antibody responses to Pneumococcus and tetanus. Examination and spirometry were normal. High resolution computed tomography of the thorax showed nodules and ground glass opacification (*Figure 1a*). There was generalized lymphadenopathy and multiple intra-abdominal masses thought to be nodes (*Figure 1b*). She was found to have low values of IgA 0.28 g/litre (0.8–2.8 g/litre) and IgG2 (0.48 g/litre (1.2–2.6 g/litre), and raised IgM 2.5 g/litre (0.5–1.9 g/litre) (normal ranges in brackets). Beta2 microglobulin was 4.5 mg/litre (1.2–2.5 mg/litre). Inguinal node biopsy showed reactive changes and a bone marrow examination showed normal cellularity, architecture and tricellular haemopoiesis. Thoracoscopic lung biopsy showed nodular lymphoid hyperplasia with minor lymphoid interstitial pneumonia.

The blood film showed a surprising absence of hyposplenic changes. The operative notes confirmed removal of the ruptured spleen but re-implantation of several small segments. A heat-damaged red cell scan showed focal accumulation in the left upper quadrant, left iliac fossa and left side of the pelvis (*Figure 2*) corresponding to the soft tissue densities on single photon emission computed tomography/computed tomography and consistent with splenic tissue.

Immunological review indicated common variable immunodeficiency on the basis of hypogammaglobulinaemia with exclusion of secondary causes. Weekly subcutaneous immunoglobulin and a gluten-free diet for newly diagnosed coeliac disease led to some improvement.

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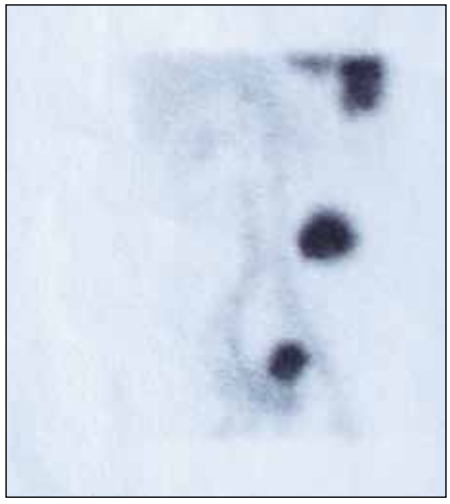
Figure 2. Technetium-labelled heated red cell scan showing multiple intra-abdominal areas of uptake indicating splenic tissue.



tissue but heat-damaged red cell scanning unequivocally identified the masses as splenic, thus avoiding a potentially dangerous biopsy or diagnostic laparotomy. **BJHM**

Bierry G, Boileau J, Barnig C et al (2009) Thoracic manifestations of primary humoral immunodeficiency: a comprehensive review. *Radiographics* **29**(7): 1909–20

Oskenhendler E, Gerard L, Fieschi C et al (2008) Infections in 252 patients with common variable immunodeficiency. *Clin Infect Dis* **46**(10): 1547–54

Peters AM (2006) Imaging the spleen. In: Cook G, Maisey M, Britton K, Chengazi V, eds. *Clinical Nuclear Medicine.* Hodder Arnold, London: 685–94

LEARNING POINTS

- Initial investigation for immunodeficiency in the context of recurrent respiratory infections may be misleadingly normal.
- The presence of hypogammaglobulinaemia and multiple abdominal masses is suspicious of lymphoma.
- Despite the history of splenectomy, the absence of characteristic hyposplenic changes on blood film examination was an important pointer to the presence of functioning splenic tissue.
- Radiolabelled heat-damaged red cell scanning is a specific, non-invasive technique for the confirmation of functioning splenic tissue.
- Although bronchial abnormalities are the commonest respiratory manifestation, histological findings of lymphoid nodular hyperplasia and lymphoid interstitial pneumonitis are highly suggestive of common variable immunodeficiency.

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