Systemic sarcoidosis with spleen involvement

A 43 year old woman had suffered from abdominal pain at the left upper quadrant for two months. She had no fever or weight loss. On physical examination the patient had tender splenomegaly. Haematological and biochemical data showed leukopenia (2.9 x 10^6/l) and she had normal liver function tests and raised serum levels of the angiotensin converting enzyme (132 U/l, normal value 18–55 U/l). On ultrasound study of the abdomen multiple low echogenicity nodules in the spleen were found. Computed tomography of the abdomen revealed an enlarged spleen with multiple small low density areas in the spleen that enhanced after contrast administration, as well as retroperitoneal adenopathy. Computed tomography of the chest showed multiple bilateral lung nodules (less than 1 cm) and mediastinal and hilar lymphadenopathy. A fibrobronchoscopic examination with transbronchial biopsy was non-diagnostic and the patient refused mediastinoscopy. Since splenic enlargement caused pressure symptoms (abdominal pain) and hypersplenism (leukopenia), and also in order to make a definite diagnosis and exclude the possibilities of malignant lymphoma or metastatic malignant tumour, splenectomy was performed. Macroscopically the spleen had a brownish appearance and elastic consistence with many nodules surfacing on the viscera. The size and weight of the spleen were 17.5 x 14 x 9 cm and 672 g, respectively. On sectional view it was found many whitish tumorous nodules filling the splenic parenchyma (fig 1). On histological examination, light microscopic appearance of the spleen was characterised by non-caseating granulomas which consisted of highly differentiated mononuclear phagocytes (epithelioid cells and giant cells) and lymphocytes (fig 2) compatible with sarcoidosis. Special stains for acid-fast bacilli and fungi were negative.

The final diagnosis was systemic sarcoidosis with spleen involvement (nodular pattern).

J L RODRÍGUEZ-GARCÍA
J PICAZO
C MIRA
F BALLESTA
J C GARCÍA-NIETO
E PRIETO
Servicio de Medicina Interna, Hospital General La Mancha Centro, Avenida de la Constitución 3, 13600-Alcázar de San Juan, Ciudad Real, Spain
Correspondence to: Dr José Luis Rodríguez-García
J.L.Rodríguez-García:jlgarcia@serconet.com
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J L RODRÍGUEZ-GARCÍA, J PICAZO, C MIRA, F BALLESTA, J C GARCÍA-NIETO and E PRIETO

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