Primary splenic sarcoidosis

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Here we report a case of primary splenic sarcoidosis presenting with isolated splenomegaly with multiple splenic nodules. The sarcoidosis was diagnosed and treated by splenectomy.

Sarcoidosis is a systemic granulomatosis disease of unknown origin, characterized by the presence of non-caseating granulomatous lesions (1). The primary site of involvement is the pulmonary system in more than 90% of cases (2). Extrapulmonary involvement usually occurs in the presence of lung involvement; isolated extrapulmonary manifestations of sarcoidosis are rare, occurring in only 10% of cases (3). Here we report the case of a 50-year-old woman who presented with a significant weight loss and isolated splenomegaly with multiple splenic nodules and was diagnosed and treated by splenectomy.

CASE PRESENTATION

A 50-year-old South Indian housewife presented with a 6-month history of loss of appetite and weight loss, associated with upper abdominal discomfort and early satiety. Her past surgical history included vaginal hysterectomy for uterine fibroids 6 years previously. Examination revealed splenomegaly, palpable 2 cm from the left costal margin. No other masses were palpable.

She had no history of any leprosy contact, and no rashes were found on physical examination. Her respiratory system, cardiovascular system, and neurological systems were normal. Baseline investigations were within normal range, including a complete blood count; serum calcium level; tests for tuberculosis, thyrotoxicosis, and immunodeficiency; renal function, liver function, blood sugar, and electrolyte testing; bone marrow aspiration; and a tumor marker profile with CA125, CA19-9, and carcinoembryonic antigen (CEA). Her erythrocyte sedimentation rate was 32 mm at 1 hour. Preliminary chest imaging showed a normal radiograph. Ultrasonography of the abdomen showed splenomegaly with multiple hypoechoic lesions in the spleen. A computed tomography (CT) scan of the abdomen and pelvis with contrast showed multiple hypodense lesions in the spleen (*Figure 1a*). The CT scan of the thorax was normal.

A diagnostic splenectomy was performed. The spleen was grossly enlarged, weighing 210 g, with multiple nodules

Figure 1. (a) CT scan of the abdomen and pelvis with contrast showing multiple hypodense lesions in the spleen. (b) Gross specimen after splenectomy showing a grossly enlarged spleen with multiple nodules throughout its surface and inside.

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Figure 2. Histopathology showing (a) splenic parenchyma with a discrete, round to oval noncaseating granuloma composed of epithelioid histiocytes and multinucleated giant cells, and (b) an asteroid inclusion body.

throughout its surface (Figure 1b). Histopathological examination showed splenic parenchyma with discrete, round to oval noncaseating granuloma composed of epithelioid histiocytes, multinucleated giant cells (Figure 2a), and asteroid inclusion bodies (Figure 2b). Reticulin stains showed an extensive reticulin network.

Postoperatively, the patient didn't receive any form of systemic treatment. She has regained her appetite and weight and remained asymptomatic 3 years after diagnosis.

DISCUSSION

In India, the exact prevalence of sarcoidosis is not known. It has been variously estimated as 61 and 150 cases per 100,000 outpatients in Delhi and Kolkata hospitals, respectively. The frequency of splenic involvement in sarcoidosis has been reported to be 10% to 50%, depending on whether it is detected on physical examination (5% to 14%), a radiological test (33% to 53%), or a tissue biopsy (24% to 59%) (4). Splenic involvement is usually asymptomatic, although left upper quadrant pain is occasionally present. Constitutional symptoms such as night sweats, fever, and malaise may occur. Massive splenomegaly is found in 3% of patients with splenic involvement. Splenic sarcoidosis may cause hypersplenism resulting in anemia, leukopenia, thrombocytopenia, or any combination including pancytopenia. Radiologically, splenic sarcoidosis manifests as multiple nodular lesions throughout the splenic parenchyma (5).

Isolated involvement of the spleen in sarcoidosis is extremely rare. Very few cases have been reported in the literature (6). Most patients with splenic sarcoidosis do not require any treatment. In our case, we performed a splenectomy because we had a strong clinical suspicion of lymphoma. The indications for splenectomy include symptomatic abdominal pain from splenomegaly, hypersplenism, functional asplenia, splenic rupture, or a strong suspicion of an alternative diagnosis.

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