

CHAPTER 11

The Spleen in Sarcoidosis

SPLENIC ENLARGEMENT

The true incidence of splenic enlargement in sarcoidosis is not known. Asymptomatic, mild enlargement of the spleen is common and requires no treatment. However, massive splenomegaly and associated hematological and immunological complications require therapeutic intervention.

The normal human spleen weighs approximately 150 g to 250 g. It becomes palpable beyond the costal margin if it has doubled in size. Massive splenomegaly is

defined as a splenic weight of greater than 1000 g or four to six times the normal weight.

Common symptoms of splenic infiltration occur in only 2% of patients with sarcoidosis and are usually confined to those with massive splenomegaly.^{1,3,5,6} These symptoms include fever, weight loss, early satiety, left upper quadrant fullness, and ache and severe pain because of splenic infarct secondary to gastric compression. Functional impairments in patients with splenic infiltration include anaemia, leucopenia, thrombocytopenia, and splenic rupture.

TABLE 11.1 Disorders Associated with Splenomegaly⁴⁻⁹

Enlargement due to increased splenic function

Reticuloendothelial system hyperplasia (RES)

- Anemias
- Early sickle cell anaemia
- Nutritional anemia
- Spherocytosis
- Thalassemia

Infections

- Viral
- Bacterial
- Fungal
- Parasitic

Immune system disorders

- Immune hemolytic anemias
- Immune thrombocytopenias
- Immune neutropenia
- Collagen vascular diseases
- Drug reactions
- Sarcoidosis
- Thyrotoxicosis (benign lymphoid hypertrophy)

Extramedullary hematopoiesis

- Myelofibrosis
- Gaucher's disease
- Marrow damages: toxins, radiation
- Marrow infiltrations: tumors, leukemias

Enlargement due to abnormal blood flow (splenic or portal)

- Cirrhosis
- Congestive heart failure
- Hepatic parasitosis (schistosomiasis, echinococcosis)
- Hepatic vein obstruction
- Portal hypertension
- Portal vein obstruction

Enlargement due to infiltrations of the spleen

- Amyloidosis
- Hyperlipidemias
- Gaucher's disease
- Hodgkin's disease
- Histiocytosis- X
- Hemangiomas
- Hamartomas
- Leukemias
- Lymphomas
- Niemann–Pick disease
- Splenic cysts

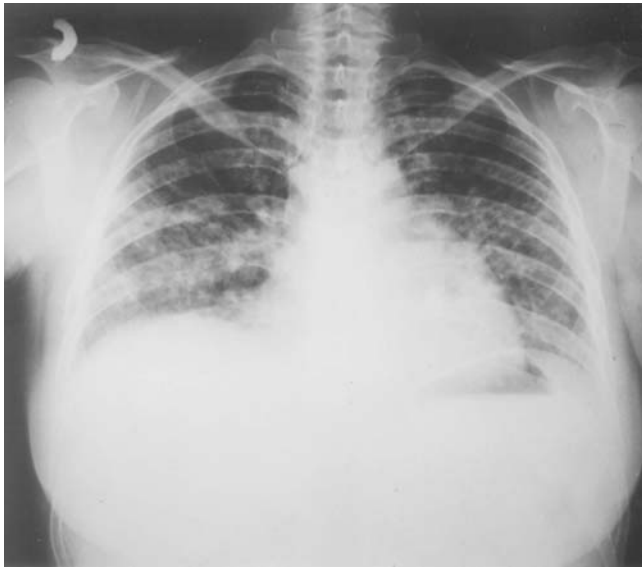


FIGURE 11.1 Chest X-ray representing the Ro stage II of lung sarcoidosis. At that time (1995), the patient had no history of abdominal symptoms and responded to corticosteroid therapy.



FIGURE 11.2 Seven years later (2002), the same female patient shown in Figure 11.1 presented with abdominal pain, left upper quadrant fullness, and ache. The chest X-ray shows the hilar lymph node enlargement with parenchymal reticular lesions.

FIGURE 11.3 Computed tomography (CT) representing multiple splenic lesions.

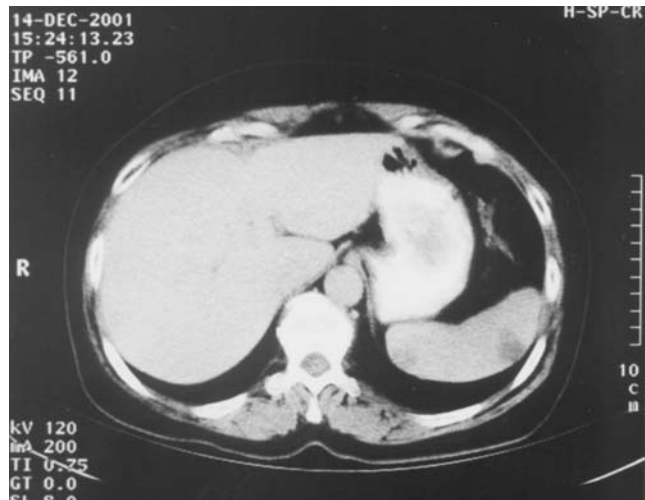
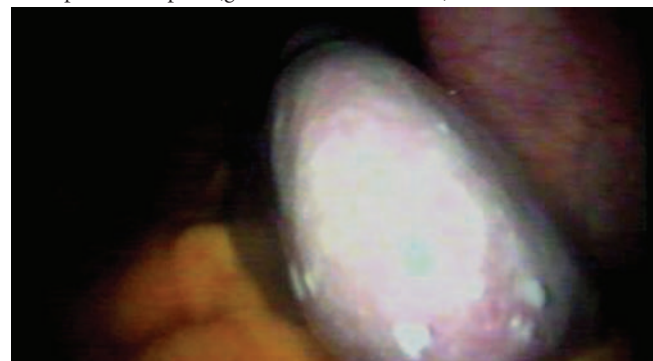


FIGURE 11.4 An ultrasound examination of an extremely enlarged spleen. (Approximately 24 cm–28 cm) It was easily palpable beyond the left costal margin. The patient has persistent hypersplenism and thrombocytopenia because of sarcoidosis.

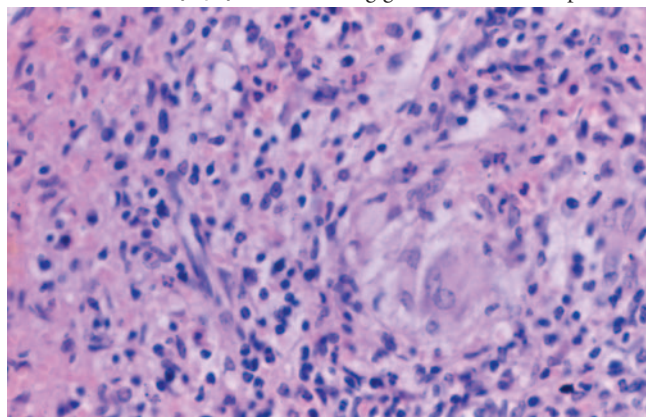
FIGURE 11.5 Endoscopic evaluation of an enlarged spleen with multiple white spots (granuloma formations).



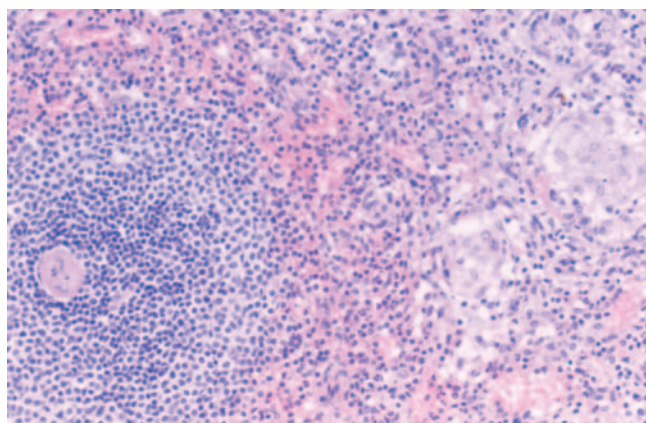
INDICATIONS FOR SPLENECTOMY

Indications for the splenectomy include the following¹: massive splenomegaly not responding to corticosteroids or other drugs, severe hypersplenism with anemia, leucopenia and thrombocytopenia, exclusion of lymphoma or a hematological malignancy, and as a precaution against spontaneous rupture of a massively enlarged spleen.

FIGURE 11.6(A,B) Noncaseating granuloma of the spleen.



A



B

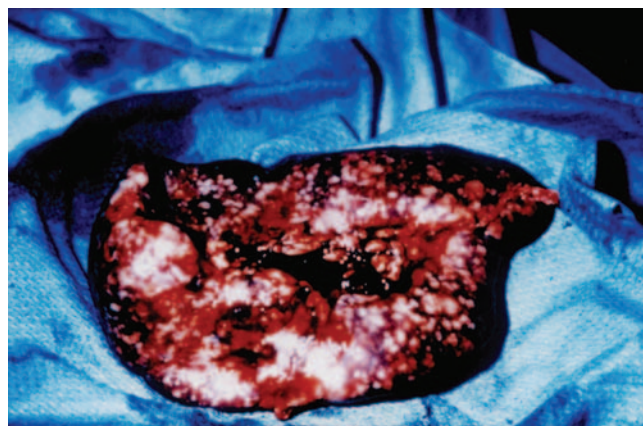


FIGURE 11.7 Splenic enlargement. This spleen is from a patient who had persistent hypersplenism. A splenectomy was advised because the patient was not able to continue prednisone as a result of side effects.¹

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