

Laparoscopic Splenectomy for Isolated Splenic Sarcoidosis

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ABSTRACT

Introduction: Sarcoidosis is an inflammatory disease with an unknown etiology. The pulmonary interstitium is mainly involved, with noncaseating granulomas and lymphadenopathy. It is a multisystemic disease, and the differential diagnosis should include infectious, neoplastic, and autoimmune diseases to prevent inappropriate treatment and unnecessary surgery. Abdominal disease without evidence of pulmonary abnormalities on chest radiography in sarcoidosis can be found in approximately 25% to 38% of cases.

The approach to isolated splenic nodules in a patient with nonspecific abdominal symptoms should be focused on exclusion of malignancies and infections, and may require computed tomography, magnetic resonance imaging, and positron emission tomography–computed tomography imaging; scintigraphy; bone marrow biopsy; breast and genital examinations; and endoscopies.

This report documents a rare case of isolated granulomatous disease of the spleen that was diagnosed and treated laparoscopically.

Case: A 29-year-old woman presented with nonspecific complaints such as nausea, vomiting, and epigastric discomfort. Further laboratory test results were normal. Abdominal ultrasonography, computed tomography, and magnetic resonance imaging revealed multiple splenic lesions. Additional examination findings were negative for occult neoplasia or infectious disease.

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Laparoscopic splenectomy was performed as a diagnostic procedure, without complications, and the final diagnosis was sarcoidosis.

Conclusion: Isolated splenic sarcoidosis is a rare manifestation of extrapulmonary disease. The final diagnosis may be achieved only by histology, requiring biopsy or splenectomy. Minimally invasive surgery is a safe and efficient method for diseases of the spleen and should be the first option when feasible. The patient did well; however, further monitoring is required to diagnose recurrence.

Key Words: Splenectomy, Laparoscopic surgery, Sarcoidosis, Splenic neoplasms, Positron emission tomography.

INTRODUCTION

Sarcoidosis is a multisystemic granulomatous disease characterized by the presence of a chronic inflammatory process of unknown etiology. The main finding is the presence of noncaseating epithelioid granulomas and alteration of normal tissue architecture of the affected organ. The disease occurs worldwide and most commonly affects young and black adults. The positive familial history is well recognized and suggests a hereditary component of the disease.¹ Sarcoidosis usually affects the lungs (90% of cases), skin, and lymphatic system. Manifestations are pulmonary hypertension, renal disease, lachrymal gland involvement, hepatopulmonary syndrome, thrombocytopenia, and cardiac involvement. The disease entails a significant impact on patients' quality of life, and 80% have chronic dyspnea. There is also an association with depression and decreased physical capacity in patients with extrapulmonary sarcoidosis.²

The etiology remains unclear for various reasons, including the heterogeneity of the manifestations of the disease, the lack of a precise definition with overlapping clinical disorders, and nonspecific nonsensitive diagnostic tests. Some studies suggest that it is caused by an unknown antigenic factor or factors determining an exaggerated cellular immune response in genetically

susceptible individuals.^{3,4} The most important diagnostic criterion is histology.⁵

The gastrointestinal tract is seldom the primary site for sarcoidosis. Symptoms may be present and are variable depending on the site of disease. Nevertheless, most patients are often asymptomatic. Hepatic and splenic sarcoidosis are common findings from biopsy and have been reported in up to 80% of cases,⁶ but often they do not manifest clinically. The disease is frequently diagnosed incidentally after investigation of abnormal liver tests. If the patient is symptomatic, ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI) are the most common diagnostic methods. Systemic symptoms may be associated with splenic involvement. Although the chest radiograph may be altered, a normal chest radiograph is seen in a quarter to one-third of patients with splenic sarcoidosis.

The imaging appearance of sarcoidosis can be similar with splenic neoplasms or infectious diseases. Biopsy may be indicated for definitive diagnosis. Needle biopsy can be associated with bleeding and tract seeding, and has variable results; therefore, laparoscopic splenectomy has become the standard surgical approach.

Conversely, patients with solid splenic lesions are uncommon, and the possible causes are lymphoma, metastasis, angiosarcoma, hemangiomas, and inflammatory pseudotumor. Metastatic disease usually derives from lung, breast, ovary, and melanoma cancers.⁷

This report documents a case of primary splenic sarcoidosis that was diagnosed and treated using laparoscopic splenectomy.

CASE REPORT

A 29-year-old woman presented with nonspecific complaints such as nausea, vomiting, and epigastric discomfort. She denied weight loss or other gastrointestinal symptoms, fever, and night sweats. Her past medical history and family history were negative, and there were no findings on physical examination. Laboratory test results showed no abnormalities. Investigation proceeded with abdominal ultrasonography, revealing a normal-sized spleen with heterogeneous parenchyma, with small hypoechoic nodules of different dimensions—most with a 1.0-cm diameter—of undetermined nature. Computed tomography (**Figures 1 and 2**) and MRI (**Figure 3**) of the abdomen confirmed the multiple lesions in the spleen (nodules with low signal on T1 and high signal on T2, with homogeneous enhancement, the largest measuring

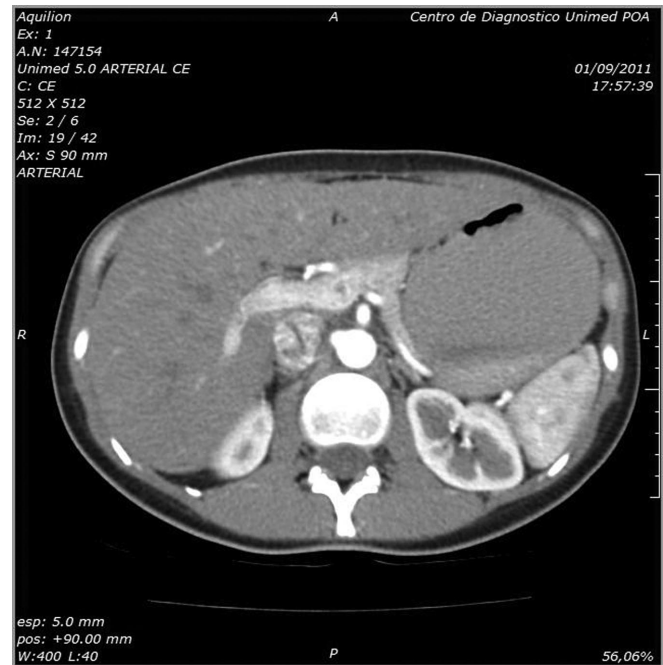


Figure 1. Splenic nodules—CT, arterial phase.

1.0 cm). There was no fluorodeoxyglucose uptake on positron emission tomography (PET)-CT. Findings of extensive occult neoplasm and infectious disease work-up were negative.

The patient thus underwent diagnostic laparoscopic splenectomy. A 5-trocar technique was used. The ligaments were divided using harmonic scalpel, and the vessels were dissected and clipped, and after full mobilization of the viscus, a stapler was used in the splenic hilum. There was no significant bleeding, and operating time was 120 minutes. The spleen was removed with a small enlargement (3 cm) in the lateral incision and morcellation. Pathological examination showed sarcoidosis. The patient recovered uneventfully and was discharged on the third postoperative day, with no complaints, adequate oral intake, and normal vital signs.

DISCUSSION

Sarcoidosis is an inflammatory disease characterized by the presence of noncaseating granulomas. Its diagnosis is based on clinical and radiological findings associated with the histology of epithelioid granulomas, although granulomas are not a specific finding of sarcoidosis, and other diseases should be excluded. These include infections caused by bacteria or fungi, carcinomas, and environmental agents.

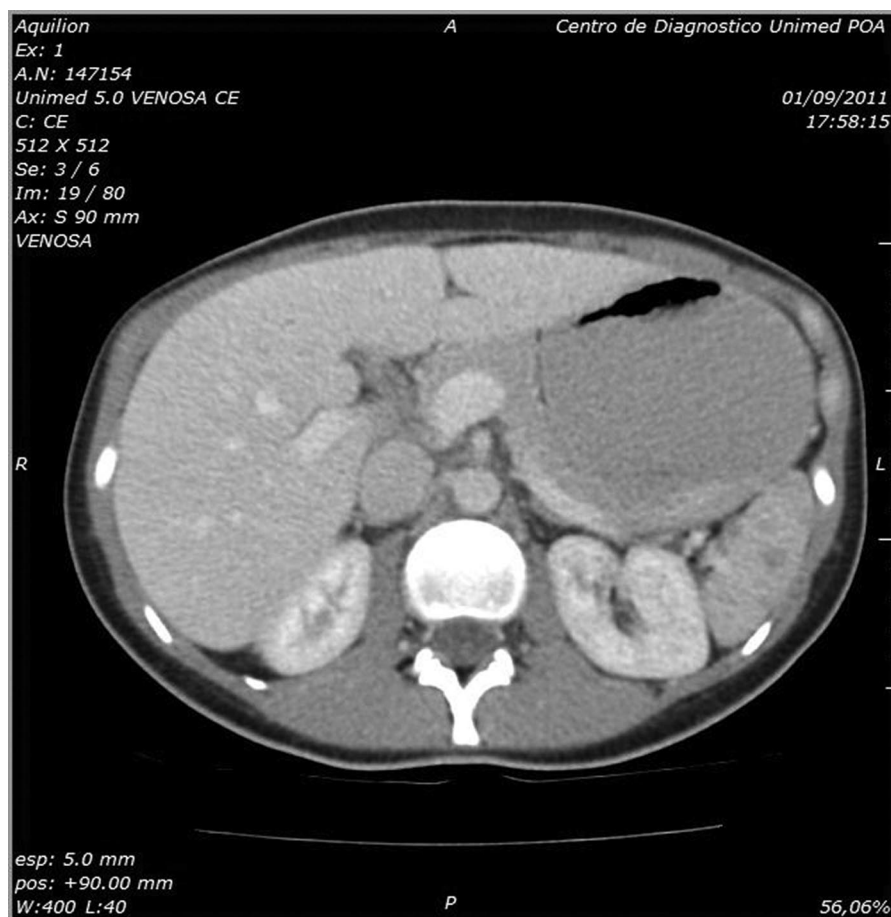


Figure 2. Splenic nodules—CT, venous phase.

Splenic sarcoidosis in the absence of clinical or radiographic pulmonary disease is extremely rare and usually does not cause symptoms. Abdominal pain and systemic symptoms such as fever, malaise, and weight loss occur in some patients. Laboratory tests are not usually helpful. Radiological findings of isolated splenic lesions are non-specific, and the differential work-up includes lymphoma, metastasis, hemangioma, hematoma, abscess, hamartoma, and angiosarcoma.⁸ Histopathological diagnosis is needed for definitive diagnosis. Correlation between pulmonary abnormalities and hepatosplenic involvement has not yet been established.⁵

The incidence of splenomegaly associated with sarcoidosis is variable, and may be as high as 40%.⁹ Imaging methods play a role in the diagnosis and treatment strategy in patients with sarcoidosis. Computed tomography, abdominal ultrasonography, MRI, and PET-CT can easily detect splenic injuries.^{10–14} PET-CT seems to be a very useful imaging method in the evaluation of

disease activity and identification of undetected areas, as well as in monitoring response to treatment in patients with sarcoidosis. PET-CT seems to be superior to scintigraphy in patients with sarcoidosis and has higher sensitivity (PET-CT provides high image quality and resolution compared with scintigraphy), lower radiation exposure, and less time between injection and imaging. Therefore, in patients with pulmonary sarcoidosis and consistent imaging of the spleen, no further testing is required.

Regarding the treatment, asymptomatic patients do not require the use of immunosuppressive drugs. When symptoms do occur, steroids or even methotrexate and azathioprine can be used. As many as 66% of patients spontaneously remit.²⁴ The lack of response or the appearance of complications may impose splenectomy.²⁰

In this case, abdominal ultrasonography revealed small hypoechoic nodules of different size, which was con-



Figure 3. Splenic nodules—MRI, T2 phase.

firmed by CT and MRI. PET-CT yielded no diagnostic advantage. Laparoscopic splenectomy has been used extensively as a tool for diagnosis and treatment for the diseases of the spleen and definition of malignancy.¹⁵ Thus, laparoscopic splenectomy was indicated for diagnostic purposes. It is a minimally invasive approach that provides shorter hospitalization, better aesthetics, decreased blood loss, and decreased postoperative complications. The benefit of rapid recovery allows the early start of chemotherapy. Splenectomy does not alter the course of disease but may be needed for treatment of complications,¹⁶ and indications include symptoms, hypersplenism, prophylaxis for splenic rupture, and neoplastic exclusion.⁵ Laparoscopic splenectomy has gradually become the gold standard for surgical removal of the spleen.

This case report highlights the diagnostic approach of splenic lesions and the value of laparoscopic splenectomy as an effective and safe procedure for diagnosis and treatment. Our case was of an extremely rare form of sarcoidosis. The isolated splenic disease is a precursor for systemic disease. Therefore, patients in this setting should be closely followed by their physicians for relapse, and immunosuppressive therapy is indicated when there is involvement of major organs (neurological, ophthalmological, or cardiac) or evidence of organ dysfunction or progressive disease in other or-

gans.³ The current data do not support routine PET-CT use to differentiate benign and malignant conditions.¹⁷

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