

# Splenic Abscess in a Patient with Wegener's Granulomatosis Treated with Laparoscopic Splenectomy

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## Abstract

Splenic abscess complicating Wegener's granulomatosis (WG) has not been previously described. We report the occurrence of a splenic abscess in a 45-year-old white male suffering from WG. The patient presented with persistent fever and abdominal pain. Magnetic resonance imaging showed two splenic cystic lesions. Differential diagnosis was splenic hematoma or abscess. The patient underwent diagnostic laparoscopy and laparoscopic splenectomy. Pathology revealed a centrally located cavity full of pus and necrotic material. Although there were no signs of active vasculitis, all other histological features were compatible with WG. The patient had an uneventful postoperative course and his disease is in remission. Laparoscopic splenectomy appears to be a safe procedure, but its impact on the management of splenic abscess needs to be determined further.

**Key Words:** Wegener's granulomatosis, splenic abscess, laparoscopic splenectomy.

## Introduction

WEGENER'S GRANULOMATOSIS (WG) is a disease characterized by necrotizing granulomatous vasculitis of small and medium-sized arteries (1). Most frequently it involves the upper and lower respiratory tract and the kidneys and has a wide range of manifestations with a broad spectrum of severity (1). Survival in WG increased dramatically after the introduction of cyclophosphamide and glucocorticoids (median survival >21 years) but often with considerable disease- and treatment-related morbidity (2, 3).

Spleen involvement in WG is not unusual but is rarely diagnosed during life and is probably underreported (4, 5). Spleen pathology in affected patients includes infarction and necrosis with central arteritis, hemorrhage, rupture, capsule adhesions, splenomegaly and function impairment (4–7). There has been no previous report of a splenic abscess in WG.

We present a patient with Wegener's granulomatosis who underwent laparoscopic splenectomy for a splenic abscess.

## Case Report

A 36-year-old white male was admitted in 1996 because of hemoptysis. Thoracic computed tomography (CT) revealed a round cavity with thick walls in the middle lobe of the right lung. There was no lymphadenopathy, and tests for specific infections (tuberculosis, parasitosis) were negative. An open lung biopsy showed granulomatous necrotic inflammation consistent with Wegener's granulomatosis. The patient was placed on methotrexate and folic acid for three years. In 1997 he had an episode of lower gastrointestinal bleeding due to an angiodysplasia of the cecum. A contrast-enhanced CT scan of the abdomen showed a hypodense multilobular area in segment V (Couinaud classification) consistent with a granuloma from WG.

In March 2005 the patient gradually developed discomfort and persistent pain of the left upper quadrant, which worsened at deep inhalation. Subsequently, he developed pain in his proximal interphalangeal joints, knees and ankles and had lost 4 kg in weight over three weeks. His temperature

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was constantly between 37.5 and 37.8°C. On physical examination he appeared ill, with a temperature of 38°C, a pulse of 110 beats per minute and a blood pressure of 115/75 mm Hg. He had severe tenderness over the left upper quadrant and splenomegaly. The hemoglobin was 9.7 g/dL, the white cell count 14,500/mm<sup>3</sup> and the platelet count 212,000/mm<sup>3</sup>; the prothrombin and partial-thromboplastin times were normal. The erythrocyte sedimentation rate (ESR) level was 95 mm/hour and the C-reactive protein (CRP) level was 102 mg/L. Levels of C<sub>3</sub>, C<sub>4</sub>, p-antineutrophil cytoplasm autoantibodies (p-ANCA), c-ANCA were normal; rheumatoid arthritis (Ra) and antinuclear antibody (ANA)-tests were also normal. The lupus anticoagulant and anticardiolipin antibodies were negative. Chest X-ray was unremarkable. Chest CT scan revealed the known lesion in the middle lobe of the right lung. A contrast enhanced CT and a magnetic resonance imaging (MRI) scan of the abdomen (Fig. 1) showed two non-enhancing splenic cysts (6.5 cm and 4.2 cm) and a focal lesion in segment IV consistent with the known granuloma. A transesophageal echocardiogram failed to detect cardiac sources of emboli.

The differential diagnosis was splenic hematoma or abscess associated with WG. Administration of methylprednisolone (500 mg IV per day for three days) and cyclophosphamide (2 mg/kg per day, orally) was begun. Methylprednisolone was continued at 48 mg per day orally and trimethoprim and sulfamethoxazole were started after three days.

Over the next two months the patient had constant low-grade fever with minimal improvement. Abdominal imaging showed that the splenic lesions were unchanged. A diagnostic laparoscopy was performed.



**Fig. 1.** Abdominal MRI scan.

## Operative Findings

The spleen was enlarged and densely adherent to the lateral abdominal wall, the diaphragm and the stomach. Two cystic lesions were identified at the splenic hilum. There was no ascites, gross lymphadenopathy or other intra-abdominal pathology. A laparoscopic splenectomy was performed. Biopsies were taken from the lesion in the right lobe of the liver.

The patient had an uneventful postoperative course and was discharged on the fourth postoperative day. Six months later, he was asymptomatic with normal levels of ESR and CRP, and his disease was in remission.

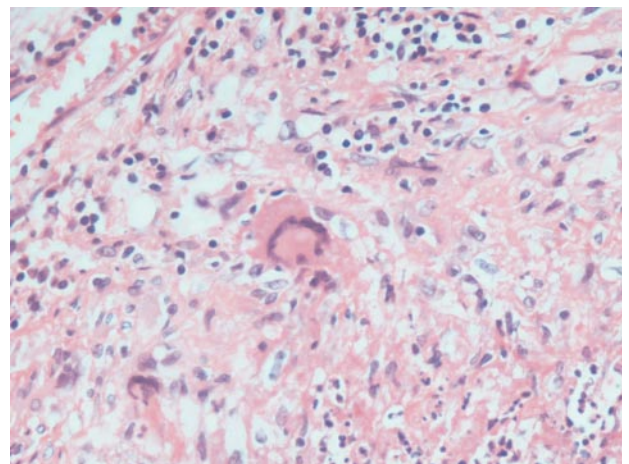
## Histopathology

### Gross Findings

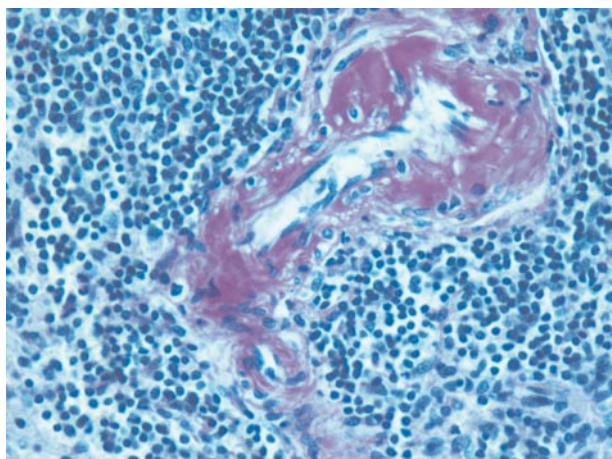
The patient's spleen measured 10×11.4×4 cm with a weight of 260 g. In sectioning, there was a central cavity measuring 4 cm containing pus and necrotic material.

### Microscopic Findings

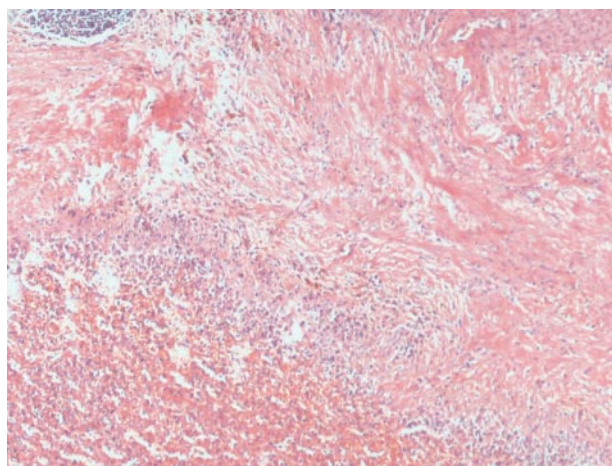
Foci of granulomatous inflammation and thrombosed vessels were found peripherally to the necrotic area (Figs. 2 and 3). Small arteries throughout the spleen had thickened walls and lumen stenosis (Fig. 4). Some small arteries had eosinophilic degeneration and chronic inflammation. Thrombosed vessels and small foci of ischemic necrosis were found distally to the necrotic cavity. Histochemistry did not reveal any microorganisms.



**Fig. 2.** Giant cells at the periphery of the necrotic area in the splenic parenchyma (H&E × 200).



**Fig. 3.** Regressive vascular necrosis: PAS + material on the vascular wall (PAS  $\times$  200).



**Fig. 4.** Palisading histiocytic inflammation around foci of splenic necrosis (H&E  $\times$  100).

### Discussion

Manifestations of splenic involvement in WG include splenomegaly, splenic vasculitis, rupture, infarction, necrosis and impaired splenic function (4–10). Splenic abscesses are a recognized complication of Felty's syndrome, rheumatoid arthritis, systemic lupus erythematosus and polyarteritis nodosa (7), but there have been no previous reports describing the occurrence of a splenic abscess in WG.

Splenic abscesses occur predominantly as a result of hematogenous spread from a septic focus (usually endocarditis) or secondary to immunosuppression (11, 12). No apparent etiology (e.g., infectious, hematologic, neoplastic) for a splenic abscess was found in the described patient. The pathogenesis remains obscure, but there are reports of aseptic splenic abscesses in other diseases asso-

ciated with circulating ANCA, such as inflammatory bowel diseases (13, 14).

Although there were no signs of active vasculitis in the pathology examination, all other histologic features (palisading granuloma around foci of necrosis, small vessel involvement) (15) were compatible with WG. It should be noted that the fundamental disease mechanism in WG is connective tissue "necrobiosis" (severe involvement of collagen and other mesenchymal tissues) and not vasculitis (15).

Diagnosis of a splenic abscess can be difficult, because the symptoms are nonspecific and can vary widely. Fever, left upper quadrant pain, nausea/ vomiting and splenomegaly with leucocytosis are the most common signs and symptoms associated with a splenic abscess (12, 16). Contrast enhanced CT scan and MRI are the most sensitive imaging techniques (12). This patient had the classic symptoms (with radiological imaging) suggestive of a splenic abscess.

Minimally invasive surgery is now the procedure of choice for elective splenectomy. Significant splenomegaly (length  $>$  22 cm or weight  $>$  700 g) is considered a relative contraindication to laparoscopic resection due to the difficulty in manipulation and the potential for significant hemorrhage (17). Hand-assisted laparoscopic splenectomy can be used in such cases, as it offers the advantages of minimally invasive surgery while allowing safer resection (17).

Splenectomy is the treatment of choice for splenic abscess (11, 12). Splenic resection is the most effective and definitive procedure for the majority of patients, although percutaneous drainage is very helpful in carefully selected cases (18). Carbonell et al. showed that laparoscopic splenectomy is a safe and effective method for treating splenic abscesses (16), as in the case of this patient. Although the number of patients in their study was small, there was no morbidity or mortality and they had shorter hospital stays compared with the open splenectomy cohort (16). The impact of minimally invasive surgery on the management of splenic abscess needs to be investigated further.

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