

# Splenectomy For Splenomegaly And Hypersplenism Manifestations Compatible With Possible Lymphoma: Diagnosis: Sarcoidosis

H Yanarda?, Y Güne?

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## Abstract

Sarcoidosis is a systemic granulomatous disorder of unknown etiology, most frequently involving pulmonary system, with or without multisystemic organ involvement. Isolated extrapulmonary involvement is unusual. Splenomegaly and secondary hypersplenism may be associated with acute and chronic infections, autoimmune states, portal hypertension or splenic vein thrombosis, and a number of infiltrative and neoplastic conditions involving the spleen. Splenomegaly has been reported in 1-40 percent of patients with sarcoidosis, but this involvement is usually self limited, rarely necessitating splenectomy (1). Clinical presentation is variable and can range from being asymptomatic to severe constitutional symptoms. Especially, the differential diagnosis of lymphoma could present a problem since both diseases could present with fever, splenomegaly and enlarged paraaortic lymph nodes (2).

We present here two cases in which splenectomy was performed for diagnostic purposes, especially with the suspicion of lymphoma, but later on found to be sarcoidosis.

## CASE 1

54 year-old female patient referred to our clinic with the complaints of non-productive coughing, hoarseness of voice and painful erythematous lesions on her legs. She had pancytopenia and splenomegaly and underwent explorative laparotomy and splenectomy for possible lymphoma last year. Physical examination revealed multiple tender erythematous rashes differing in size on lower extremities bilaterally. After consultation with dermatology department lesions concluded to be consistent with erythema nodosum. Hemogram and routine biochemical laboratory exams, including protein and immunoglobulin electrophoresis, were normal. Last year, the patient had liver and bone marrow biopsies also, but no specific diagnosis could be established. But blood count returned to normal and patient became asymptomatic after splenectomy. Mediastinal and right paratracheal lymphadenomegalies (LAMs) of milimetric in size, but no hilar pathology, were found on thorax computerized tomography (CT) during that time. After detecting bilateral hilar LAM on chest-ray and in the presence of erythema nodosum we suspected of sarcoidosis. ACE level was 115 U/L (8-52), CD4/CD8 rate from

bronchoalveolar lavage was 1.8. thorax CT showed that paratracheal LAMs became pathologic in size. Biopsy taken from paratracheal lymph nodes was consistent with sarcoidosis. Reevaluation of splenectomy specimen was inconsistent with sarcoidosis again. Biopsy taken from larynx due to hoarseness, also didn't show any specific finding. Echocardiography and fundoscopy were also normal. With the diagnosis of sarcoidosis steroid treatment instituted and continued for one year. Hilar LAM, erythema nodosum, hoarseness and complaints of patient all regressed totally. She is still attending our department.

## CASE 2

56-year old female patient was evaluated with the complaints of left lumbar pain, weight loss and low grade fever. She had anemia and splenomegaly. Biopsies from left axillary LAM, liver and bone marrow didn't lead any specific diagnosis. She was splenectomized for diagnostic purposes and with the suspicion of tuberculous adenitis antituberculous therapy was started. 6 months later bilateral hilar LAM and interstitial pulmonary infiltration were detected on chest-X ray and thorax CT. Then she was referred to our department. White blood cell count was 12,000 /mm<sup>3</sup> other routine laboratory findings were normal. Spirometric examination showed mild obstruction (FEV<sub>1</sub>/FVC= 76 %). ACE level was 72.2 U/L (8-52). Acid-

fast bacilli were not detected with Ziehl-Nielsen staining. Bronchoalveolar lavage CD4/CD8 rate was 2.02. Transbronchial endoscopic biopsy taken from nodular granulomatous lesions demonstrated to have non-necrotizing granulomatous inflammation and also findings related with non-specific bronchitis. Splenectomy material was reevaluated by an experienced pathologist and reported to have non-caseating granulomatous inflammation and dystrophic calcifications. After antibiotherapy and bronchodilator treatment respiratory functions returned to normal. Again steroid treatment was started and continued for one year. All clinical and radiological findings regressed totally. She is also still under our follow-up.

## **DISCUSSION**

Sarcoidosis is a granulomatosis disease of unknown origin with a variable clinical presentation. The reported frequency of splenomegaly in sarcoidosis ranges from 1% to 40%. Autopsy results on series of patients with generalized sarcoidosis suggests that the spleen is the second most common site of involvement. The prevalence of splenomegaly was 63 % in autopsy findings by Longcope and Freiman (3).

Compared with patients without splenomegaly, patients with splenomegaly had evidence of more extensive extrathoracic sarcoidosis. Whereas degree of pulmonary involvement is similar in patients with or without splenomegaly. And there is no difference in longterm follow-up of patients with or without splenic enlargement since the degree of pulmonary involvement is the determining factor of morbidity and mortality. (4) But there're reports that patients with splenomegaly had evidence of extensive pulmonary and extrathoracic sarcoidosis with a poor prognosis in spite of steroid therapy (5). Although individual case reports have documented the occurrence of hematologic abnormalities in patients with splenic involvement, there are no adequate studies that indicate the frequency with which manifestations of hypersplenism occur in such patients. Although the natural history of sarcoidosis is usually unchanged after splenectomy, symptoms like abdominal pain and hematological abnormalities can resolve completely (2, 4, 6).

The patients with splenomegaly have an increased incidence of abdominal pain and constitutional symptoms, articular symptoms, hepatomegaly, and peripheral lymphadenopathy. Splenic rupture may occur rarely. (1, 4, 5)

The lungs are radiographically involved in more than 90 % of patients with sarcoidosis. Only 5-10 percent of patients present with a normal chest roentgenogram. Neither chest roentgenograms nor pulmonary function tests are diagnostically sensitive or specific. Bronchoscopy with transbronchial biopsy is a high-yield procedure and obtains characteristic non-caseating granuloma in 90 % to 100 % of patients, in some series, including patients with normal chest x-ray (4, 7).

## **CONCLUSIONS**

In summary, sarcoidosis should be considered in differential diagnosis of splenomegaly even in the absence of overt pulmonary involvement.

## **CORRESPONDENCE TO**

YANARDAG Halil, MD. Department of Internal Medicine, Cerrahpasa Faculty of Medicine, University of Istanbul, TR-34303, Cerrahpasa, Istanbul, TURKEY Phone and Fax no : +90 212 5862928 E-mail : yilmazleman@e-kolay.net

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**Author Information**

**Halil Yanarda?**

Cerrahpasa Faculty of Medicine, Internal Medicine Respiratory Diseases Department, Istanbul University

**Yilmaz Güne?**

Institute of Cardiology, Istanbul University