

CASE REPORT OF EXTRA PULMONARY SARCOIDOSIS PRESENTING AS SPLENOMEGALY IN ABSENCE OF PULMONARY INVOLVEMENT

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ABSTRACT

Sarcoidosis is a granulomatous disease with multisystem involvement mostly pulmonary. Extrapulmonary involvement without pulmonary involvement is very rare. We report a case of extra pulmonary sarcoidosis presenting as splenomegaly in absence of pulmonary involvement. 40 year old male presenting with fever and splenomegaly and axillary lymphadenopathy. Chest roentgenogram and CECT chest were normal. Serum ACE levels were high. Lymph node biopsy is suggestive of sarcoidosis. Patient responded to steroids

KEYWORDS: Sarcoidosis is multisystem disorder characterised by granulomatous inflammation.

INTRODUCTION

Sarcoidosis is multisystem disorder characterised by granulomatous inflammation in any part of body usually in more than one areas.^[1]

In most of cases sarcoidosis is first identified on chest roentgenogram of asymptomatic patients.^[2] The incidence of splenomegaly in sarcoidosis ranges from 11-30%.^[2] However the incidence of large splenomegaly extending 4 cm or more below left costal margin is 4.7% in Kataria and Whitcomb's series.^[2]

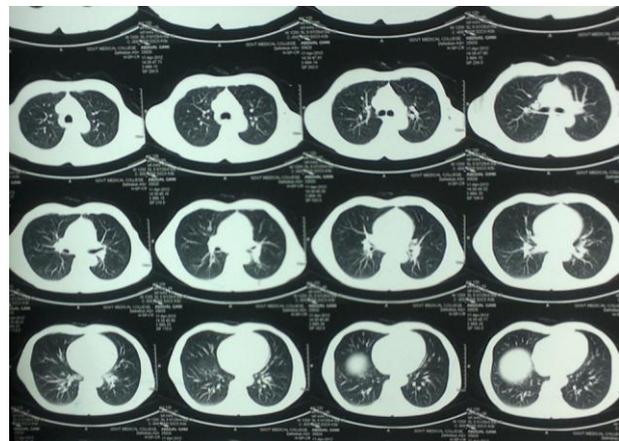
Patients with splenomegaly have high incidence of constitutional symptoms and tend to have disseminated disease.^[3]

CASE REPORT

40 year old male working as carpenter presented with chief complaint of fever for 7 weeks and fullness in left upper quadrant. Patient was in his usual state of health when in evening after returning from work started with fever recorded 100-101^oF not relieved by local medication associated with evening rise, chills and rigours, myalgias and malaise. It was associated with pain and fullness in left upper quadrant. Patient received injectable ceftriaxone 1 gram bid for 7 days.

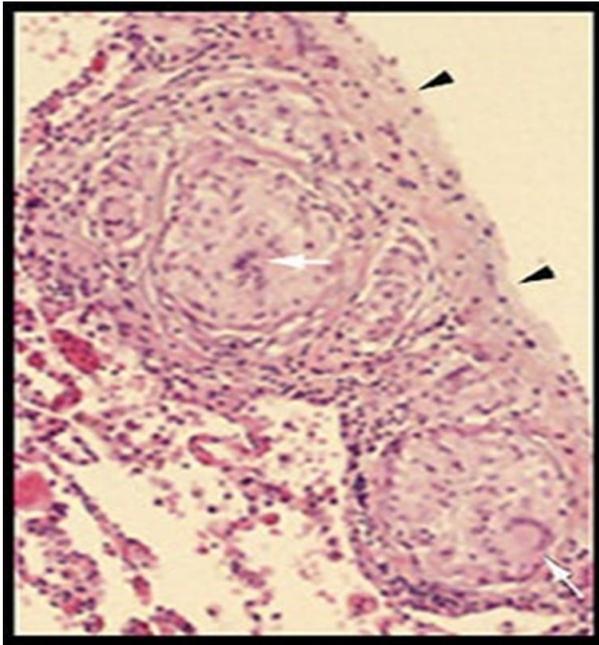
On examination patient was conscious cooperative oriented to time place and person, febrile temp 101^oF, pulse 110 beats regular synchronous good volume, blood pressure 110/70, respiratory rate 20 breaths per

minute. Bilateral axillary lymphadenopathy (medial group) was present.



CECT Chest and Abdomen

• B/L lung fields normal No lung parenchymal lesion seen No mediastinal LAP



Lymph node excision biopsy granulomatous inflammation with giant cell

Chest cardiovascular and central nervous system examination was normal.

On abdominal examination splenomegaly was present 9cm below left costal margin. CBC shows Hb 12.4 TLC 4600, DLC N₆₅ % L₂₁ %, Platelets 2.15 lacs ESR 49mm/hr. Serum Urea 42 mg/dl serum creatinine 1.0 mg/dl, serum sodium 134 meq /dl potassium 3.8meq/dl calcium 11.08mg/dl, glucose 112mg/dl. serum Bilirubin 0.82mg/dl S. proteins 7.76mg/dl, Albumin 3.5mg/dl SGOT 13IU/dl SGPT 12 IU/dl ALP 72 IU/dl CRP 11gms/dl LDH 216 IU/dl ANA Negative EGD Shows early oesophageal varices. CXR PA view normal, ECG normal, USG moderate splenomegaly

Blood and urine culture sterile

Widal titers; T (O) 1; 80 T (H) 1:80 Brucella titers; non-reactive Montoux; non-reactive sputum for AFB 2 samples negative. Echo normal. Bone marrow examination shows mild hypo plastic marrow with few prominent.

Serum electrophoresis shows hypergammaglobulinemia, no definite m bands seen. CECT chest and abdomen shows bilateral normal lung field and parenchyma and no lymphadenopathy. Spleen is enlarged showing homogenous contrast enhancement. Hepatobiliary system is normal.

Lymph node excision biopsy: shows lymph node parenchyma replaced by well circumscribed cluster of epithelioid histiocytes with occasional large Langerhans giant cells, no caseating necrosis seen suggestive of sarcoidosis ACE levels; 140 IU/dl (8-65) high 24 hr. urinary calcium: 292 mg /dl (100-300)

TSH: 2.4 IU

Patient was managed with tab prednisolone 40 mg od, patient improved symptomatically, fever subsided within 14 days, patient is on oral steroids and regular follow up with regression of splenomegaly

DISCUSSION

Giant splenomegaly in other reports [2-4] was always associated with disseminated disease and constitutional symptoms. This patient was having symptoms of fever from 7 months and left sided fullness due to splenomegaly. The absence of chest involvement as suggested by lack of pulmonary symptoms, normal chest roentgenogram and CT scan chest is quite unusual in presence of splenomegaly.

Kataria and whitcomb, s² noted hypersplenism in 6 of 11 patients with splenic size >4cm below left costal margin. The elevated levels of ACE confirm the sensitivity of this test as a marker of disease activity⁵. In this patient excisional lymph node biopsy is confirmatory of sarcoidosis. The resolution of constitutional symptoms, fever and splenomegaly, after prednisolone therapy is surprising since Kataria noted diminution of spleen size after 1 year of corticosteroid therapy in patients with splenic size >4cm. This case further illustrates the heterogeneity of sarcoidosis, a granulomatous disease of unknown aetiology.

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