

Case Report

Sarcoidosis with Splenic Involvement: Two Case Reports from a Single Centre from Northern Sri Lanka

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Abstract: Splenic sarcoidosis is a rare clinical entity which can have heterogeneous clinical presentation. We highlight two different clinical encounters of splenic sarcoidosis with coexistent pulmonary sarcoidosis, which needs high degree of clinical suspicion to diagnose in complexed burnt out state and importance of timely diagnosis and early initiation of corticosteroids to minimize disease progression. Case 1; 58 year old previously healthy female presented with marked constitutional symptoms and left side upper abdominal discomfort over 6 months period with unremarkable clinical examination on admission. Her chest X ray on admission simulated the appearance of lymphangitic carcinomatosis, however ultrasound guided splenic tru-cut biopsy confirmed the diagnosis of splenic sarcoidosis and patient responded to corticosteroid therapy. Case 2; 21 year old young male was evaluated for pyrexia of unknown origin for three months. His clinical examination was only significant for bilateral cervical discrete lymph node enlargement and chest X ray on admission showed bilateral hilar enlargement and initial ultrasound abdomen revealed multiple hypoechoic splenic lesions. Finally bronchoscopy guided lung biopsy confirmed the diagnosis and patient was remarkably improved with corticosteroid treatment. Splenic sarcoidosis has heterogeneous clinical presentation. Judicial use of imaging, appropriate tissue biopsy together with correct clinical judgment will enable early diagnosis and early treatment with corticosteroid will have favorable patient outcome.

Keywords: Splenic Sarcoidosis, Pyrexia of Unknown Origin.

INTRODUCTION

Sarcoidosis is a rare multisystem disorder which is often under diagnosed in Sri Lankan setting and renders high degree of clinical suspicion particularly to diagnose in complexed burnt out stage with multisystem involvement. The most common organ system involved is the lung which accounts for 90 percent of presentations. The involvement of spleen in systemic sarcoidosis is a rare clinical entity and there are only few case reports published in international literature with hepatosplenic involvement. However data on systemic sarcoidosis with coexisting pulmonary and splenic involvement, isolated splenic involvement and hepatosplenic sarcoidosis is lacking in local literature. Here we report two patients with systemic sarcoidosis with coexisting splenic and pulmonary involvement that presented to the university Medical Unit of Teaching Hospital Jaffna.

CASE 1

58 year old female presented with significant loss of appetite and weight over 6 months duration. Detailed systemic inquiry did not revealed any positive history apart from recent onset vague left hypochondrial

discomfort. Her physical examination was unremarkable apart from small goitre. Her baseline blood investigations were well within normal limits except a marginally raised alkaline phosphatase level (135U/L). Her chest X-ray on admission showed bilateral diffuse reticulonodular shadowing simulating lymphangitis carcinomatosis. Ultrasound abdomen and breasts was arranged in view of excluding primary malignancy which revealed multiple hypoechoic splenic lesions with a spleen size of 9 cm. Contrast enhanced CT scan of the chest was interpreted as multiple lung metastases with enlarged right paratracheal and subcarinal lymph nodes (Figure 1).

In view of marked constitutional symptoms on admission the possibility of mycobacterial infection was reasonably ruled out by three negative sputum samples for Acid Fast Bacilli (AFB) and negative tuberculin skin test. Contrast enhanced CT of abdomen further reconfirmed hypodense splenic lesions and otherwise indeterminate. A bronchoscopy guided lung biopsy was done, however histology did not revealed any significant findings. In the absence of a potentially diagnostic clue it was decided to go ahead with image

guided splenic biopsy with extreme precautions to avoid bleeding. Histology of the biopsy revealed presence of well formed non- necrotising epithelioid granulomas with splenic tissue showing a slight excess of eosinophils. The PAS and Ziehl- Neelsen stain to exclude fungal and acid- fast bacilli were negative. Based on history, favourable radiological and histological findings a diagnosis of splenic sarcoidosis was made. Subsequently an elevated ACE level of 197.62U/L (8-52U/L) strengthened the provisional diagnosis. She was treated with oral prednisolone 1 mg/kg daily and continued for a month and tailed off over 3 months period and continued with 5 mg per day maintenance dose. Follow up chest radiograph taken at 3 months after commencing treatment showed remarkable improvement.



Fig-1: HRCT thorax showing bilateral nodular lesions mimicking multiple lung metastasis

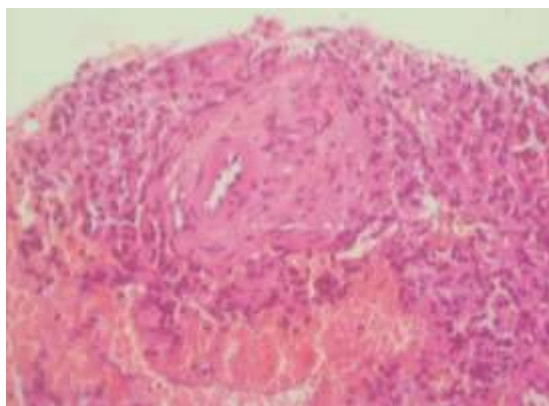


Fig-2: H&E (40x40) section of USS guided splenic tru-cut biopsy showing a non- necrotizing epithelioid granuloma

CASE 2

22 year old male was evaluated at the University Medical Unit, Teaching Hospital Jaffna for pyrexia of unknown origin for three months. His physical examination was positive for bilateral posterior triangle discrete cervical lymph node enlargement, otherwise the system examination did not reveal any significant findings. On admission his blood investigations revealed high ESR of 118mm/1st hour (Normal range <15 mm/1st hour) and CRP of 73.3mg/dl

(Normal range 0.0-0.8mg/dl) Chest X ray showed bilateral hilar lymphadenopathy. Mycobacterial infection was reasonably ruled out by three negative sputum AFB and negative skin tuberculin test. Ultrasound abdomen was performed to rule out intra abdominal lymphadenopathy a characteristic feature of lymphoma however it revealed enlarged spleen (12 cm) with multiple hypoechoic areas. High resolution CT scan of thorax revealed enlarged mediastinal, paratracheal and subcarinal lymph nodes. Histological examination on bronchoscopy guided biopsy revealed non caseating granulomatous inflammation. Considering the history, radiological findings and biopsy findings systemic sarcoidosis was entertained as unifying diagnosis and serum ACE level which was done as a supportive evidence was found to be 51 U/L (Normal range 8-52 U/L) patient was treated with oral prednisolone 1 mg/Kg daily. At two weeks review he was clinically doing well with remission of radiological findings. It was decided to continue same dose prednisone for another six months.

DISCUSSION

Sarcoidosis is a multi system disorder, histologically characterized by non caseating granulomatous inflammation. Though 90% of clinical presentations are featured by lung parenchymal involvement, splenic sarcoidosis is a clinically rare entity even though splenic involvement is reported in 40% to 60% of patients in literature.

Most commonly splenic sarcoidosis is clinically silent, however they can have wide range of presentations from local pressure symptoms to marked constitutional symptoms as reported in case 1 and even it can present as massive splenic infarctions [1]. Imaging plays a key role in early identification of splenic sarcoidosis. In a poor resource setting ultrasonography and contrast enhanced CT abdomen are reasonable choice of investigations sensitive enough to detect splenic involvement though MRI allows more comprehensive assessment.

Sarcoidosis is a close mimic of tuberculosis infection in terms of clinical presentation; preliminary haematological findings; biochemical investigations and radiological features. Hence it creates great degree of diagnostic dilemma particularly in tropics where Mycobacterium tuberculosis infection is highly endemic. In contrast, the characteristic histological finding of non caseating granulomatous inflammation provides solid evidence for the diagnosis of sarcoidosis along with a negative tuberculin skin test. This approach is simple but reasonable enough to exclude tuberculosis infection in tropical settings [2]. Further the elevated angiotensin converting enzyme (ACE) level and presence of subtle characteristic radiological differentiating features will help to further strengthen the clinical diagnosis.

As mentioned in case 2, splenic sarcoidosis can present with pyrexia of unknown origin. In such instances diagnosis of splenic sarcoidosis is often dismissed in tropics because of high prevalence of infective aetiologies that present with PUO with splenomegaly such as typhoid, typhus, infectious mononucleosis, chronic malaria, leishmaniasis, schistosomiasis, infective endocarditis etc. [3].

Corticosteroid still stands the mainstay of treatment for sarcoidosis. In most instances pulmonary sarcoidosis is being managed by pulmonologists based on collaborative guidelines laid down by American Thoracic Society (ATS) European Respiratory Society (ERS) and World Association of Sarcoidosis and Other Granulomatous Disorders (WASOG) and British Thoracic Society guidelines. Both guidelines recommend corticosteroids for initial management of progressive symptomatic pulmonary involvement. ATS/BRS/WASOG guideline recommend 20mg – 40mg of oral prednisolone as the initial dose to be continued for 1-3 months before weaning and thereafter a maintenance dose to be continued for at least one year. However BTS guideline recommends more longer treatment duration of 6 to 24 month depending on clinical improvement [4]. Further, in the absence of favorable response to steroids or in the presence steroid induced adverse effects, introduction of steroid sparing immunosuppressive agents is of proven benefit according to current literature [4]. Though there are robust data on management of pulmonary sarcoidosis, evidence on treatment strategies for splenic sarcoidosis is scarce. However apart from corticosteroid therapy surgical intervention becomes a suitable option when splenomegaly in sarcoidosis is complicated by hypersplenism [5].

In conclusion splenic sarcoidosis is a rare clinical entity and it is a challenging diagnosis in the presence of heterogeneous aetiologies for tropical splenomegaly. However judicious use of available investigations to arrive at an early diagnosis and early initiation of prompt corticosteroid therapy will minimize disease progression and ensure good patient outcome.

DECLARATIONS

Ethical approval

Ethical clearance for the publication for the two case reports was obtained from the Ethical Review Committee, Faculty of Medicine, Teaching Hospital, Jaffna, Sri Lanka.

Consent for Publication

Informed written consent for publication of the personal clinical details and the images was obtained from the two patients prior to submission of this article.

Availability of data and materials

All clinical data and images used in this case report is available with the corresponding author on reasonable request

Competing Interests

The authors declare that they have no competing interests.

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Authors' contributions

DDD was involved in writing the manuscript, TK, GS and JAP involved in writing as well as editing the initial manuscript and all authors read and approved the final manuscript.

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