Case Report

Pulmonary Sarcoidosis with Extra-pulmonary Involvement of Supra-clavicular Lymphnode

Biswas M A[1], Haque M S[2], Shawon M H H[3], Podder T[4]

Abstract

This is a case report of pulmonary sarcoidosis as bilateral hilar lymphadenopathy with extrapulmonary involvement of right supraclavicular lymph node. Patient was 32 years old, male, ex-smoker, government service holder hailing from Faridpur and presented with fever, cough, shortness of breath & supraclaviclar lymphadenopathy. Chest x-ray P/A view showed bilateral hilar lymphadenopathy with pulmonary infiltrations. Serum ACE level was high. Lymph node biopsy is suggestive of sarcoidosis. Patient responded to steroid.

Keywords: Sarcoidosis, Bilateral hilar lymphadenopathy, Pulmonary infiltration, Lymph node biopsy, Serum ACE level.

Introduction

Sarcoidosis is a granulomatous disease with multisystem involvement mostly pulmonary. Extrapulmonary involvements are quite common.[1] Sarcoidosis is a chronic inflammatory granulomatous disease characterised by granuloma without caseum, which more frequently affects the lung, although multi-organ involvement is common. Changes in chest imaging are described at some stage of the disease in 90% of patients, more common as bilateral hilar lymphadenopathy and interstitial lung disease. Atypical manifestations, such as pseudotumours, can also occur.[2] The etiology of sarcoidosis is not fully understood; the association between sarcoidosis and tuberculosis remains controversial.[3]

Case Summary

A 32 years old, ex-smoker, male, government job holder patient was brought to hospital with complaints of fever associated with cough and shortness of breath for 3 days. Fever was continuous in nature and which was subsided after proper medication by paracetamol. Cough was productive, mucoid, with foul smell and bloody discharge. Patient also had breathlessness which has no history of previous attack and there was no association with allergic condition, diurnal or seasonal variation.

1. Dr. Muhammad Asaduzzaman Biswas
Associate Professor, Department of Respiratory Medicine
Diabetic Association Medical College, Faridpur.

2. Dr. Mohammad Saiful Haque
Associate Professor, Department of Radiology
Diabetic Association Medical College, Faridpur.

3. Dr. Md. Hashibul Hasan Shawon
Intern Doctor, Department of Medicine
Diabetic Association Medical College, Faridpur.

4. Dr. Tonmoy Podder
Intern Doctor, Department of Medicine
Diabetic Association Medical College, Faridpur.

Correspondence to:
Dr. Muhammad Asaduzzaman Biswas
Associate Professor, Department of Respiratory Medicine
Diabetic Association Medical College, Faridpur.
E-mail: drasaddtcd@gmail.com

On examination**,** Patient was conscious, well alert, oriented without any pallorness and cyanosis. Temperature was 102º F, Pulse Rate 90 beats/minute, which was regular, normal in volume and character. Blood pressure was 130/90 mmHg, respiratory rate 35 breaths/minute, oxygen saturation was 95%, breath sound was vesicular. Right supra-clavicular lymph node was enlarged, non-tender, with regular margin, firm in consistency, mobile and measuring about 2.2 cm x 1.5 cm x 1.0 cm. Sputum for C/S shows Pseudomonus and sensitive to Floroqunolone and 3rd generation cephalosporine. So it treated with sensitive injectable antibiotic and other supporting management. Patient become afebrile and sputum production also stopped but some dry cough was persisting. After one month follow up, number of lymph node involvement is increasing gradually and bilateral cervical lymphadenopathy was persist. Rests of general and systemic examinations were normal.

Meanwhile, the patient was treated symptomatically and undergone several investigations. Routine investigations revealed the following findings:

Name of investigations	Findings
Hb%	13.0 g/dl
Total WBC count	9.48 x 10^3
CRP	<6 mg/dl
Chest X-ray	Bilateral lymphadenopathy without pulmonary infiltration. (Figure 2)
The findings of liver and kidney functions along with echo-cardiography were within normal limits	

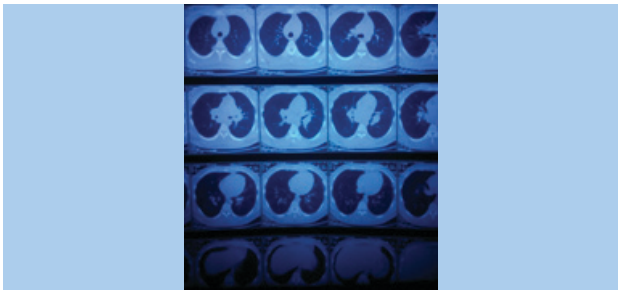

Figure 1: HRCT

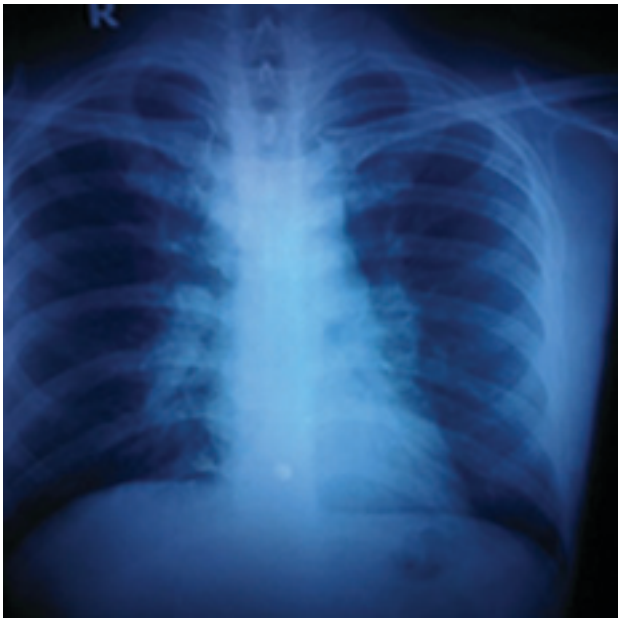

Figure 2: Chest X-Ray P/A view

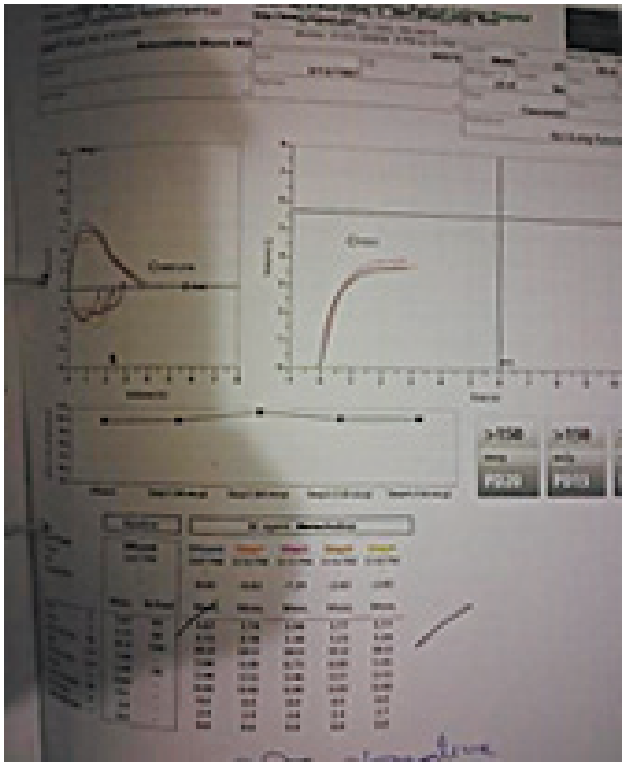

Figure 3: Spirometry

In addition, Sputum for AFB & Gene Xpert for Micobacterium Tuberculosis (MTB) were negative. Biopsy of right supra-clavicular lymph node shows non-caeseating granulomatous lesion suggestive of sarcoidosis. Serum ACE level was high (86 U/L). Spirometry showed FVC 65%, FEV1 68%, FEV1: FVC was 85.9%, which was suggestive of restrictive lung disease (Figure-3). Considering the presentation & investigations, the patient was diagnosed as Pulmonary sarcoidosis (Stage-1) with extrapulmonary involvement of right supra-clavicular lymph node.

Treatment and follow up

Prednisolone (1mg/kg) was started and patient gradually decries the size of lymph nodes and finally disappeared at month 3, but in the meantime iatrogenic Cushing's syndrome has been developed, presenting central obesity, acne, and plethora. So prednisolone was replaced by methylprednisolone (1mg/kg) one more month and Chest X-ray shows hilar lymphadenopathy disappeared. Serum ACE level also declined to 12.4U/L and then we tapered the dose and now patient is clinically good health.

Discussion

Sarcoidosis is a chronic granulomatous disease that affects multiple organs. Pulmonary involvement is frequent but usually not with this presentation. In about 5% cases superficial lymph node involvement is seen. Some authors contend that MTB plays a role in the etiology of sarcoidosis, due to the similar clinical and histopathological outcomes.[4] In recent decades, studies have been published documenting the presence of Micobacterium tuberculosis DNA in patients with sarcoidosis.[5] Similiarly, there are reports of sequential occurrence of sarcoidosis and Tuberculosis.[6,7] Currently, some authors propose that MTB Ag can induce sarcoidosis in genetically predisposed individuals.[8,9] In this case, it is isolated from MTB as sputum and gene Xpert both was negative and biopsy shows non- caeseating granuloma. The goals of sarcoidosis management are to prevent or control organ damage, relieve sign symptoms and improve the patient quality of life. While a significant percentage of sarcoidosis do not need treatment. However prednisolone should be commenced immediately in presence of Pulmonary, Renal impairment and uveitis. In patients with severe disease MTX (10-20mg/wk), Azathioprine (50-150mg/day) and use of specific TNF-α inhibitors have been effective. Hydroxyclouroquine may be useful in cutaneous sarcoid with limited pulmonary involvement. The overall mortality is low (1-5%) and usually reflects cardiac involvement or pulmonary fibrosis.

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