■ *Case Report*

Pulmonary sarcoidosis: a case report with typical imaging features

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Abstract

Sarcoidosis is a granulomatous disorder multisystem involvement; however, pulmonary manifestations typically dominate with abnormal chest radiographs in majority of the patients. Pulmonary function tests like vital capacity and total lung capacity are reduced. The typical radiographic feature of sarcoidosis is bilateral hilar lymphadenopathy with involvement of right paratracheal lymph nodes concomitantly. Computed tomographic scan is ahead of conventional chest radiographs in depicting parenchymal, mediastinal and hilar abnormalities. Transbronchial lung biopsy and transbronchial needle aspiration are important for diagnosing pulmonary sarcoidosis.

Keywords: Pulmonary sarcoidosis, Bilateral hilar lymphadenopathy, Corticosteroids, Transbronchial lung biopsy, Lung transplantation.

Introduction

Sarcoidosis is a multisystemic disorder of unknown etiology. Its characteristic feature is the presence of non-caseating granulomas that can affect various body parts; however, pulmonary involvement is seen in more than 90% of patients[1]. The clinical course and expression of pulmonary sarcoidosis are variable, may be asymptomatic or can present with severe respiratory symptoms and even can cause death[2]. Restrictive pattern of disease is seen in more than 20% of patients at initial diagnosis; however obstructive pattern have also been noted[1,3]. In about two thirds of patients there is spontaneous regression but it may lead to chronicity in about 30% of patients. Chronic form of sarcoidosis may result in progressive loss of pulmonary function and there could be mortality in 1 to 4% of patients.

Case presentation

A 42 years old male presented with two months history of cough, intermittent fever, fatigue, night sweats and weight loss. Cardiovascular system was normal. Total calcium, serum phosphorus and alkaline phosphatase levels were within normal limits. Complete blood count (CBC) was normal except low lymphocytes (13%), Erythrocyte sedimentation rate (ESR) was high (115 mm/hr). Saturation of peripheral oxygen (SPO2) was 97% of room air. Serum ACE inhibitor was 32 U/l (normal). Montoux test was negative.

Chest radiograph shows prominent bronchovascular markings in bilateral lung fields, bilateral hilar soft tissue prominence with air bronchogram possibly due to enlarged lymph nodes and prominent horizontal fissure.

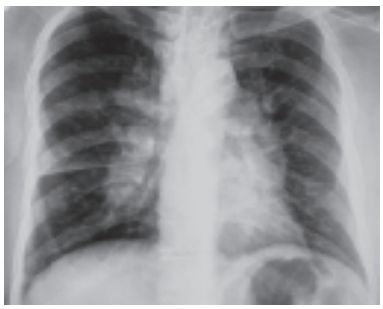

***Fig 1**: Chest radiograph showing prominent bronchovascular markings in bilateral lung fields, bilateral hilar soft tissue prominence possibly due to enlarged lymph nodes and prominent horizontal fissure.*

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Computed tomographic (CT) scan of chest revealed bilateral massive hilar and mediastinal lymphadenopathy and patchy infiltrates in both lower lobes. Ophthalmologic examination was within normal limits. No features s/o healed or active uveitis.

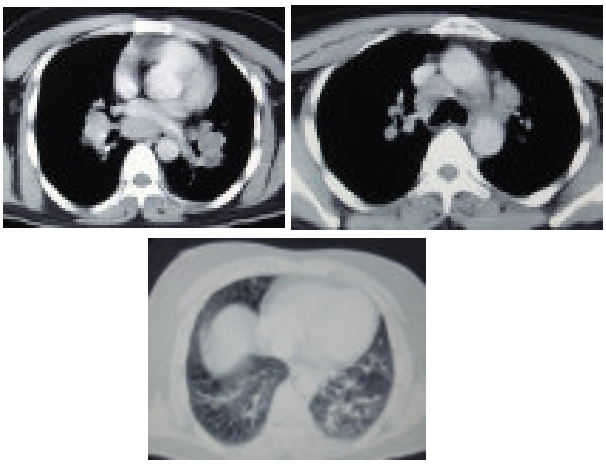

Fig 2a, 2b and 2c: *Axial section CT scan of chest in mediastinal window showing bilateral massive hilar and mediastinal lymphadenopathy (Figure 2a and 2b) and in lung window showing patchy infiltrates in both lower lobes (Figure 2c).*

Bronchoscopy showed splayed secondary carina in right upper lobe with chronic inflammation suggestive of sarcoidosis. Cytopathology of the bronchoalveolar fluid (BAL fluid) was done which revealed macrophages, few respiratory epithelial cells and occasional benign squamous cells in a mucoid background. No acid fast bacteria seen on Ziehl-Neelsen stain. No fungal elements or granuloma or atypical cells seen. Transbronchial needle aspiration (TBNA) shows numerous respiratory columnar epithelial cells, foamy and carbon laden macrophages, and occasional cluster of squamous metaplastic cells and neutrophils in a mucoid background. No granuloma or atypical cells seen.
Patient was advised oral antibiotics and prednisolone along with salbutamol inhaler for 1 month and there was clinical improvement in the symptoms.

Discussion

Sarcoidosis has propensity to involve multiple systems, but pulmonary involvement usually dominates[5]. Skin, eyes and peripheral lymph nodes are each involved in 15 to 30% of patients[5,6]. Involvement of spleen, liver, heart, central nervous system, bone or kidney occurs in 2 to 7% of patients[4].

It presents with cough, dyspnea or bronchial hyperreactivity with significant endobronchial or pulmonary parenchymal involvement[1]. However, 30 to 60% of patients are asymptomatic and diagnosed incidentally on chest radiographs[7].

The typical radiographic feature of sarcoidosis is bilateral hilar lymphadenopathy with involvement of right paratracheal lymph nodes concomitantly[7]. Other groups of mediastinal lymph nodes may be involved and could be detected by computed tomographic (CT) scans. Unilateral hilar lymphadenopathy on CT is seen in about 10% of patients[8]. Pulmonary parenchymal infiltrates (with or without hilar lympadenopathy) are seen in 20 to 50%[1,7]. The pattern of infiltrates may be patchy or diffuse with predisposition for upper and mid lung zones. There could be presence of reticulonodular infiltrates, macroscopic nodules, consolidation or masslike lesions[5].

Computed tomographic scan is ahead of conventional chest radiographs in depicting parenchymal, mediastinal and hilar abnormalities[10]. On CT scan, the typical features of sarcoidosis is mediastinal and/or hilar lymphadenopathy, nodular opacities and micronodules along bronchovascular bundles, predilection for mid and upper lung zones, an axial distribution, pleural or subpleural nodules, septal and nonseptal lines, confluent nodular opacities with consolidation and groundglass opacities11. In advance stage of the disease, pulmonary fibrosis occurs with volume loss, hilar retraction, coarse linear bands, large bullae, cystic radiolucencies, distortion, mycetomas, bronchiectasis and enlarged pulmonary arteries may be observed[2,3,9].

There are four stages of pulmonary sarcoidosis mentioned below:
stage 0: normal
stage I: bilateral hilar lymphadenopathy without pulmonary infiltrates
stage II: bilateral hilar lymphadenopathy plus pulmonary infiltrates
stage III: parenchymal infiltrates without bilateral hilar lymphadenopathy
stage IV: not universally adopted) refers to extensive fibrosis with distortion or bullae

In stage I sarcoidosis, there is abnormality in pulmonary function tests (PFTs) are detected in 20%. Reductions in vital capacity and total lung capacity are characteristic findings. There is increased level of serum angiotensin converting enzyme (ACE) in approximately 30–80% of patients and could be a marker of total granuloma burden[4].

The diagnostic yield of flexible fiberoptic bronchoscopy with transbronchial lung biopsy (TBLB) is about 60–90%, even in radiographic stage I disease[12]. Transbronchial needle aspiration biopsies (TBNA) are diagnostic in 63–90% of patients with mediastinal and/or hilar adenopathy on chest CT[13]. The combination of TBNA and TBLB may have a higher accuracy than either procedure alone[13].
Specific complications of sarcoidosis are necrotising sarcoid angiitis, bronchostenosis, myecetomas, pleural involvement, superior vena cava syndrome and pulmonary embolism.

Corticosteroids are the mainstay treatment option for severe or progressive sarcoidosis (pulmonary or extrapulmonary), and often gives significant regression of the disease[2,14]. Patients not responding or having adverse effects from corticosteroids, then immunosuppressive, cytotoxic or immunomodulatory agents are preferred. In patients with end-stage pulmonary sarcoidosis refractory to medical therapy, lung transplantation is a viable option[15].

Summary

Sarcoidosis is a multisystem disorder with predominantly pulmonary involvement. The diagnosis of pulmonary sarcoidosis is suggested by bilateral hilar lymphadenopathy, with or without parenchymal changes on chest radiographs and is supported by noncaseating granulomata in tissue biopsies. Radiographic staging of pulmonary sarcoidosis as well as clinical and laboratory findings can be prognostic.

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