

## Pneumothorax - a Rare Presenting Sign of Sarcoidosis

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### Abstract

Sarcoidosis is a non-infectious, inflammatory disease of unknown etiology which is characterised histologically by presence of noncaseating granulomas. It is a common pulmonary disease in Indian Subcontinent with tuberculosis as the commonest differential diagnosis. Spontaneous pneumothorax have been described in late stages of sarcoidosis but pneumothorax in early disease or the first presenting sign is very rare. This article discusses a rare case of Sarcoidosis where pneumothorax was the first presenting sign.

**Keywords:** Sarcoidosis; Pneumothorax; Spontaneous; Sign

### Introduction

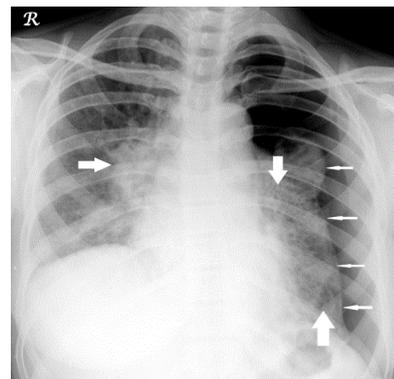
Sarcoidosis is an idiopathic, multisystem, non-infectious, inflammatory disease characterised by histological presence of noncaseating granulomas [1]. Pulmonary manifestations of the disease predominate and are associated with maximum morbidity & mortality [2]. Pneumothorax is an atypical manifestation of sarcoidosis noted in only 2% of cases with few cases reported in literature [3,4]. When encountered, pneumothorax is usually a late presentation of disease [5], rarely it may also be the first manifestation of disease as seen in our index case [6].

### Case Report

A 25-year old female with history of sudden dyspnea at rest presented to the emergency department of our hospital. There was no history of chest pain, fever or asthma. Leading question revealed history of slight progressive dyspnea on performing household work since last few months. There was no occupational or other history suggesting exposure to any allergen.

Clinical examination revealed signs of dyspnea at rest including flaring of nares, deep supraclavicular fossa, inward drawing of intercostal spaces, restricted movement of left hemithorax and predominant abdominal breathing. Auscultation revealed lack of breathing sounds in left hemithoracic region with normal pulses. Percussion revealed hyperresonant note in left hemithorax. Laboratory tests and ECG were all unremarkable.

Chest radiograph in PA projection was advised and it revealed presence of moderate to gross pneumothorax in left hemithorax causing near-complete compression atelectasis of left lung without obvious mediastinal shift; enlargement of right hilar shadow probably secondary to adenopathy and multiple, air-filled, cystic spaces randomly distributed in all lobes of both lungs but more easily identified in compressed left pulmonary parenchyma (Figure 1).



**Figure 1:** Radiograph of chest taken in PA projections shows pneumothorax on left side causing near-complete collapse of left lung (multiple thin arrows); multiple cystic spaces in left lung (thick white arrows) and right hilar enlargement representing adenopathy (arrow in right hemithorax) in patient of pulmonary sarcoidosis.

Based on these findings, sarcoidosis was suggested as the radiological diagnosis. Cystic disease of left lung with secondary pneumothorax and infection was the other differential diagnosis.

Patient was then advised tube thoracostomy to relieve dyspnea and further evaluation with contrast-enhanced computed tomography (CECT) of chest & flexible bronchoscopy for nodal biopsy.

Due to financial constraints but for completion of diagnostic work-up, CECT thorax was deferred and patient was directly taken up for flexible bronchoscopy when nodal biopsy (without US guidance) confirmed presences of noncaseating granulomas while bronchoalveolar fluid revealed raised angiotensin-converting enzyme (ACE) levels, thus confirming sarcoidosis.

Patient was then discharged with ICDT in situ and on a prescription with short-course of oral corticosteroid, bronchodilator & an antibiotic. Patient was then lost to follow-up.

## Discussion

Though sarcoidosis is a multisystem disease yet its pulmonary manifestations predominate with more than 90% patients having abnormal chest radiographs at the time of diagnosis [7]. The most characteristic findings include near-symmetrical, bilateral hilar & superior mediastinal adenopathy and parenchymal infiltrative lesions including reticular, reticulonodular & focal alveolar opacities [3]. However, less than one-quarter present with atypical symptoms including cavities with or without fungal balls, pleural effusions, solitary pulmonary nodules, bullae or pneumothorax [8]. Bullae seen in sarcoidosis have a different etiopathogenesis than localised cystic airspaces that are seen in end-stage disease [9].

Spontaneous pneumothorax in sarcoidosis may be unilateral or bilateral, usually secondary to rupture of bullae or necrosis of granuloma in sub pleural location which may coexist with hemorrhagic or nonhemorrhagic pleural effusion [6,10]. Pneumothorax has usually been described in the medical literature as a late manifestation of sarcoidosis with fibrotic or bullous disease [5]. However, very few cases have been described in the medical literature where pneumothorax is the presenting sign or the first sign of relapse [6,11-13].

Management of pneumothorax has not been conspicuous with some authors suggesting the role of corticosteroids in preventing relapse of pneumothorax [13]. Thoracoscopic bullectomy has been suggested in cases of severe bullous disease with pulmonary function impairment nonresponsive to corticosteroid or pneumothorax unresponsive to tube drainage or frequent relapses [4,11-13].

## Conclusion

To summarize, pneumothorax is a rare presenting sign of sarcoidosis which should be suspected in young patients especially in the absence of history of smoking. Pneumothorax usually represents late manifestation of sarcoidosis.

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