

Spontaneous Pneumothorax Complicating Pulmonary Sarcoidosis: A Case Report.

Abstract:

Sarcoidosis is a multisystem granulomatous disease predominantly affecting the lungs, which may progress to chronic fibrotic forms responsible for respiratory complications. Spontaneous pneumothorax is a rare but potentially serious complication of pulmonary sarcoidosis, occurring most often in advanced stages of the disease.

We report the case of a 57-year-old nonsmoking man with stage IV fibrotic pulmonary sarcoidosis who presented with acute right-sided chest pain and severe respiratory distress. Chest radiography revealed a complete right-sided pneumothorax. Initial chest tube drainage resulted in incomplete lung re-expansion, requiring subsequent surgical management. Thoracic surgery revealed subpleural bullae, which were resected, allowing complete lung re-expansion. The postoperative course was favorable, and the patient remained clinically stable without pneumothorax recurrence during follow-up at 1, 2, and 3 months.

Through this case, we discuss the proposed pathophysiological mechanisms of pneumothorax in sarcoidosis, management approaches, and the risk of recurrence. This complication should be considered in any patient with diffuse pulmonary involvement who develops respiratory distress, particularly in advanced fibrotic forms.

Keywords: Pulmonary sarcoidosis; Spontaneous pneumothorax; Pulmonary fibrosis; Respiratory complications.

Introduction

Sarcoidosis is a multisystem granulomatous disease with a predilection for mediastinal and pulmonary involvement. Diagnosis is based on a combination of clinical, radiological, and histological findings after other granulomatous diseases have been excluded. Corticosteroid therapy is the first-line treatment, while immunosuppressive agents constitute second-line therapy. The course of the disease may be complicated by serious events, although these remain rare.

Case Presentation

A 57-year-old nonsmoking man was admitted to the emergency department with right-sided, pleuritic chest pain that was exacerbated by deep inspiration and coughing, with no history of chest trauma.

The patient had been followed for two years for diffuse interstitial lung disease progressing to fibrosis, with a history of occupational exposure to dust and mold.

A diagnosis of sarcoidosis had been considered and established three months earlier in the setting of exertional dyspnea and dry cough, based on several findings: radiological evidence of an interstitial pattern predominantly involving the mid and upper lung zones, classified as stage IV; histological findings on bronchial biopsy showing epithelioid and giant-cell granulomatous inflammation without caseous necrosis; and bronchoalveolar lavage showing lymphocytosis of 25%.

The patient had been receiving systemic corticosteroid therapy for three months, with marked clinical improvement that was maintained after gradual dose reduction.

Pulmonary function testing demonstrated a mixed obstructive and restrictive ventilatory defect, with a reduced FEV1/FVC ratio of 0.46, forced vital capacity (FVC) of 2.31 L (55% of predicted), forced expiratory volume in one second (FEV1) of 1.07 L (33% of predicted), total lung capacity (TLC) of 4.31 L (64% of predicted), and diffusing capacity for carbon monoxide (DLCO) of 63% of predicted.

On admission, examination revealed tachypnea at 25 breaths/min, oxygen saturation of 75% on room air, and distension of the right hemithorax, with absent tactile vocal fremitus on palpation and reduced breath sounds over the right lung field. The patient was hemodynamically stable, with no signs of acute right heart failure.

Chest radiography (Figure 1) showed a complete right-sided pneumothorax.



Figure 1. Chest right-sided

radiography (Figure 1) revealed a complete pneumothorax.

A chest tube was inserted in the right fourth intercostal space. Follow-up chest radiography showed satisfactory lung re-expansion, with a small residual apical pneumothorax.

Clinical and radiological improvement was slow. After a 14-day hospital stay, the patient was transferred to the thoracic surgery department for surgical management. Surgery consisted of resection of superficial bullae (Figure 2), allowing the lung to re-expand against the chest wall.



Figure 2. Thoroscopic views showing subpleural bullae (red arrows) in the lower lobe (A), interlobar fissure (B), and middle lobe (C).

Immediate postoperative chest radiography (Figure 3) showed complete re-expansion of the right lung, with persistent bilateral interstitial changes related to underlying parenchymal fibrosis. At postoperative follow-up visits at 1, 2, and 3 months, the patient remained clinically stable, with no recurrence of pneumothorax.



Figure 3. Postoperative frontal chest radiograph showing complete lung re-expansion and diffuse fibrotic changes.

Discussion

Spontaneous pneumothorax is a rare but well-described complication of pulmonary sarcoidosis, reported in approximately 1–2% of cases, and occurs most often in chronic fibrotic forms of the disease [1,2]. The present case illustrates this complication in a patient with advanced mediastinopulmonary sarcoidosis (stage IV) and severe impairment of respiratory function.

Several pathophysiological mechanisms have been proposed to explain pneumothorax in sarcoidosis, including rupture of subpleural bullae, necrosis of subpleural granulomas, and weakening of fibrotic lung parenchyma leading to alveolar rupture [2,3]. These mechanisms are comparable to those described in other systemic diseases with diffuse and cystic pulmonary involvement, particularly neurofibromatosis type 1, in which the risk of pneumothorax is also elevated [4]. In such cases, slow recovery after chest

86 tube drainage, as seen in our patient, often reflects poorly compliant diseased lung tissue resulting from
87 chronic parenchymal remodeling.

88 Pneumothorax is generally regarded as a late complication of sarcoidosis, occurring preferentially in
89 fibrotic or bullous forms [2]. However, it may occasionally be the presenting manifestation of the disease
90 or the first sign of relapse [2,3]. In our patient, pneumothorax developed despite clinical improvement
91 under corticosteroid therapy, highlighting that control of inflammation does not prevent mechanical
92 complications arising from irreversible structural damage.

93 Initial management of pneumothorax complicating sarcoidosis relies on chest tube drainage, in
94 accordance with recommendations for secondary pneumothorax. Nevertheless, the possibility of
95 recurrence and the risk of persistent air leakage may justify pleurodesis or surgical management in
96 selected patients with an unfavorable clinical course [2,5]. The role of corticosteroid therapy in
97 preventing pneumothorax recurrence remains controversial, as the available evidence is limited and
98 does not establish a consistent protective effect [2].

99 Finally, this case emphasizes that sarcoidosis should be considered in patients presenting with
100 spontaneous pneumothorax and diffuse pulmonary involvement, particularly when respiratory
101 functional impairment is disproportionate or suggestive extrapulmonary signs are present. Early
102 recognition of this association enables appropriate comprehensive management and improved
103 assessment of respiratory prognosis.

104 **Conclusion**

105 Spontaneous pneumothorax is a rare complication of pulmonary sarcoidosis, generally associated with
106 advanced fibrotic forms. Its occurrence requires prompt management and comprehensive reassessment
107 of the disease, given the potential for progression and recurrence.

108 **Declarations**

109 **Patient Consent for Publication**

110 Written informed consent for publication of this case report and the accompanying clinical images was
111 obtained from the patient.

112 **Conflicts of Interest**

113 The authors declare no conflicts of interest.

114 **Funding**

115 The authors received no specific funding for this work.

116 **Author Contributions**

117 All authors contributed to the clinical management, preparation, and critical revision of the manuscript.
118 All authors read and approved the final version.

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References

1. Valeyre D, Prasse A, Nunes H, Uzunhan Y, Brillet PY, Müller-Quernheim J. Sarcoidosis. *Lancet*. 2014;383(9923):1155–1167. doi: [10.1016/S0140-6736\(13\)60680-7](https://doi.org/10.1016/S0140-6736(13)60680-7).
2. Manika K, Kioumis I, Zarogoulidis K, et al. Pneumothorax in sarcoidosis. *J Thorac Dis*. 2014;6(Suppl 4):S466–S469. doi: [10.3978/j.issn.2072-1439.2014.09.11](https://doi.org/10.3978/j.issn.2072-1439.2014.09.11).
3. Froudarakis ME, Bouros D, Voloudaki A, et al. Pneumothorax as a first manifestation of sarcoidosis. *Chest*. 1997;112(1):278–280. doi: [10.1378/chest.112.1.278](https://doi.org/10.1378/chest.112.1.278).
4. Ayed Della S, Kotti A, Ben Sik Ali H, et al. Pneumothorax spontané et maladie de Recklinghausen : à propos d'un cas. *Rev Pneumol Clin*. 2012;68(3):202–204. doi: [10.1016/j.pneumo.2011.08.001](https://doi.org/10.1016/j.pneumo.2011.08.001).
5. Light RW. *Pleural Diseases*. 6th ed. Philadelphia: Lippincott Williams & Wilkins; 2013. Chapter 24: Pneumothorax.