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# Diffuse bronchiectasis as the primary manifestation of endobronchial sarcoidosis

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ABSTRACT

Sarcoidosis is an idiopathic disease that most commonly involves the lungs and is characterized by granulomatous inflammation. Bronchiectasis is one pulmonary manifestation of sarcoidosis, although it is almost always observed as traction bronchiectasis in the setting of fibrotic lung disease. A 50-year-old woman was evaluated for chronic cough and bronchiectasis with a small amount of peripheral upper lobe honeycombing and no significant pulmonary fibrosis or lymphadenopathy. After an extensive laboratory and imaging evaluation did not identify a cause of her bronchiectasis, bronchoscopy was performed to assess for primary ciliary dyskinesia and revealed a diffuse cobblestone appearance of the airway mucosa. Endobronchial biopsies and lymphocyte subset analysis of bronchoalveolar lavage fluid were consistent with a diagnosis of sarcoidosis. We believe endobronchial sarcoidosis should be included in the differential diagnosis of patients presenting with bronchiectasis.



## 1. Introduction

Sarcoidosis is an idiopathic granulomatous disease that can affect every organ system, most commonly the lungs, which are involved in over 90% of cases [1,2]. There are numerous clinical manifestations of pulmonary sarcoidosis, including pulmonary nodules, fibrotic lung disease, airway hyperreactivity, mucosal nodules and plaques, bronchostenosis, and bronchiectasis [3]. The most common cause of bronchiectasis in sarcoidosis is traction bronchiectasis and has been reported in up to 40% of patients with fibrotic lung disease due to sarcoidosis [4]. We present a rare case of diffuse bronchiectasis due to endobronchial sarcoidosis.

### 1.1. Case report

A 50-year-old-woman with allergic rhinitis and infertility (three attempts at *in vitro* fertilization were unsuccessful) was referred to pulmonary clinic for bronchiectasis and chronic cough. She reported several years of a chronic cough productive of pale yellow sputum, as well as recurrent respiratory and sinus infections that would respond to oral antibiotics. She denied any smoking history and occupational or environmental exposures, and her medications included gabapentin, levocetirizine, and intranasal fluticasone. Her vital signs and physical exam were within normal limits. Pulmonary function tests revealed an isolated reduction in the DLCO to 66% predicted. A high-resolution chest CT demonstrated diffuse upper lobe-predominant bronchiectasis and a small amount of peripheral honeycombing in both upper lobes and scattered tree-in-bud and ground glass opacities (Fig. 1); the bronchiectasis and peripheral honeycombing had progressed slightly compared to a chest CT performed eight years prior and there was no significant pulmonary fibrosis or hilar or mediastinal lymphadenopathy.

A laboratory evaluation was initiated in an attempt to identify the cause of her bronchiectasis. Screening tests for connective tissue disease including rheumatoid factor, anti-CCP antibody, antinuclear antibody, anti-Smith antibody, anti-SSA and SSB antibodies, and anti-RNP antibody were all negative. The serum IgE was elevated to 187 IU/mL, but *Aspergillus* precipitins and serum-specific IgE antibodies to *Aspergillus fumigatus* were negative. Quantitative immunoglobulins and IgG subclasses were normal. A sweat chloride test was negative and a CFTR gene mutation analysis was normal.

In order to obtain targeted samples for culture and test for primary ciliary dyskinesia, a bronchoscopy was performed. Endobronchial biopsies of the main carina were collected with forceps and brushes for transmission electron microscopy, which showed

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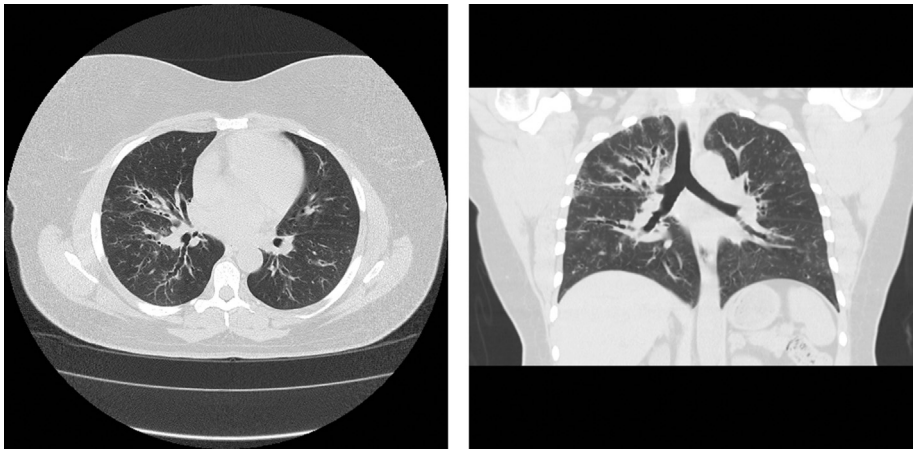

**Fig. 1.** a - HRCT Axial. b - HRCT Coronal.

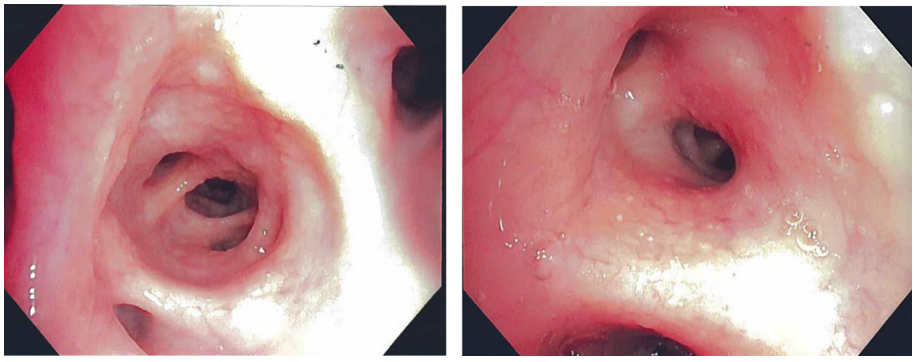

**Fig. 2.** a - Right Lung Bronchoscopy. b - Left Lung Bronchoscopy.

no evidence of primary ciliary dyskinesia. Bacterial, fungal, and mycobacterial cultures obtained from bronchoalveolar lavage (BAL) fluid and brushes of the right upper lobe grew no organisms. Airway survey unexpectedly revealed diffuse cobblestoning of the endobronchial mucosa distal to the secondary carinas (Fig. 2), so additional endobronchial forceps biopsies and transbronchial biopsies of the right upper lobe were collected for histopathology and the BAL fluid was sent for lymphocyte subset analysis. The CD4:CD8 ratio of the BAL fluid returned elevated at 8.9 and the endobronchial biopsies showed well-formed, non-caseating granulomas with chronic inflammation and giant cells with Schaumann bodies (Fig. 3). Given the diffuse endobronchial cobblestoning, elevated BAL CD4:CD8 ratio, non-caseating granulomas with Schaumann bodies, and negative cultures, she was diagnosed with sarcoidosis.

## 2. Discussion

The non-caseating granuloma is the histopathologic hallmark of sarcoidosis and in one prospective study of 34 patients with suspected sarcoidosis was found on endobronchial biopsy in 61.8% of

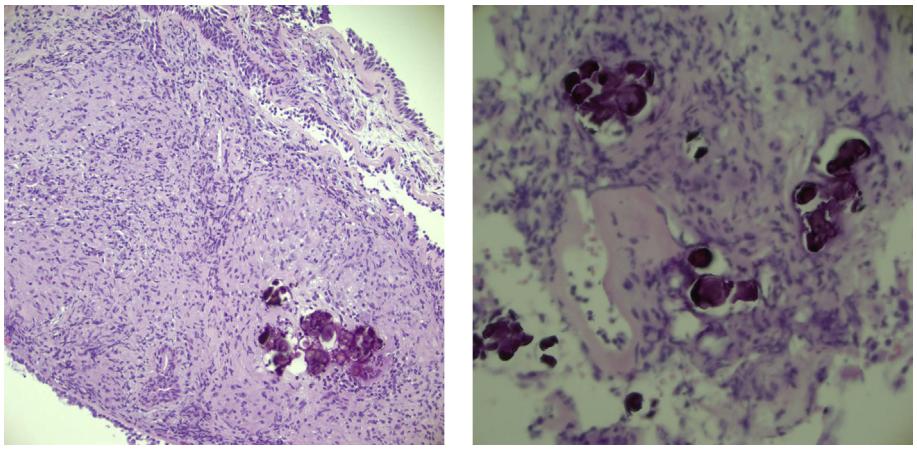

**Fig. 3.** a - Endobronchial Granulomas. b - Schaumann Bodies.

patients. Endobronchial biopsy was more likely to be positive in patients with an abnormal-appearing airway mucosa, but still revealed granulomas in 30.0% of patients with a normal-appearing airway mucosa [5]. Schaumann bodies, also known as conchoidal bodies, are concentric calcifications containing calcium oxalate crystals that are commonly seen in sarcoid granulomas [6,7], although they are not pathognomonic of the diagnosis [8].

When bronchiectasis is described in patients with sarcoidosis, the pathogenesis is almost exclusively attributed to traction bronchiectasis from fibrotic lung disease [3,9–12]. Several other mechanisms have been described, however, including chronic or recurrent infections [13] and extrinsic airway compression from enlarged lymph nodes causing a "middle lobe syndrome." [14,15] Endobronchial sarcoidosis has been reported as a cause of bronchiectasis; however, this is typically in association with bronchostenosis due to granulomatous inflammation and edema, an endobronchial mass, or scarring [9,13,16].

This case is unique due to the presence of diffuse bronchiectasis without any significant fibrotic lung disease, bronchostenosis, or lymphadenopathy in a patient with endobronchial sarcoidosis. In this patient, bronchiectasis may have developed as the result of extensive granulomatous inflammation that caused bronchial wall damage [9,16]. Since the majority of cases of non-cystic fibrosis bronchiectasis are reported to be idiopathic [17,18], we suggest that bronchoscopy with endobronchial biopsy be considered as part of the diagnostic evaluation in these patients to assess for possible endobronchial sarcoidosis.

## Conflicts of interests

None.

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## Disclaimer

The views expressed herein are those of the authors and do not reflect the official policy or position of Brooke Army Medical Center, the U.S. Army Medical Department, the U.S. Army Office of the Surgeon General, the Department of the Army, the Department of the Air Force and Department of Defense or the U.S. Government.

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