Title

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CLINICAL VIGNETTE

Endobronchial Sarcoidosis

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Introduction

Sarcoidosis involving thoracic lymph nodes and pulmonary parenchyma is familiar to most clinicians. Airway involvement has been observed in nearly two-thirds of patients but is often overlooked.

Case

A 58-year-old African American female with no significant past medical history presents with three months of dyspnea on exertion with occasional wheezing and non-productive cough. Symptoms progressed to dyspnea with walking three blocks and with any incline. She had a fifteen pound weight loss over three months. She denied fever, chills, night sweats, or rashes. Chest X-ray showed bilateral hilar adenopathy and increased reticular markings (Image 1). Computed tomography (CT) scan of the chest demonstrated bulky mediastinal adenopathy with mass effect, anatomic distortion of pulmonary vessels, and airway compression in addition to perilymphatic micro- and macro-nodules (Image 2). Bronchoscopy showed a slightly splayed carina, submucosal infiltration (Image 3a) and an approximate fifty percent luminal obstruction of the right upper lobe posterior segment by an irregular endobronchial mass (Image 3b). Bronchoalveolar lavage was negative for bacterial, fungal, and acid fast organisms. Endobronchial biopsy of the right upper lobe endobronchial mass and endobronchial ultrasound-guided fine needle aspiration from the subcarinal lymph node showed coalescing epithelioid granulomas. The aspirate cultures were negative for fungal and mycobacterial organisms. She was diagnosed with sarcoidosis with endobronchial involvement and was started on oral corticosteroid therapy.

Discussion

Sarcoidosis is a multi-system granulomatous disease of unknown cause that is characterized by the formation of noncaseating epithelioid cell granulomas. Sarcoidosis involves the respiratory system in more than 90% of cases, typically involving hilar and mediastinal nodes and lung parenchyma. Airway involvement, based on clinical features, physiologic testing, imaging techniques, bronchoscopy, and airway mucosal biopsy, has been observed in nearly two-thirds of patients with sarcoidosis.[1]

Sarcoidosis can involve any portion of the respiratory tract. Nasopharyngeal, laryngeal, and tracheal disease can be seen in approximately 5% of cases; these cases need to be differentiated from granulomatosis with polyangiitis.[2]

Endobronchial involvement is common in sarcoidosis. Even normal appearing mucosa has shown epitheloid granulomas on endobronchial biopsy.[1,3,4] The most common initial abnormality is mucosal edema, erythema and fine granularity as seen in Image 3. Sarcoid nodules can be seen in the bronchi and may cause post-obstructive atelectasis, airway luminal narrowing and distortion. They classically are 2-3mm waxy yellow mucosal lesions that are diffuse and coalesce to form the classic mucosal cobblestone appearance.[1]

In 2001, Schorr and colleagues evaluated 34 subjects with sarcoidosis in which the addition of endobronchial biopsy increased the yield of fiberoptic bronchoscopy with transbronchial biospies by 20.6%. Although more frequently positive in abnormal-appearing airways, endobronchial biopsy provided diagnostic tissue in 30.0% of patients with normal-appearing endobronchial mucosa as well.[4] This is less diagnostically relevant now that endobronchial ultrasound guided fine needle aspiration of lymph nodes has a diagnostic yield of approximately 80-90% in patients with mediastinal adenopathy and clinical suspicion of sarcoidosis. However, it certainly highlights the high frequency of airway involvement even in the absence of visualized abnormalities.

Our patient's endobronchial lesion (Image 3b) had a mass-like appearance and did not fit the usual description of a sarcoid nodule. Biopsy was therefore important to rule out other etiologies including infection and malignancy. Ultimately, all clinical, radiographic, and bronchoscopic findings were highly suggestive of sarcoidosis, and pathology was consistent with sarcoidosis.

Conclusion

Sarcoidosis most commonly involves the thoracic lymph nodes and pulmonary parenchyma though any portion of the respiratory tract, and any organ system of the body can be involved. Endobronchial involvement is quite common with presentation ranging from normal mucosa to a cobblestone appearance to endobronchial mass lesion, as in our patient.

Images

Image 1: Chest X-ray showing increased size and density of bilateral hilar regions with abnormal interface in the AP window. Constellation of findings is suspicious for lymphadenopathy. Increased reticular markings seen throughout the lung fields may represent peribronchial thickening versus interstitial thickening.

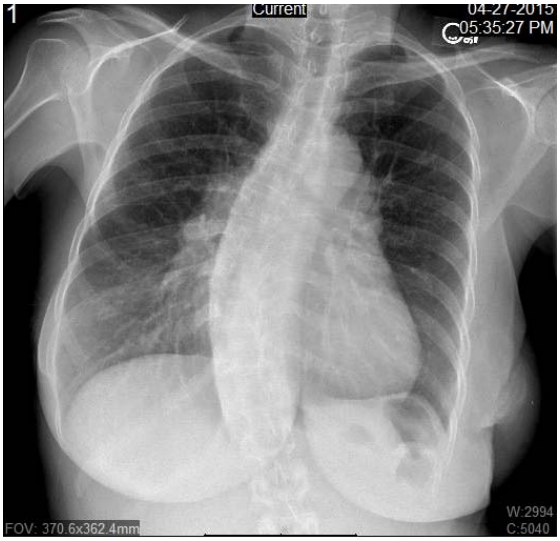
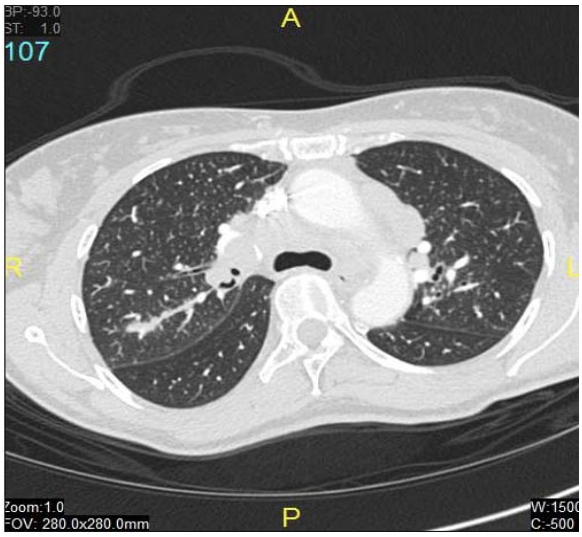


Image 2: Chest CT with contrast showing bulky mediastinal adenopathy involving bilateral paratracheal, aortopulmonary window, periaortic, subcarinal, bilateral tracheobronchial, interlobar, lobar, and intrapulmonary station lymph nodes. Upper lobe greater than lower lobe, predominantly perilymphatic peribronchovascular and lymphangitic nodular thickening is demonstrated with micro- and macro-nodules, including along the fissures.



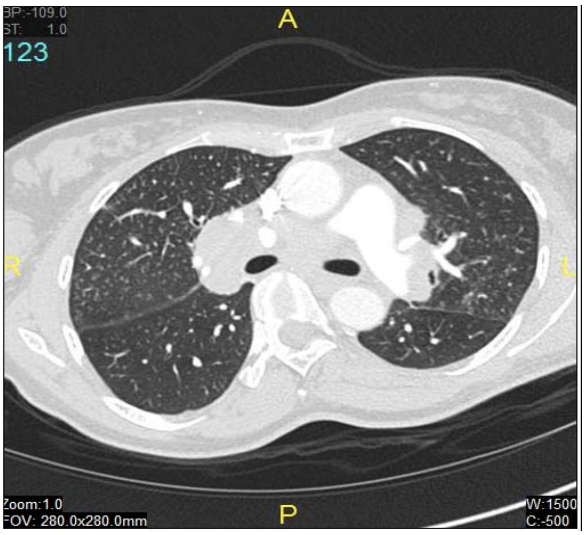
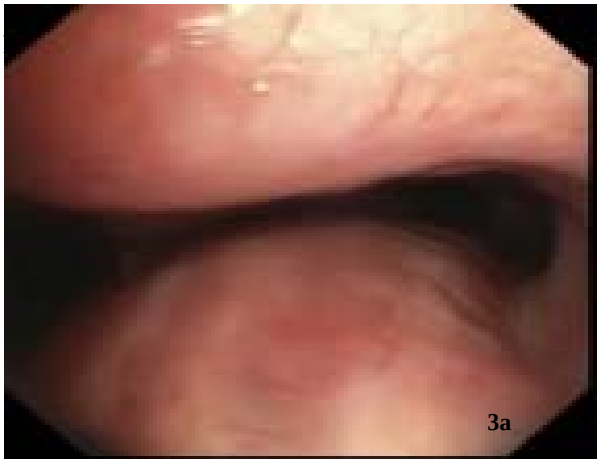
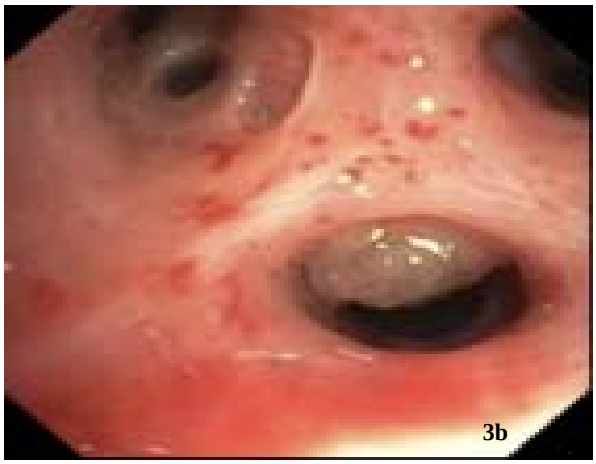


Image 3: Flexible videobronchoscopy showing submucosal infiltrated and edematous mucosa (3a) and approximately fifty percent luminal obstruction of the right upper lobe posterior segment by an irregular endobronchial mass (3b).





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