Granulomatous lung disease with an unusual presentation

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Background

Isolated granulomatous inflammation of the tonsils is rare, and a nonspecific diagnosis favors sarcoidosis, a granulomatous inflammatory disease with unknown etiology. Pulmonary actinomycosis is a rare and confusing diagnosis that can mimic sarcoidosis and share clinical features with chronic suppurative lung infections. Here we present a case of a young immunocompetent female with chronic tonsillitis found to have granulomatous tissue on biopsy. She had a subsequent lung involvement with a lung biopsy suggestive of a non-caseating granulomatous disease, along with microbiological evidence of pulmonary actinomycosis. Her clinical and radiological features failed to improve with antibiotic therapy for actinomyces treatment but showed a dramatic response to steroids supporting the diagnosis of sarcoidosis with a primary tonsillar involvement.

Case

40-year-old female with history of chronic tonsillitis and tonsillolith s/p elective tonsillectomy at outside hospital 8 months ago, presented with progressively worsening non-productive cough without hemoptysis, exertional dyspnea, 25lbs weight loss and generalized weakness post procedure. She denied smoking cigarettes or recent travel. Her TB IGRA, Hepatitis panel and HIV screen were negative. She was on room air, with diffuse bilateral rhonchi on exam. Her HRCT Chest showed bilateral multifocal nodules and cavitary lesions, mediastinal lymphadenopathy, with right lower lobe dense consolidation. She underwent transbronchial biopsy and lavage which was negative for fungus, acid-fast bacilli; pneumocystis and aspergillus PCR were also negative. The lung biopsy was positive for actinomyces odontolyticus filaments in BAL with background pathology of non-caseating non necrotizing granulomatous tissue. EBUS-guided lymph node sampling was non-diagnostic with normal pattern flow cytometry. Her ACE level later came elevated to 83 Ug/L and ANA was normal. Upon further inquiry, her initial tonsillectomy specimen was consistent with non-caseating granulomatous changes which was thought to be due to non-specific chronic inflammation. She completed IV antibiotic therapy with ceftriaxone for 4 weeks for actinomyces pneumonia treatment without much improvement but after initiating systemic steroid therapy, her symptoms improved drastically. Her repeat CT Chest 4 weeks after initiating steroid showed improving lung nodules and mediastinal lymphadenopathy.

Discussion

Very few cases of initial presentation of tonsillar sarcoidosis are described in literature. Isolated tonsillar sarcoidosis may develop as a granulomatous response to the usual stimuli for chronic tonsillitis. In the absence of any identifiable cause of granulomatous reaction, a nonspecific diagnosis of sarcoidosis is made. Tonsillar sarcoidosis, when present, often supports a subsequent multisystemic disease. About 80-90% of sarcoidosis patients have the lungs and the mediastinal lymph nodes involvement. Sarcoidosis may promote an anaerobic milieu to favor actinomycete growth.

Images

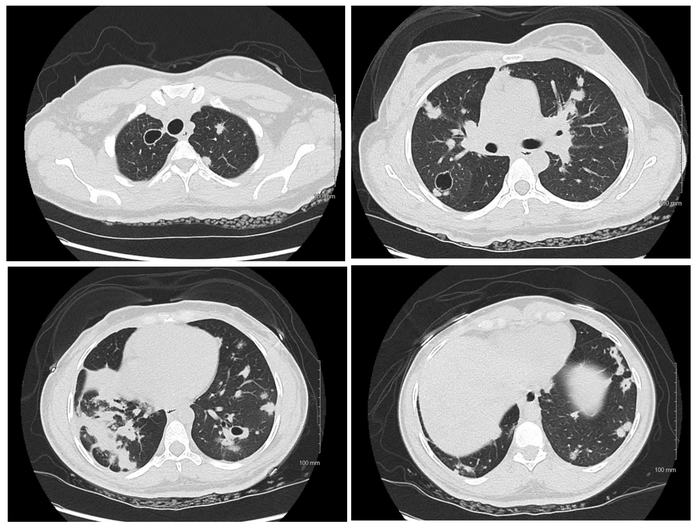

Figure 1: HRCT Chest showed bilateral multifocal nodular and cavitary lesions, mediastinal and hilar lymphadenopathy, with right lower lobe consolidation

Pulmonary actinomycosis clinically resembles other chronic suppurative lung infections, such as tuberculosis, fungal infections, and lung abscesses, as well as lung malignancy. It may have varied radiological presentation including patchy air-space consolidation, multifocal nodular appearances, cavitation, pleural thickening, pleural effusions and hilar, and/or mediastinal lymphadenopathy which can add to the diagnostic uncertainty. Sputum samples and bronchial brushings can also be nonspecific due to absence of adequate culturing technique and previous antibiotic therapy. Histopathologically, actinomycosis is mostly described a granulomatoid-like reaction.

The samples positive for actinomyces from sputum, BAL or brushings often represent colonization or contamination with saliva in absence of Sulphur granules in purulent infected tissue. Usually, improvement in chest radiographic findings is expected within 4 weeks of appropriate antibiotic treatment in pulmonary actinomycosis.

In addition to formation of abscesses, untreated actinomycosis may invade the pleura, chest wall, soft tissues, and bony structures through formation of sinus tracts especially with prolonged systemic steroids treatment in symptomatic pulmonary sarcoidosis.

Teaching Points

-The sensitivity and specificity of radiographic findings and bronchoalveolar lavage in diagnosis of pulmonary actinomycosis is low.

-The diagnosis of pulmonary actinomycosis becomes further complicated in patients with chronic granulomatous disease who have a higher risk of developing an invasive disease.

-The diagnosis of sarcoidosis is not standardized and the exclusion of alternative causes of granulomatous diseases remains one of the three major diagnostic criteria.

References

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