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Pulmonary Hyalinising Granuloma: A rare pulmonary disorder

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Abstract

Pulmonary hyalinising granulomas are rare, non-infectious fibrosclerosing lesions of the lung which can mimic metastatic disease. It was first described in literature by Engleman et al in the year 1977. Its etiology is unknown but they may be caused by an exaggerated immune response. The patient typically presents with cough, chest pain, dyspnoea or haemoptysis in association with multiple bilateral parenchymal nodules. We report the case of a 20 years old male who presented with a 12-month history of

worsening dry cough. His plain chest radiograph and subsequent CT scan revealed bilateral pulmonary nodules. A CT guided biopsy of the pulmonary lesions was consistent with Pulmonary Hyalinising Granuloma [PHG].

Keywords: Pulmonary Hyalinising Granuloma, Lung nodules.

Introduction

Pulmonary hyalinising granulomas (PHG) are rare, non-infectious fibrosclerosing lesions of the lung which can

mimic metastatic disease. Their etiology is unknown but they may be caused by an exaggerated immune response to an unknown antigen.¹ The clinical symptoms are nonspecific, including cough, fever, fatigue, dyspnea, pleuritic chest pain, sinusitis, and pharyngitis but several patients are completely asymptomatic.² Radiography and CT findings showed randomly distributed nodules and masses with well-defined borders, some with and some without calcium. Although not commonly seen, the calcification is usually focal, central and irregular.³ Open Biopsy normally reveals haphazard dense collagen bands surrounded by lymphoplasmacytic infiltrate that sometimes form germinal centers. In general, the pathology is very characteristic: the centre of the lesion consists of extracellular, eosinophilic hyalinised collagen usually arranged in parallel lamellae, but it may be quite disorganized. The peripheral areas tend to be more cellular with clusters of lymphocytes, histiocytes and plasma cells and germinal centres may be seen.¹ The appearance is easily confused with nodular amyloid. The clinical course is usually self limited and benign, but 30% of patients develop progressive inflammatory disease, with enlarging lesions and increasing dyspnoea.⁴

First described by Engleman et al in 1997 affects mostly adults from the 20s to the 70s showing no sex predilection and typically runs an indolent course.⁵ No specific medical interventions have been shown to influence granuloma size. Patients generally remain asymptomatic or may develop mild constitutional disturbance. Although the nodules tend to grow slowly, spontaneous regression or disease stabilization has been described.^{5,6} There is no effective treatment for pulmonary hyalinising granuloma; however, a few cases, especially those with progressive disease, have been reported that showed improvement with corticosteroids.^{1,3,7} The prognosis is usually excellent.

Case History:

We report a case of a 20 years old male who presented with 12 months history of worsening dry cough. There was no significant history of dyspnoea, chest pain, haemoptysis or fever. The patient denied any history of rash, arthralgia, oral ulcer or haematuria. Prior to this he reportedly had no physical ailments or functional impairment. In his prior workup, a chest radiograph had shown bilateral nodular densities, for which he had taken various antibiotics including a seven months course of anti-tuberculous drug therapy prescribed by his primary care physician. The sputum smears were negative for Acid fast bacilli (AFB) and there had been no improvement in his symptoms or his chest radiograph with this therapy.

He did not have any history of pulmonary tuberculosis (TB) or Asthma. He was a shopkeeper by profession and a non-smoker. There was no history of TB contact or a family history of respiratory or autoimmune illnesses.

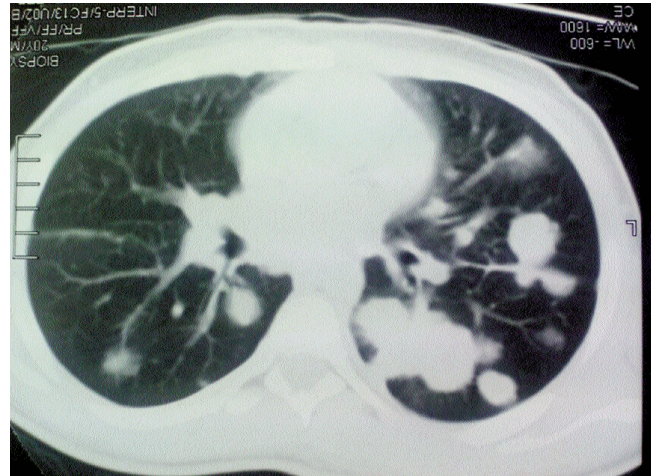


Figure-1: CT scan chest showing well circumscribed nodular soft tissue densities bilaterally.

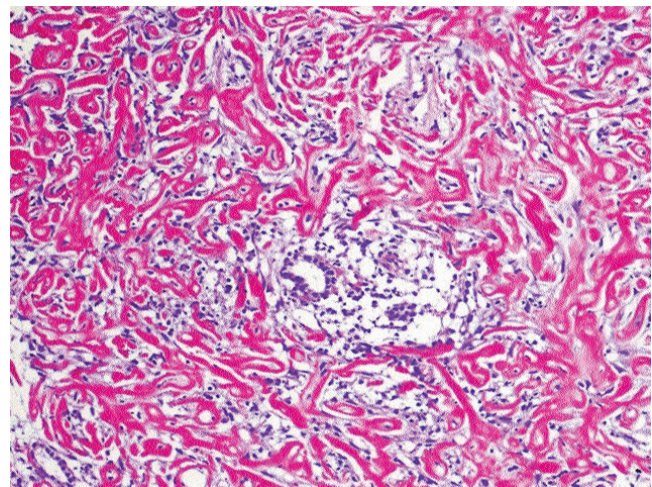


Figure-2: Keloid type thick interweaving collagen bands, with entrapped respiratory epithelium (Haematoxylin and Eosin. Original magnification x 200).

On examination he was thin, and lean with no signs of respiratory distress. His oxygen saturations were 97% on room air. There were no significant findings on general physical as well as on chest examinations.

His CT scan of the chest was done which showed bilateral nodular soft tissue densities with no significant lymphadenopathy or pleural effusion (Figure-1). CT guided biopsy of his lung nodules revealed linear cores of tissue predominantly consisting of keloid type thick interweaving collagen bands, focally arranged in concentric lamellae around the blood vessels (Figure-2). The material was negative for Congo Red, GMS, Silver red and PAS special stains. Entrapped respiratory epithelium (positive for CK AE1/AE3) showed sparse chronic inflammatory cell in-

filtrate. The edge of tissue showed a mixed acute and chronic inflammation. Foci of calcification were also noted. These features were highly suggestive of Pulmonary Hyalinising Granuloma.

Discussion

This case report describes one of the youngest patients known to have PHG (the youngest patient reported was aged 19 years).⁸ He was relatively asymptomatic, the only complaint being chronic cough, despite significant lesions on the chest radiographs, which on biopsy turned out to be PHG.

PHG typically presents as bilateral, multiple nodules in the sub-pleural or intrapulmonary lung tissue. Also in the literature there are some reports about solitary, unilateral and central manifestation.⁴⁻⁶ Therefore PHG should not only be considered in patients showing multiple pulmonary nodules but also in patients showing solitary nodules.⁹

The pathogenesis of PHG is still obscure. Several authors proposed that the lesion represents a continuing immune response to agents such as fungal organisms (e.g. Histoplasmosis) or tubercle bacilli.^{5,6} Support for this hypothesis has come from the demonstration of a variety of auto-antibodies in sera of patients with PHG. To date, there have been reports of anti-antinuclear anti-bodies (ANA), rheumatoid factor (RA factor), anti-neutrophil cytoplasmic antibodies (ANCA), anti-smooth muscle antibodies (ASMA), anti-mitochondrial antibodies (AMA) and Coombs-positive hemolytic anemia.^{6,10}

Our patient did not demonstrate any clinical features suggestive of an autoimmune process however a detailed autoimmune workup could not be undertaken due to financial constraints.

The patient is being followed up over the last 6

months and he has not shown any signs of deterioration symptomatically and there have been no significant change in the size of the pulmonary lesions on chest radiography.

Conclusion

Pulmonary hyalinising granuloma, a usually benign condition, should be kept in mind when encountered with patients presenting with nonspecific chest symptoms and bilateral pulmonary nodules on chest radiographs. Every possible effort should be made for obtaining tissue diagnosis because; although rare but certain benign conditions like PHG do exist which rarely can have prognostic significance in patient's long-term survival.

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