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Unusual Lung Presentation of Wegener's Granulomatosis- A Case Report

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Wegener's Granulomatosis, vasculitis, anti-neutrophil cytoplasmic antibody, Pleural effusion.

Abstract
Wegener's granulomatosis Is Well Known For Its pulmonary symptoms (WG). however, there hasn't been any mention of a pleural effusion as a first symptom. Here We present a case of pleural effusion which is later diagnosed as Wegener's granulomatosis. A 50-year-old male presented with pleuritic chest pain, CT Chest revealed pleural thickening and nodule, on examination there was palpable nodule on right side of chest. Pleural biopsy and lung

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biopsy showed thick chronic inflammatory infiltration in pleural biopsy and necrotizing granulomatous vasculitis. Laboratory investigations are positive for Wegener's granulomatosis and patient is started on cyclophosphamide and methylprednisolone, for which he showed improvement.

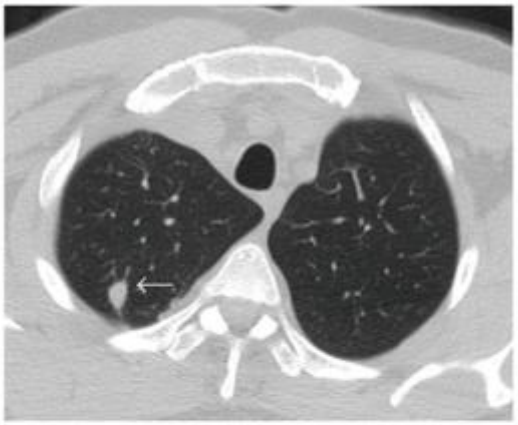
1. Introduction

Wegener's granulomatosis (WG) is a vascular disorder of small and medium-sized arteries. Although the symptoms vary, pulmonary abnormalities are typical. Most common thoracic lesions are intrapulmonary nodules, typically having irregular margins in cortical and excavated but less common pleural involvement and consolidation of parenchyma, rarely in pleural effusion noted in initial presentation.

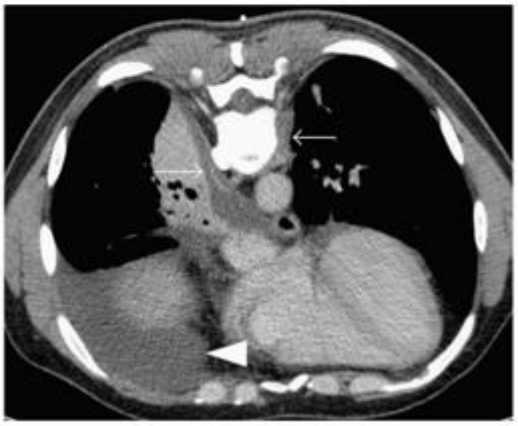
2. Case Report

A 50 years old gentleman came with complaints of severe right pleuritic chest pain over dorsal side requiring morphine. No history of smoking or alcohol intake. He is a worker at metal factory for 25 years. Except for asthenia and neurological symptoms, the physical examination was normal (pain in the right thoracic region, both sides paresthesia, anddysesthesiaC8-D1). Initial chest-x-ray showed no significant abnormality. Later CT Chest showed Right side upper lobe having a nonspecific1.5cm nodule, pleural effusion, bilateral subpleural thickening in the 9thvertebral region. WBC count was 12000/mm3 and eosinophil count was normal. Renal function test is normal. Bronchoscopy showed inflammation of right lower lobe bronchus and biopsy is nonsignificant. Later thoracocentesis is showed pleural fluid was sterile exudative lymphocytic predominance (lymphocytosis 85%, neutrophilsis10%, monocytes is 5%) and no malignant cells during surgical resection of the nodule, pleural biopsy taken.

Case Reports



(a)



(b)

Figure1: (a) Plain CT shows right upper lobe has nonspecific nodule(arrow). (b) Contrast CT shows at 9th vertebrae, pleural effusion in the right side (arrowhead) and both sides sub pleural thickening (arrow).

According to histological study of pleura, a thick inflammatory infiltration, predominates around the periphery of the arteries, most commonly having lymphocytes and plasma cells. Pulmonary specimen showed a mass which is centrally necrotic surrounded by palisading histiocytes (Fig 2).

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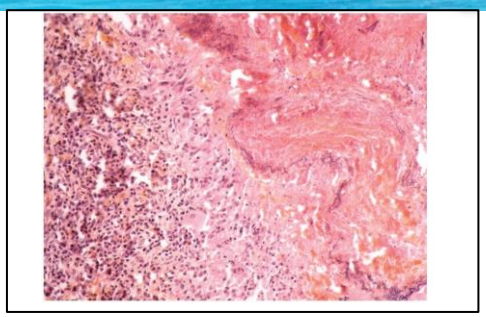

Figure 2: Histoscopic examination shows that pulmonary nodule has necrotizing area in the center surrounded by histiocytes in which few giant and multinucleated. Elastic staining shows partial destruction of the elastic lamnia by granulomatous inflammation (Magnification of elastic strain×200).

Few multinucleated histocytes had granulomas within pulmonary artery wall. In serologies, ANCA antibodies and antiproteinase 3antibodies (APR3) (129IU; normal30) were highly positive. A renal biopsy was not done in view of normal urine routine examination and renal function test. Skin examination showed nonspecific ulceration in the forehead. A sinus scan showed perforation of septum and thickening of ethmoidal sinus.

Hence, the patient is diagnosed as Wegener's Granulomatosis by histopathology, ANCA and APR3 positivity and treated with intravenous methylprednisolone initially, followed by oral prednisolone and cyclophosphamide. Four months later Azathioprine and low dose of glucocorticoid given. The patient's condition improved, and the peripheral neuropathy symptoms is reduced. Antibody levels decreased (ANCA+, APR3), and repeat thoracic scan recovered to normal (no residual thickening and no pleural effusion) (6 IU).

3. Discussion

Differentiated diagnosis of pleural effusion with thoracic pain, asthenia and lymphocytes in pleural fluid is purulent pleurisy, tuberculosis, malignant disease, parietal and neurological disorders (epiduritis/trauma). Pleural damage is uncommon in Wegener Granulomatosis (48 percent–73 percent at presentation, 85 percent–92 percent during the disease), while parenchymatous lung manifestations are prevalent (48 percent–73 percent at presentation, 85 percent–92 percent during the disease). Only ten percent of WG patients with early lung involvement have pleural lesions. The most common features are pleural thickening and pleural effusion. Pleural thickness is due to prior effusion or cicatricial alterations caused by inflammation of nodules extending into the pleura. Commonly pleural effusion is unilateral with small volume and significant pachypleuritis. Pleural fluid tapping and biopsies are challenging procedures. Except with infectious nodule and ruptures with in the pleural space, with sterile pleural exudate and associated with hypoglycopleury and predominate neutrophils. The diagnosis is guided by the presence of additional pulmonary injury (excavated or non - excavated nodule, infiltration) on imaging. Kumasaka et al. [5] recorded one case of endotoxin-induced small vessel vasculitis with pleuropulmonary symptoms. Immuno histochemistry withIL-1 and VCAM-1 shows vasculitic lesions having activated monocytes and endothelial cells. In one of the two cases, effusion shows increased eosinophilic but normal peripheral blood eosinophilic count was documented in isolated and inaugural pleurisy described in WG [2]. Few other cases published with similar presentation are (A66 year old female with fever, unilateral pleural chest pain, raised WBC counts, A 76 year old female with chest pain, dyspnea, left pleural effusion, WBC counts12000, nil eosinophils in exudate). The patient in our case had thickening of pleura and effusion. The diagnosis was based on clinical, radiological, histological, and biological findings.

4. Conclusion:

It is pointed out that pleural effusion can be initial presentation of Wegener's granulomatosis and also vasculitis. other differential diagnosis should be ruled out like infections and malignant conditions. As this disease is responding to immunosuppressive drugs and other localizations of Wegener's granulomatosis, based on clinical, radiological, histopathological and other findings diagnosis is supportive for Wegener's granulomatosis.

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