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Sarcoidosis: An Unusual Case of Pleural Effussion

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Abstract: ***Sarcoidosis is a multisystem granulomatous disease of unknown etiology in which pleural effusion (PE) is very rare. Tubercular as well as malignant pleural effusion is much more common and first need to be excluded. We are presenting a case of 58 year old female patient who developed pleural effusion (PE) unresponsive to Antitubercular therapy.***

Keywords: sarcidosis, granulomatous, etiology, pleural effusion

1. Introduction

Sarcoidosis is a multisystem granulomatous disease of unknown etiology in which pleural effusion (PE) is very rare. Tubercular as well as malignant pleural effusion is much more common and first need to be excluded.

We are presenting a case of 58 year old female patient who developed pleural effusion (PE) unresponsive to Antitubercular therapy.

2. Case Report

A 58 years old female who was a known case of hypertension for 10 years (on regular medication)and hypothyroidism for 5 years presented with dry cough for 7 months . She was noted to have low grade fever and cervical lymphadenopathy during this period. She was evaluated in another hospital where a computed tomography (CT) Scan thorax showed bilateral pleural effusion and mediastinal lymphadenopathy. The pleural fluid was exudative on aspiration but ADA was in the normal range, a Sputum acid fast bacillus (AFB) was negative. Cervical lymph node fine needle aspiration cytology (FNAC) showed granulomatous lymphadenitis. Cervical lymph node biopsy was done which showed necrotising granulomatous lymphadenitis. On the basis of biopsy

Antitubercular therapy (ATT) was started. But patient got no relief in symptoms and she presented to this hospital, by then she had completed 6 months of ATT. The routine Lab investigations were found to be normal.

A chest X-Ray thorax revealed bilateral pleural effusion (R>L), [FIG 1] that had increased, cervical lymphadenopathy was unchanged. Contrast enhanced computed tomography (CECT) showed borderline enlarged mediastinal lymph nodes, left axillary lymphadenopathy was noted. The USG abdomen was non contributary. Mantoux test was < 2mm in induration. Serum ACE level was 73 U/L (normal upto 60). Left Cervical Lymph node FNAC was repeated and showed non necrotizing granulomatous lymphadenitis, no AFB was seen.

Pleural fluid aspiration was exudative (protein- 5.1 g/dl) with low LDH (145 IU/L) and ADA of 9.1. Predominant cells were lymphocytes and no malignant cells were seen. AFB/Fungal/ Gram stains and aerobic culture/ Gene Xpert were negative in the pleural fluid. A USG breast was done which showed multiple well defined hypoechoic areas in bilateral axillary and intramammary region suggestive of lymph nodes. Mammography revealed dense left axillary nodes.

Left axillary lymph node biopsy was done which showed multiple non caseating granulomas comprising of many epitheloid cells, few foreign bodies and langerhans type of giant cells and scanty lymphocytes. No AFB was demonstrated. There was no evidence of malignancy. HLA Typing in blood showed presence of HLA DRB1*14 allele. (1) The tests for ANA, RA factor were negative.

The supporting evidence for sarcoidosis were a) raised ACE b) lymphocytic predominant pleural fluid with raised protein but low LDH c) lymph node biopsy showing non caseating granuloma d) positive HLA typing for sarcoidosis. The evidence for ruling out tuberculosis were no response to ATT, negative Mantoux test, pleural fluid-gene expert negative, AFB stain negative, ADA negative. Malignancy was ruled out as there was no evidence of malignant cells in pleural fluid and biopsy. There was no evidence of primary tumor in breast and abdomen as shown by mammography and USG abdomen.

Thus the patient was diagnosed as sarcoidosis presenting as pleural effusion with lymphadenopathy, glucocorticoid (prednisilone 30 mg per day) was started and now patient improved significantly with regression of effusion and lymph glands. [FIG 2]

3. Discussion

Pleural involvement in sarcoidosis accounts for 2 - 4 percent of cases. It may manifest as a) pleural effusion (previously thought to exclude sarcoidosis) b) pneumothorax c) pleural thickening d) hydropneumothorax e) trapped lung f) haemothorax g) chylothorax (2)

Szwarcberg and colleagues published their findings in a series of 61 patients with sarcoidosis using thoracic CT scans. The authors noted that 25 (41%) of the 61 patients had ‘‘pleural involvement’’ detected on CT. Of the 25 patients with pleural involvement, 20 (80%) had pleural thickening and five (20%) patients had pleural effusions; {8.2% (5/61)}. But, cause of pleural disease in this cohort of sarcoidosis patients was not determined. (3) In our own series of 153 patients pleural effusion was not found in Indian population with sarcoidosis.

Prevalence of pleural effusion in sarcoidosis is less than 3 percent in which right side accounts for 45 percent, left side 33 percent and bilateral 22 percent. (4) Effusion can be exudative or transudative. Diagnosis is made generally by

pleural biopsy which shows non - caseating granulomas. Majority resolve spontaneously in 1 to 3 months. (5)

It is not entirely certain as to why pleural involvement is rare in sarcoidosis, despite the fact that pulmonary parenchymal and nodal involvement is virtually present in almost all reported cases. The frequency of occurrence of pleural effusion in patients with sarcoidosis has been reported by Huggins et al. (2) as 2.8% (5 of 181 patients) with only 2 of the 181 PEs (1.1%) caused by sarcoid pleural involvement. The reasons for low incidence could be:- (a) mere presence of pleural effusion associated with sarcoidosis cannot be considered to be caused by sarcoidosis, (b) small pleural effusions can be missed on routine CXR (c) in a tuberculosis (TB) endemic country like ours, most pleural effusions are wrongly diagnosed as tubercular and empirically treated with ATT (d) lack of histopathological evidence of pleural involvement. Pleurisy in sarcoidosis has been considered to be related to either inflammation of visceral and parietal pleura caused by peripheral lung granulomas, or disturbance of venous and lymphatic circulations. They are typically – 1.paucicellular 2.lymphocytic predominant 3.exudative protein discordance with low LDH further supporting the view of increased capillary permeability with minimal pleural space inflammation. Our case was peculiar as there was no involvement of lung parenchyma, with multiple areas of extrapulmonary lymphadenopathy (cervical, axillary, mammary) along with pleural effusion was present. Pleural fluid was exudative, lymphocytic with protein LDH discordance. Axillary lymph node biopsy showed non caseating granuloma. Glucocorticoids were started and patient improved significantly with regression of pleural effusion and lymph glands.

The management of a sarcoid pleural effusion depends on the symptoms and biopsy evidence of disease. In the absence of symptoms one can expect an effusion to resolve spontaneously but if the effusion is symptomatic or recurrent, resolution will usually occur with glucocorticoid therapy. Development of pleural effusion during steroid therapy points more towards infections secondary to usual or opportunistic organisms.

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