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Systemic Primary Amyloidosis Presenting as Generalized Lymphadenopathy and Massive Pleural Effusion

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ABSTRACT
A 73 year- old man with cough, dyspnea, generalized lymphadenopathy and left sided pleural effusion was admitted with primary impression of lymphoproliferative disorders. The precise evaluation showed systemic primary amyloidosis with the rare presentation of generalized lymphadenopathy and massive pleural effusion without any other organ involvement as the available tests showed. ***(Tanaffos 2005; 4(16): 69-71)***
Key words: *Amyloidosis, Lymphadenopathy, Pleural effusion*

CASE SUMMARIES

A 73-year-old man was presented with progressive productive cough and dyspnea since 3 months before admission in hospital for a left sided hydropneumothorax. He had no specific antecedent except being heavy smoker (30 pack a year) which was discontinued since 26 years ago. In physical examination large, firm, bilateral lymphadenopathy in neck, supraclavicular and axillary regions were present in addition to decreased breathing sound in left hemithorax. Chest x-ray revealed left sided hydropneumothorax and CT-scan showed extensive middle and posterior mediastinal adenopathies with mild pericardial effusion as well. Lung parenchyma seemed normal in right side and partially collapsed in left side. With primary impression of lymphoproliferative disorders, a chest tube was inserted as a symptomatic treatment evaluating pleural effusion characters (the pneumothorax regarded to be iatrogenic).

Analysis of this fluid showed a transudative effusion with 95% mononuclear cells predominance in cytology.

All evaluations for AFB were negative. Echocardiopraphy showed normal left ventricular function (EF: 50%) with mild pericardial effusion. Laboratory exams were as follows: Urea: 25; Creat: 0.9; FBS: 107; Hgb: 14; and ESR: 13. Chest tube drainage was enormous (1-2 lit/d) despite the completely expanded left lung. For making diagnosis, at first left supraclavicular lymph node biopsy was preformed followed by a right supraclavicular lymph node biopsy; both pathologic reports were as amyloid lymphadenopathy. (both

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seen by pathologists from two different pathologic wards). In order to find the cause of persistent drainage of pleural cavity and evaluation of mediastinal lymph nodes, left thoracoscopy was performed in which the same pathologic result was achieved as "extensive deposition of amyloid material in mediastinal lymph node and chronic nonspesific inflammation with deposition of amyloid around the vascular channels".

Therefore, with the impression of systemic primary amyloidosis, work up for finding other organ involvements were carried out. Kidney and liver functions were normal. Protein electrophoresis showed abnormal monoclonal band in gamma region (mono-gammopathy) which reversed to normal after 2 months of beginning medical treatment, with negative Bence Jones protein in Urine.

FANA, C3, C4, CH50, DS-Anti DNA were all normal. Pleurodesis was performed to control effusion with bleomycin (60 mg) which was unsuccessful at first but became effective in the 2nd time. Patient was referred to a rheumatologist for follow up and treatment. He is under treatment now with cyclophosphamide(BD) and prednisolone (5 mg/d).

DISCUSSION

Amyloidosis is a subset of diseases with insoluble deposition of abnormal proteins in extra cellular regions (1, 2). More than 20 different proteins form clinically relevant amyloid deposits (1). Systemic amyloidosis is neoplastic, inflammatory, genetic or iatrogenic in origin, while localized amyloidosis or organ limited amyloidosis is associated with aging and diabetes (2). Though its clinical presentation depends on site and rate of amyloid deposition (1), it may have no apparent clinical consequences or may be associated with severe pathophysiologic changes (2).

Proteinuria is the most common presentation, (1, 2) and renal lesion seems to be progressive and irreversible (2), making its complications of the most common causes of death in these patients, followed by cardiac failure and arrhythmias (1).

Diagnosis is usually by pathologic confirmation of involved organ (Congo red staining, polarized beam) and use of immunohistochemistry. Deposition of amyloid may occur everywhere but its presentation with generalized lymphadenopathy is rare and may mimic lymphoma (3). Up to 37% of cases of generalized primary and secondary amyloidoses demonstrate lymph node involvement (4), but isolated lymph node amyloidosis is exceedingly rare (2 reported cases in literature) (4).

On the other hand, involvement of respiratoy system in systemic amyloidosis is more in the form of bronchopulmonary disease (5).

Pleural involvement is rare (5, 6, 7, 8) and its associated pleural effusion (uni- or bilateral) which is usually resistant to treatment might be transudative (6,7,9) or exudative (10). Thoracoscopy and biopsy can confirm it (5, 11), and pleurodesis is the present treatment modality (5), although decortications may be another option (12).

Even in the presence of cardiac involvement, pleural effusion might be due to pleural deposition rather than being the result of CHF (9).

To our knowledge, this case is the first one presenting with generalized lymphadenopathy without renal, cardiac, pulmonary or hepatic involvement, associated with pleural deposition and resistant transudative pleurisy. It belongs to systemic primary amyloidosis group.

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