

Short reports

Nodular pulmonary amyloidosis occurring in association with pulmonary lymphoma

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Abstract

A rare association between primary pulmonary lymphoma and pulmonary nodular amyloidosis in a 56 year old man is described.

Pulmonary amyloidosis is a rare condition, not related to any systemic disease and often asymptomatic, being discovered on a routine chest radiograph.¹ We describe a case associated with a pulmonary lymphoma, which as a primary tumour is also rare.

Case report

A 56 year old ex-coalminer presented with a 20 year history of pleuritic pain in the left upper chest. Two days before admission the pain increased and hemoptysis occurred. He had no clinical evidence of coalworkers' pneumoconiosis and had smoked heavily for 35 years. On examination there were wheezes over the left upper chest. There was no supraclavicular or cervical lymphadenopathy. A chest radiograph showed a left hilar mass and an infiltrate in the left upper lobe (fig 1). Fiberoptic bronchoscopy showed narrowing of the apicoposterior segment of the left upper

lobe and biopsy specimens were suggestive of a small cell carcinoma. Computed tomography showed no mediastinal lymphadenopathy but bilateral dense nodules were present in the pulmonary parenchyma, which were thought to be secondary to coalworkers' pneumoconiosis. Isotopic bone scanning and computed tomography of the brain showed nothing abnormal, and the patient was referred for surgical treatment of his early stage disease. At thoracotomy a mass surrounding the left upper lobe bronchus was palpable. Tumour also appeared to be infiltrating the tissue around the left vagus nerve. This was diagnosed as a small cell carcinoma on frozen section. Several subaortic lymph nodes showed no histological evidence of malignancy. In addition, there were multiple 1 cm diameter nodules throughout both lobes of the lung; wedge resections were performed on two of the nodules from the left upper lobe.

PATHOLOGICAL FINDINGS

The biopsy specimen from the left upper lobe bronchus measured about 1.5 × 0.5 × 0.5 cm and consisted of sheets of dark, round cells with a very high ratio of nucleus to cytoplasm. Immunohistochemical examination showed that the tumour cells were positive for leucocyte common antigen, indicating that the tumour was a small cell non-Hodgkin's lymphoma, not a small cell carcinoma as originally thought. The wedge biopsy specimens from the left upper lobe each measured about 4 × 2 cm and contained three nodules up to 1.5 cm in diameter. Histologically, the nodules consisted of eosinophilic masses, which obliterated the parenchyma and stained positively for amyloid. Within and around the masses were focal collections of lymphocytes and plasma cells. There were also occasional giant cells and areas of calcification and ossification (fig 2). The amyloid was resistant to permanganate oxidation,² indicating that it was derived from immunoglobulin. Immunohistochemical studies showed a disproportionate number of cells with kappa light chains in both the lymphoma and the amyloidomas.

The patient made an uneventful recovery and is currently undergoing treatment with chlorambucil and prednisolone at monthly intervals. At the last follow up he was tolerating the chemotherapy and the hilar mass had diminished in size on the chest radiograph. The amyloid nodules appeared to be slightly smaller also.

Discussion

Pulmonary amyloidosis is classified histopathologically into three major types—tracheo-bronchial, diffuse alveolar-septal, and nodular parenchymal types. The last form usually occurs in the older age group, with an average age of 63 years and a range of 38–95 years in one study. The prognosis is good, with no

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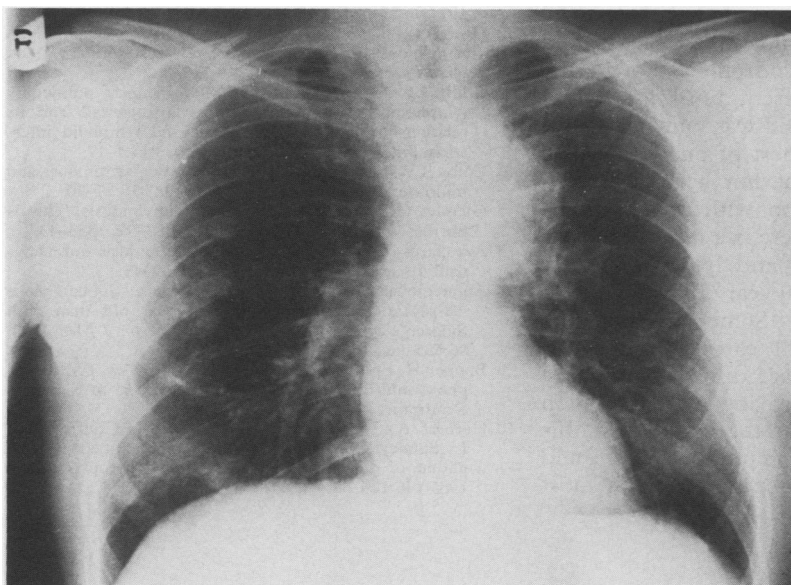
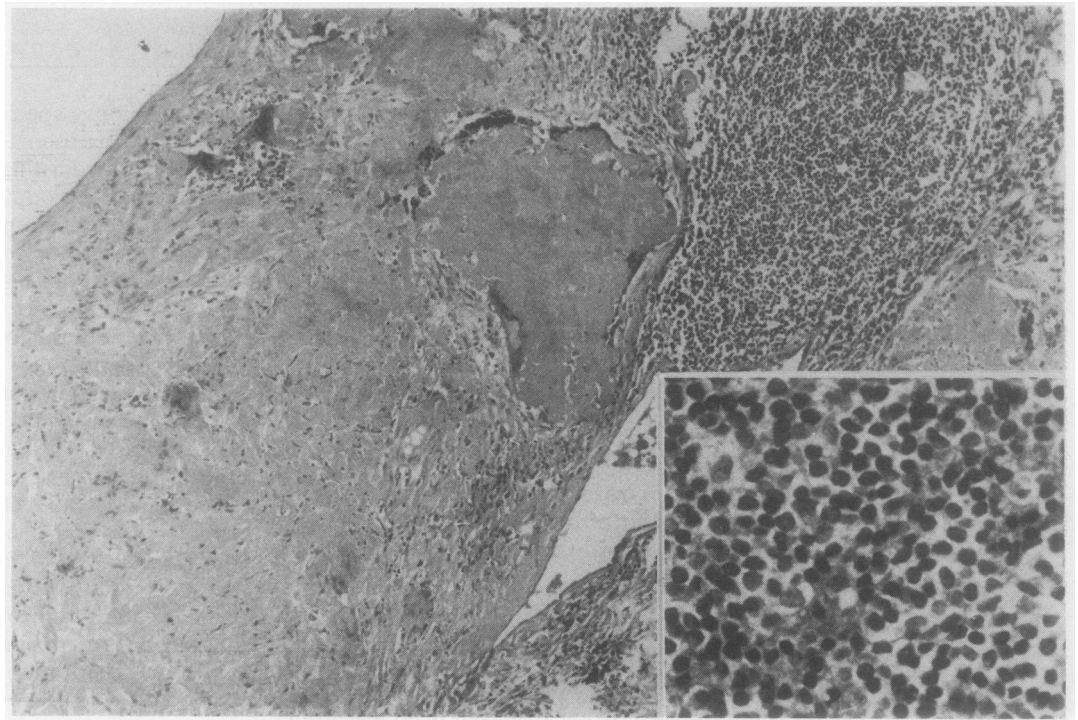


Figure 1 Posteroanterior chest radiograph showing a left hilar mass and multiple nodules in the middle and lower zones.

Figure 2
Photomicrograph showing an amyloidoma with foci of calcification, ossification, and giant cells. Inset: Well differentiated non-Hodgkin's lymphoma.



deaths in 28 reported cases.³ The tracheo-bronchial and diffuse alveolar forms usually present with respiratory symptoms and carry a worse prognosis.

The small lymphocytic type of primary lymphoma tends to run an indolent course and many patients are successfully treated by surgery alone.⁴ This case illustrates the usefulness of immunocytochemistry in diagnosis as this tumour was originally typed as small cell carcinoma. Sometimes amyloid is present within the stroma of the tumour⁵ but is not present in separate discrete nodules. In this case it is tempting to speculate that the amyloid was associated with the pulmonary lymphoma. The association between systemic amyloidosis and Hodgkin's lymphoma, and to a lesser extent non-Hodgkin's lymphoma, is well recognised.^{6,7} Nodular pulmonary amyloidosis occurred in association with a nodal immunoblastic lymphoma in a 72 year old man with Sjögren's syndrome,⁸ though the lymphoma did not present until four years after the nodular amyloid was known to have been present. To the best of our knowledge nodular amyloidosis has not been described previously in association with primary pulmonary lymphoma. There is a report of coexisting nodular amyloid and lymphoid interstitial pneumonia in a 59 year old woman with Sjögren's syndrome.⁹ Some investigators argue that some or all cases of lymphoid interstitial pneumonia are diffuse pulmonary lymphomas.¹⁰ The permanganate oxidation technique indicated that the amyloid in this case was derived from immunoglobulin, and agrees with an earlier report in which most

cases (8/11) were resistant to permanganate.³ The nodular amyloid in our case may have been present for a considerable length of time and was discovered only because of the onset of the pulmonary lymphoma. The finding of monoclonal kappa light chain in both the lymphoma and the amyloidomas, however, suggests that they are related. In a previous study all 11 amyloidomas tested were polyclonal in type.³ There is a hypothesis that pulmonary amyloid tumours arise from a local monoclonal proliferation of immunocytes,⁶ and this might be the case in our patient.

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