Amyloidosis with pleural involvement

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ABSTRACT: Although amyloidosis of the respiratory tract is well recognized, pleural involvement is very rare with only two cases being reported in the past. We report a case of primary amyloidosis with pleural effusion and suggest that pleural involvement and pleural effusion be added to the classification of pulmonary amyloidosis, and that amyloidosis be added to the list of causes of a pleural effusion.

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Amyloidosis of the lung is rare though its occurrence is well recognized [1, 2]. Involvement of the lower respiratory tract occurs in both the tracheobronchial tree and lung parenchyma. Pleural involvement is very rare, there has only been a single case report of pleural effusion [3] and pneumopleural amyloid "tumour" [4].

We report the case of a patient with primary amyloidosis who developed a pleural effusion and had amyloid deposition identified in both lung and pleura at thoracoscopy.

Case Report

A 52-yr old man presented to a cardiologist with a nine month history of dull central chest pain which was associated with exercise and was relieved by rest. The pain often radiated to his throat, was worse after meals and was accompanied by breathlessness. There was no relief with sublingual nitrate preparations. He had undergone a cholecystectomy and vagotomy ten years earlier and two years before presentation he had severe post-operative dyspnoea following reduction of a wrist fracture. Although no definite diagnosis was made, he was thought to have had a pulmonary embolus or aspiration pneumonia. There was no family history of heart disease nor was there any relevant occupational history. He was a life long non-smoker, drank no alcohol and his only medication was cholestyramine for slightly elevated blood cholesterol levels.

Physical examination at presentation was unremarkable apart from an intermittent 4th heart sound. The chest radiograph was normal. Electrocardiogram showed T-wave inversion in the lateral chest leads. Exercise ECG using the Bruce protocol was negative. Cardiac catheterization showed normal coronary arteries, the right coronary artery being dominant. There was some diffuse narrowing following ergometrine which was associated with chest pain and S-T segment depression.

It was concluded that there was no evidence of coronary artery disease and there was functional overlay to his symptoms. Over the next twelve months he was treated with nitrates, beta-blockers and calcium slow-channel antagonists, with no effect. He was referred to the pain relief clinic, and acupuncture was also unsuccessful.

One year after presentation he was admitted with severe breathlessness on exertion. He was found to have a tachycardia of 110 b·min−1, an elevated jugular venous pressure, bilateral ankle oedema, a 4th heart sound and signs consistent with a pleural effusion at the right base. Chest radiography showed cardiomegaly and confirmed the pleural effusion, there was no evidence of parenchymal lung disease. Echocardiogram showed both ventricles to be hypokinetic. Cardiac catheterization was repeated and again showed normal coronary arteries with global hypokinesis. Cardiac biopsy was performed (fig. 1) and showed obliteration of interstitial and myocardial fibres by an eosinophilic amorphous material, which was orange with Congo red and apple green under polarization. There was also amyloid infiltration of vessel walls. Electron microscopy was performed and confirmed the presence of amyloid.

The pleural fluid was tapped and biochemically was an exudate. The effusion reaccumulated two days later, by which time the cardiac failure had been resolved following diuretic treatment. Because the fluid was an exudate and thought to be due to an inflammatory process rather than cardiac failure, thoracoscopy was performed. The procedure was carried out using a Storz thoracoscope under local anaesthesia. Following induc-
Fig. 1. — Pleural biopsy, internal scale bar equals 10 μ. Congo Red Stain with polarization, showing amyloid material in the pleural interstitium.

Fig. 1.

 tion of pneumothorax, inspection revealed the lungs to have oedema of the interlobular septa, the pleural surfaces were hyperaemic and the chest cavity contained 500 ml of a brown turbid fluid. Biopsies were taken of the pleura and lung and both had been infiltrated by amyloid.

Comment

Amyloidosis affecting the lung was first described in a case of secondary amyloidosis by Virchow in 1857 [5]. Amyloid localized to the lower respiratory tract was reported by Lesser in 1877 [6]. Since that time the occurrence of amyloid in the lung has been well documented, indeed it has been suggested that in generalized primary amyloidosis, particularly when there is cardiac involvement, infiltration of the lung parenchyma is common [7, 8].

Most authorities agree that amyloidosis, when it affects the lung, can be classified into two main types; the tracheobronchial type which can be multifocal submucosal plaques or a tumour-like mass, and the parenchymal type in which the deposition may be nodular, either multiple or solitary, or may be that of a diffuse alveolar septal infiltration [2]. A review of the literature shows that pleural involvement is very rare. Pleural effusion with pleural amyloidosis was the subject of a case discussion at the Massachusetts General Hospital [3] and a pneumonopleural tumour was described in 1961 [4].

We suggest that pleural involvement be added to the classification of amyloidosis of the lower respiratory tract and that amyloidosis be added to the list of rare causes of a pleural effusion. Furthermore, the thoracoscope should be considered in making the diagnosis in cases of unusual infiltrative disease which may involve the lung and pleura. One must be aware, however, of the possibility of bleeding tendencies which may occur in amyloidosis. These in turn have been ascribed to amyloid involvement of the blood vessels and in a few cases a deficiency of factor X [9, 10].

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References


RÉSUMÉ: Quoique l’amyloidose du tractus respiratoire soit bien reconnue, l’atteinte pleurale est très rare, avec seulement deux cas rapportés dans la littérature. Nous présentons un cas d’amyloidose primaire avec épanchement pleural et nous suggérons que l’atteinte pleurale et l’épanchement pleural devraient être ajoutés à la classification de l’amyloidose pulmonaire, et que l’amyloidose devrait être jointe à la liste des causes d’épanchement pleural.