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CASE STUDY

Pleural amyloidosis: thoracoscopic aspects

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Pleural amyloidosis: thoracoscopic aspects. F. Bontemps, I. Tillie-Leblond, M.C. Coppin, P. Frehart, B. Wallaert, P. Ramon, A.B. Tonnel. ©ERS Journals Ltd 1995.

ABSTRACT: We describe the case of a 60 year old man with primary amyloidosis, who suffered from peripheral neuropathy and cardiomyopathy and who presented with recurrent right pleural effusion. For this reason, he was admitted to hospital for thoracoscopic examination with pleurodesis.

Macroscopic examination of the parietal pleura revealed a diffuse inflammation and light brown deposits that where covered with nodules. Biopsy specimens confirmed the amyloid deposition.

This macroscopic appearance is described for the first time, and suggests that a local pleural synthesis of amyloid substance may occur during the course of systemic amyloidosis.

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Amyloidosis is characterized by the extracellular deposition of amyloid substance [1, 2]. In primary systemic amyloidosis, peripheral nerves, heart, skin, muscle and, less frequently, the pulmonary tract may be involved [3–5]. Tracheobronchial or parenchymal localizations have been described [6–8]. Pleural effusions rarely occur, and are often attributed to congestive heart failure, resulting from advanced amyloid cardiomyopathy. A local synthesis of amyloid substance has been suspected in some cases when the effusion was an exudate or when the presence of an abnormal protein was detected in the pleural fluid [9–12].

Case report

A 60 year old man was admitted to hospital for pleurodesis in January 1994. His grandfather, father, uncle and brother suffered from systemic amyloidosis. Therefore, in 1977 neuromuscular biopsy was performed on the patient, which revealed amyloid deposits. Symptoms began in 1978 with pedal paresthesia. In 1988, dyspnoea developed and congestive heart failure, attributed to amyloidosis (endomyocardial biopsy) was diagnosed. In 1991, a right pleural effusion appeared, without any clinical evidence of heart failure. Amyloid proteins were detected in the pleural fluid following staining with Congo red. Repeated thoracentesis was the indication for pleurodesis by thoracoscopy.

On admission, the patient complained of dyspnoea on moderate exertion. Physical examination revealed mild ankle oedema, dullness to percussion and decreased breath sounds in the right lower lung field. The pleural effusion was confirmed by chest roentgenogram. Arterial blood gas measurements on room air revealed pH 7.44, arterial carbon dioxide tension (P_{a,CO_2}) 4.8 kPa (36 mmHg), and arterial oxygen tension (P_{a,O_2}) 9.2 kPa (69 mmHg). Blood cell count, serum creatinine, serum electrophoresis, immunoelectrophoresis, complement components, antinuclear factor, rheumatoid factor, hepatitis B surface antigen, and $beta_2$-microglobulin were negative or in the normal range. Erythrocyte sedimentation rate was 40 $mm \cdot h^{-1}$. Serum immunoglobulins M, A, G and E (IgM, IgA, IgG and IgE) values were, respectively, 172, 316 (normal range 74–237 $kU \cdot L^{-1}$), 174 (normal range 90–167 $kU \cdot L^{-1}$), and 10 $kU \cdot L^{-1}$. Serum protein level was 59 $g \cdot L^{-1}$ (normal range 60–75 $g \cdot L^{-1}$) and serum albumin level was 26.8 $g \cdot L^{-1}$ (normal range 30–48 $g \cdot L^{-1}$). Lactate dehydrogenase (LDH) in serum was 201 $U \cdot L^{-1}$ (normal range 160–330 $U \cdot L^{-1}$). Liver functions showed elevated alkaline phosphatases (461 $U \cdot L^{-1}$, normal range 80–220 $U \cdot L^{-1}$), and aspartate aminotransferase 20 $U \cdot L^{-1}$. Cholesterol level in serum was 1.33 $g \cdot L^{-1}$ (normal range 1.5–2.4 $g \cdot L^{-1}$).

The electrocardiogram showed a diffuse microvoltage and an ST-segment depression in V5 and V6, and echocardiography revealed hypertrophied left and right ventricles with diffuse hypokinesia of the left ventricle, the ejection fraction being 55%.

Fibreoptic bronchoscopy was normal, as were the biopsies performed on the bronchial wall. Computed tomography (CT)-scan of the lung showed no parenchymal or mediastinal abnormality. Cytological analysis of pleural fluid showed 40% macrophages, 20% mesothelial cells, 20% lymphocytes and 20% non-leucocytoclastic neutrophils. Biochemical analysis of the pleural fluid showed protein 27 $g \cdot L^{-1}$, albumin 16.9 $g \cdot L^{-1}$, amylase 66 $U \cdot L^{-1}$, LDH 107 $U \cdot L^{-1}$ and cholesterol 0.41 $g \cdot L^{-1}$.

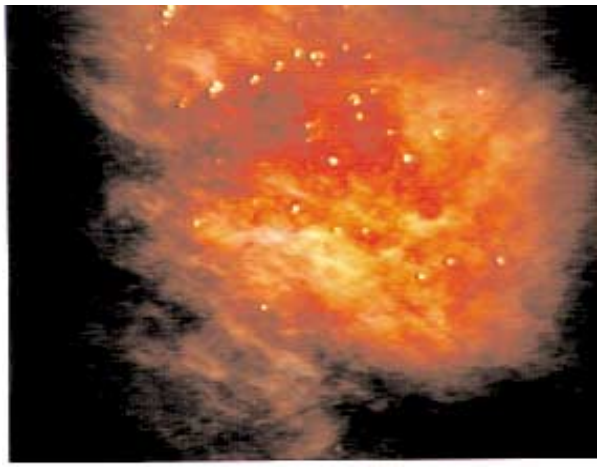

Fig. 1. – Thoracoscopic examination of the parietal pleura showing diffuse inflammation and light brown "nodules", measuring about 5 mm in diameter. (bar line = 2.34 cm).

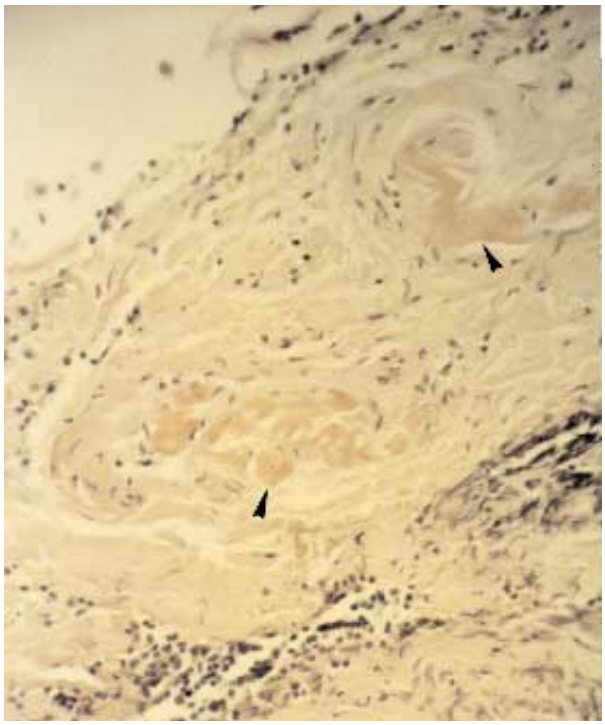

Fig. 2. – Histopathology of a pleural specimen showing amyloid deposition stained with Congo red (arrowheads), together with an inflammatory infiltrate. (Bar line = 41 μm, magnification ×100).

Thoracoscopic examination was performed under local anaesthesia. Macroscopic evaluation revealed a normal visceral pleura. The parietal pleura was covered with brown nodules of about 5 mm in diameter (fig. 1). The nodules were tough under the forceps. Biopsies were performed and the pathological examination revealed typical amyloid tissue. Staining with Congo red demonstrated an amyloid deposition, associated with an infiltrate of inflammatory cells, and pleural thickening (fig. 2). This typical aspect was confirmed by apple-green birefringence using polarization microscopy. Talc pleurodesis was efficient with no recurrence of pleural effusion so far.

Discussion

Pleural effusion is rare in systemic amyloidosis. The pathogenesis remains unclear and the amyloid deposition is not constant. This report deals with a patient suffering from amyloidosis with cardiomyopathy and amyloid deposition in the parietal pleura accompanied by a transudate. It assesses the roles played in the production of the pleural effusion by the congestive heart failure on one hand, and local inflammatory response to amyloid protein production on the other. The pleural effusion may result either from impaired fluid resorption (occlusion of the stomata by fibrin or amyloid proteins, pulmonary venous hypertension secondary to congestive heart failure), or from increased fluid production induced by inflammatory reaction [11–13].

In the case described by ASTOUL *et al*. [12] the lesions were disseminated on the parietal, visceral and diaphragmatic pleura, in a patient with no clinical evidence of heart failure. The diffuse aspect of the lesions indicated that the mechanism responsible was a local inflammatory reaction, secondary to amyloid deposition.

The individuality of our observation is the description of unusual macroscopic abnormalities, not previously described. Indeed, the thoracoscopic aspects showed brown nodules and inflammation, involving the parietal pleura (fig. 1). Since pleural fluid resorption is performed through the stomata of the parietal pleura, these findings favour primarily local production of amyloid substance, with deposition on the stomata leading to their occlusion. This may be responsible for the impaired fluid resorption and may, in part, explain the recurrence of the effusion. The fact that the effusion had not reappeared 6 months after pleurodesis as well as the lack of clinical evidence of heart failure indicated that cardiomyopathy did not play the major role in the production of the effusion. Moreover, the macroscopic abnormalities seen in this case may favour a local pathogenesis of the pleural amyloidosis. It would be interesting to perform "serum amyloid protein scintigraphy" to elucidate the exact pathogenesis of pleural amyloidosis [14].

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