Three Cases of Primary Pulmonary Amyloidosis

Takumi Yoshida, Akiho Obara, Kohei Yamauchi, Toshihide Nakadate, Akiko Shiba, Masayuki Ohura, Hiroshi Inoue and Nobukazu Tomichi*

In three cases of primary pulmonary amyloidosis the chief complaint was hemosputum. The diagnosis of amyloidosis was made using histochemical analysis of bronchial wall biopsy in all cases; multiple nodular lesions were observed in trachea and bronchi on flexible fiberoptic bronchoscopy. The surface of the tracheobronchial mucosa was smooth but bled easily. In one patient, chest X-ray film showed a solitary nodular shadow in the left lower lung field. These three cases were tracheobronchial amyloidosis, and one case was combined with nodular parenchymal type amyloidosis. (Internal Medicine 37: 687-690, 1998)

Key words: tracheobronchial amyloidosis, localized amyloidosis, nodular parenchymal amyloidosis

Introduction

Amyloidosis is characterized by the deposition of amyloid fibrils in various tissues, and is usually recognized by its staining reaction with Congo red and the subsequent demonstration of green dichroism. Amyloidosis in the lung is classified as primary pulmonary amyloidosis or a part of pulmonary amyloid deposition in systemic amyloid infiltration. Primary pulmonary amyloidosis is relatively rare. Lesser (1) reported the first autopsy case of amyloidosis localized to the lower respiratory tract in 1877. Recently, primary pulmonary amyloidosis is divided into tracheobronchial, nodular parenchymal and diffuse parenchymal types (2). In Japan, there were very few reports of amyloidosis localized to the lower respiratory tract. Here, we report three cases of primary pulmonary amyloidosis. The distribution of amyloid deposition in these three cases was tracheobronchial, and one case was combined with nodular parenchymal type.

Case Report

Case I

A 78-year-old Japanese male was admitted to Iwate Medical University Hospital with hemosputum and cough as the chief complaints. His history included surgical operation for laryngeal cancer, and he also presented with idiopathic thrombocytopenic purpura (ITP) and non-A and non-B type chronic hepatitis. In his blood test, the thrombocyte number was decreased due to ITP, and the values of glutamic oxaloacetic transaminase (GOT) and glutamic pyruvic transaminase (GPT) were slightly higher than normal, probably due to chronic hepatitis. There were no abnormal data in the urinalysis. Flexible fiberoptic bronchoscopy was performed to examine the origin of bleeding. Multiple coalesced nodules were revealed in the wall of trachea and bronchi. The bronchial lumen was irregular and narrowed on bronchoscopy (Fig. 1A). These nodular lesions were detected in the surface of trachea through bilateral main bronchi and intermediate bronchus. In the biopsy specimens of these nodular lesions, amyloid deposits were detected histochemically (Fig. 2A, B). The patient suffered from disseminated intravascular coagulation (DIC) and acute renal failure due to severe pneumonia during the treatment for ITP, and finally died. The amyloid deposits were examined in autopsy. They were observed in the trachea and limited areas of the bronchial wall, but were not observed in the pulmonary parenchyma and other organs. The type of amyloid deposition was tracheobronchial amyloidosis (Table 1).

Case II

A 75-year-old Japanese male was admitted to Iwate Medical University Hospital because of recurrent hemosputum. He had shown the same symptom 10 years before. He had smoked for about 50 years. His past history and family history were non contributory. Laboratory data on admission showed no abnor-
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Figure 1. Bronchoscopical findings in each case are shown. Multiple nodular lesions were observed at carina and bilateral main bronchi. The surface of the mucosa appeared smooth but bled quite easily (A). Irregular nodules and plaques, and vascularity were observed at the left main bronchus (B). Bronchial lumen appeared irregular and narrowed at the left 2nd carina (C).

mality of peripheral blood cell count, biochemistry, serology or urinalysis. No abnormal finding was observed in his chest X-ray films and computed tomography (CT) films. Flexible fiberoptic bronchoscopy was performed to examine the origin of the bleeding. The complete bronchial mucosa was injected and bled easily. There were discrete, occasionally pedunculated, irregular nodules with intact overlying mucosa. These nodular lesions extended from the left main bronchus into the left second carina (Fig. 1B).

Histologically, eosinophilic nodular deposits with foci of ossification were observed in the submucosa of bronchus (Fig. 2C). These deposits were positive for Congo red staining (Fig. 2D). In this case, the type of amyloid deposition was tracheobronchial amyloidosis (Table 1). After these examinations, no hemosputum was observed for 1 year in his clinical record as an outpatient.

Case III

A 78-year-old Japanese male was admitted to Iwate Medical University Hospital complaining of hemosputum. He had smoked for about 50 years. There was nothing else noteworthy in his past history. The breath sound was weak at the left back portion of the lung. On admission, there were no abnormal data in peripheral blood cell count, biochemistry, serology or urinalysis. The chest X-ray film showed a solitary nodular lesion, 5 centimeters in diameter in the left lower lung field and the margin of the left diaphragm and costophrenic angle were obliterated (Fig. 3).

Flexible fiberoptic bronchoscopy was performed to examine the origin of bleeding. Multiple nodular lesions were observed from the left upper lobe bronchus to the left lower lobe bronchus (Fig. 1C). Amyloid deposits were detected in the biopsy specimens of these bronchi and in the solitary nodular lesion in the left lower lung (Fig. 2E, F). In this patient, the type of amyloid deposition was tracheobronchial with nodular parenchymal type (Table 1). After these examinations, no hemosputum was observed for 5 years without therapy.

Discussion

Pulmonary amyloidosis occurs as a localized process restricted to the lung, or as a part of systemic infiltration. The term 'primary amyloidosis' has been applied to cases without accompanying diseases. The first report of amyloidosis in the lower respiratory tract was by Lesser in 1877. Subsequently, Spencer, Rubinow et al, Thompson and Citron and Chen et al reviewed localized respiratory amyloidosis and described the classification, prognosis, and management. The lung is frequently involved in all types of systemic amyloidosis. Primary amyloidosis limited to the lower respiratory tract is relatively rare. Primary pulmonary amyloidosis is traditionally divided into tracheobronchial amyloidosis, nodular parenchymal amyloidosis and diffuse parenchymal amyloidosis. The patients in the present paper correspond to tracheobronchial type (case I, case II and case III) and one of them was combined type, tracheobronchial with nodular parenchymal type (case III).

Hui et al described the chest X-ray findings of patients with primary pulmonary amyloidosis. The chest X-ray findings of the tracheobronchial type showed narrowing of the distal trachea, irregular surface and narrowing of main stem bronchi, and chronic atelectasis. The nodular parenchymal amyloidosis was well circumscribed. Most patients with pulmonary nodular amyloidosis had no symptoms, only an abnormal chest X-ray finding as the initial manifestation of disease. In the present paper, a pulmonary solitary nodular shadow in the chest X-ray film was observed in one case (case III). In this case, the solitary nodular shadow in the chest X-ray film was similar to that of lung cancer. We performed flexible fiberoptic bronchoscopy and lung biopsy in three cases. Multiple polypoid nodules were observed in the trachea and bronchi in each case. The wall of the trachea and bronchi appeared irregular and hemorrhagic, but the surface of the mucosa was preserved. In case II, the polypoid bronchial nodules were rough compared with the other 2 cases, because of calcification and osseous metaplasia that could be
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Figure 2. Bronchial biopsies. Submucosal deposits of amyloid were seen (Congo red, ×100) (A). The amyloid deposit showed apple green birefringence on polarization microscopy (Congo red stain, ×200) (B). Amyloid in bronchial mucosa. Eosinophilic nodular deposits with osseous metaplasia were found under the surface epithelium of bronchus (HE stain, ×160) (C). Congo red staining (×160) (D). Amyloid in pulmonary nodular parenchyma (HE stain, ×100) (E). Congo red staining in pulmonary nodular parenchyma (×100) (F).

observed right under mucosa. The clinical features, radiographic changes, and endoscopic appearance of pulmonary amyloidosis may be indistinguishable from those of endobronchial neoplasms and benign tumors (8, 9). In the present patients, it was difficult to make a differential diagnosis.

The cause of amyloidosis has not been elucidated. The protein of amyloid fibrils may be light chains synthesized by increased plasma cells and secreted extracellularly, cleaved by lysosomal enzymes in tissue macrophages, polymerized into AL amyloid fibrils, and then deposited (10, 11). In the present three cases, there were plasma cell infiltrations in the amyloid deposits. There have been very few reports of long-term obser-
Table 1. The Types of the Amyloid Deposition in Three Cases

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<th>Type of primary pulmonary amyloidosis</th>
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Figure 3. A 5 cm diameter tumor is observed in the left lower lung field.

vation of primary pulmonary amyloidosis. Further research is necessary to follow the clinical course of such patients with primary pulmonary amyloidosis.

References