Primary Diffuse Tracheobronchial Amyloidosis Treated by Bronchoscopic Nd-YAG Laser Irradiation

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A case of primary diffuse tracheobronchial amyloidosis in a 45-year-old woman is reported. Because of tracheal stenosis immediately beneath the vocal cords, due to amyloid deposits, she was treated with Nd-YAG laser irradiation. Immediately after treatment, symptoms such as cough, wheezing, and shortness of breath on exertion improved. Bronchoscopic Nd-YAG laser irradiation proved useful for the removal of amyloid deposits from the trachea in this patient.

Key words: Tracheal stenosis, Amyloid AA

Primary pulmonary amyloidosis is a very rare disease. Multiple or diffuse bronchial amyloidosis often leads to progressive airway obstruction. Thus, a rapid, effective, and safe treatment for maintaining airway patency is necessary. We describe a case in which bronchoscopic Nd-YAG laser irradiation was used to remove amyloid deposits safety and successfully.

CASE REPORT

On May 6, 1988, a 45-year-old housewife was admitted to our hospital because of progressive dry cough, wheezing, and shortness of breath for the previous 2 years. Physical examination at admission revealed inspiratory and expiratory stridor on auscultation of the chest, but was otherwise normal.

Pulmonary function tests showed a mildly reduced FEV\textsubscript{1,0} of 1.7 l (FEV\textsubscript{1,0\%} 62\%), a moderately reduced peak flow of 2.1 l/s, and a normal vital capacity of 2.7 l (% VC 103\%). Other data were as follows: hemoglobin 12.0 g/dl; white blood cell count 5,900/mm\textsuperscript{3} with a normal differential; platelet count 232,000/mm\textsuperscript{3}; serum total protein, 7.5 g/dl; albumin, 4.1 g/dl; a slightly elevated erythrocyte sedimentation rate, 12 mm/h. Serum electrolytes, blood urea nitrogen, creatinine, hepatic function profile, and urinary sediment were all normal. The arterial blood gas level in room air was as follows: pH, 7.4; PaCO\textsubscript{2}, 34 Torr; and PaO\textsubscript{2}, 92 Torr.

The chest X-ray appeared normal, but the chest CT scan (Fig. 1) demonstrated tracheal stenosis due to thickening of the tracheal wall. On bronchoscopy at admission (Fig. 2a), the larynx was shown to be normal, but the trachea was narrowed to less than 4 mm in diameter by numerous yellowish deposits just beneath the vocal cords. The distal portions of these deposits could not be observed due to the tracheal stenosis.

Biopsy specimens were obtained from these deposits during bronchoscopy. Bronchial biopsies showed infiltration with eosinophilic material that contained amyloid which stained with congo red and showed green birefringence under polarized light. Electron microscopic examination demonstrated amyloid fibrils (Fig. 3). Immunohistochemically, immunoreactivity with amyloid AA (Amyloid A, DACO Co., Ltd.) was observed in these fibrils;
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Fig. 1. Chest CT scan on admission showing evident wall thickening indicating stenosis.
(a) Level just under the vocal cords, (b) Subcarcina level.

therefore an early diagnosis of AA type amyloidosis was made. Gastrointestinal and rectal examination revealed no evidence of amyloidosis, and bone marrow aspiration disclosed no abnormality. A complete roentgenologic skeletal survey revealed no osseous abnormalities. The electrocardiogram was not remarkable. Thus, she was diagnosed as having primary diffuse tracheobronchial amyloidosis.

Because of exertional dyspnea, cough, and wheezing, removal of the amyloid deposits by bronchoscopic Nd-YAG laser irradiation (Laser Sonic, Model 6000; M & M Co., Ltd.) was performed. Amyloid deposits just beneath the vocal cords were irradiated four times to a total dosage of 2,251 J. Marked side effects such as infection or hemorrhage were not observed. After this therapy, bronchoscopy (Fig. 2b) showed that the tracheal diameter had widened, and disclosed an extensive lesion consisting of projecting deposits extending in an irregular fashion from the vocal cords down to the carina (which was considerably broadened) and into both main bronchi. Both main bronchi were severely narrowed by deposits. After this therapy there was symptomatic improvement, and pulmonary function tests showed an increase of FEV₁ to 2.1 L (FEV₁ 67%) and an increase in peak flow to 2.6 L/s. She was discharged on June 17, 1988, and two years after treatment she is well and free of symptoms.

DISCUSSION

Amyloidosis is a rare disease which may involve various organs of the body. Clinically, primary pulmonary amyloidosis was classified by Spencer (1) as:

I. A localized deposit in a bronchus.
II. Multiple or diffuse bronchial deposits.
III. Localized diffuse parenchymal deposits.
IV. Diffuse amyloid infiltration of the capillary walls and blood-vessels in the lungs.

Multiple or diffuse bronchial deposits as in the present case, often cause progressive airway obstruction and lead to distal atelectasis, secondary infection and respiratory failure. Various treatments such as pneumonectomy (2), radiotherapy (3), and intermittent bronchoscopic resection (4), have been reported, but none of them have proved to be successful. In addition, hemorrhage due to structural weakness of increased friability of arterial walls from amyloidosis involvement has been reported (5). From this point of view therefore, Nd-YAG laser irradiation was thought to be advantageous. In the present case, bronchoscopic Nd-YAG laser irradiation was performed safely and successfully, thus this procedure may be considered useful for maintaining airway patency in cases of tracheobronchial amyloidosis.

REFERENCES