We report on a 65-year-old female who complained of recurrent bronchopulmonary infections since 1999. She suffered from permanent cough and progressive dyspnea. The diagnosis of amyloidosis was made by bronchoscopic tissue biopsies, during which severe bleeding occurred. Argon-plasma-laser treatment stopped the bleeding and resulted in a successful recanalization of the left main bronchus. The patient noticed a decrease in dyspnea shortly after the intervention. Further diagnostic procedures did not show any signs of systemic or malignant disease. This led us to the diagnosis of a rare form of isolated tracheobronchial amyloidosis.

Key words: amyloidosis, endobronchial ultrasound, tracheobronchial amyloidosis

INTRODUCTION

Primary isolated tracheobronchial amyloidosis is a very uncommon disease. The clinical symptoms vary and include progressive dyspnea, cough, hemoptysis, and wheezing. Narrowing of the airways can cause atelectasis and recurrent pneumonia or bronchopulmonary infections. Solitary nodules or elongated airway narrowing may be mistaken for neoplasia. In this article we describe a localized form of primary tracheobronchial amyloidosis.
Case report

A 65-year-old non-smoking female complained of recurrent bronchopulmonary infections over the last six years. She suffered from permanent cough and progressive dyspnea. She was treated with inhaled bronchodilators, but the treatment was unsuccessful. The repetitive chest X-rays and CT-scans revealed a narrowing of the left main bronchus caused by circumferential thickening of the bronchial wall and atelectasis in the left upper lobe (Fig. 1 and Fig. 2). In view of the computed tomography findings, the patient was subjected to a fibreoptic bronchoscopy examination. After the first biopsy, a bleeding occurred. This made any further ambulant diagnostics impossible. Therefore the patient was admitted to our department. Physically examination and, particularly, auscultation of the chest were normal. Pulmonary function tests showed a mildly reduced FEV₁ of 1.5 l (64.8% of predicted), a moderately reduced peak flow, and an almost normal vital capacity of 2.2 l (76.3% of predicted). The airway dysfunction was partly reversible. All biochemical parameters and blood cells were within normal limits. The arterial blood gas levels in room air were as follows: PaCO₂ 40 Torr, PaO₂ 76 Torr, and pH 7.4.

Because of the bleeding the patient underwent rigid bronchoscopy with endobronchial ultrasound (EBUS). The examination showed an extreme stenosis of the left main bronchus by tumor-like, vulnerable tissue. In addition, there were white-yellowish, hyaline nodules at the main bronchial walls and in parts of the right upper lobe (Fig. 3). EBUS showed a thickening of the bronchial mucosa without infiltration of the surrounding tissue. The left part of the bronchial tree could not be evaluated due to the narrowing of the left main bronchus.

![Fig. 1. Chest CT scan showing airway narrowing and mural thickening of the left main bronchus.](image)
The diagnosis of amyloidosis was made by broncoscopically gained tissue biopsies. They showed infiltrations with eosinophilic material that contained amyloid which stained with Congo red and showed green birefringence under polarized light. The biopsy, again, caused a severe bleeding. Argon-plasma-laser treatment stopped the bleeding and resulted in a successful recanalization of the left main bronchus. The investigations for systemic involvement such as bone marrow aspiration, 24h urinary protein, and electrophoresis disclosed no abnormality. Neither the electrocardiogram nor ergometric and spiro-ergometric tests nor echocardiography were remarkable. There could not be found any other
organ dysfunction or a malignoma. Thus, the patient was diagnosed as having primary tracheobronchial amyloidosis.

The patient benefited from the laser treatment, as she noticed a decrease of dyspnea and cough shortly after the procedure. An increase in the FEV₁ to 1.7 l (73.4% of predicted) was measurable. She was discharged on September 15, 2005 and two years after the treatment she is still free of symptoms. In hindsight of the present diagnosis we affirmed that a CT imaging taken already in 2001 showed progressive airway narrowing, calcifications, and mural thickening of the left main bronchus; the three major consequences of amyloid infiltrations.

DISCUSSION

Pulmonary amyloidosis is a rare disease. It was found localized in the respiratory tract first by Lesser in 1877 (1). The most important pulmonary manifestations are tracheobronchial, nodular, and diffuse interstitial infiltrations (2-4). Although tracheobronchial amyloidosis is rare, it is still the most common manifestation of pulmonary amyloidosis (4). Clinical symptoms are usually similar to those caused by various airway diseases. The most common are dyspnea, cough, hemoptysis, and hoarseness (5). To prove the diagnosis, a tissue biopsy is essential.

Tracheobronchial amyloidosis typically presents after the fifth decade of life (2). It can be part of a systemic disease, but if it becomes symptomatic as a first manifestation of amyloidosis it is very often an isolated pulmonary form.

The treatment of the localized form is symptomatic only (4). Elongated stenosis can be reopened by repetitive laser therapy, balloon dilatation or stenting, all of which lead to good results in the majority of cases (4-7). Operative resection is rarely necessary. In case of localized AL (amyloid light chain) amyloidosis of the lung, radiotherapy can also be considered as a treatment option. Spontaneous regression has also been reported (8). As the disease usually progresses slowly and today most of the characteristic endobronchial changes can be treated with bronchoscopic techniques, the prognosis has improved over the last years.

Overall there have been very few reports of long-term observation of primary pulmonary amyloidosis and there is still no universally accepted treatment for the localized form. A definitive treatment does not always seem to be necessary.

REFERENCES


Author address: G. Laier-Groeneveld, Evangelisches und Johanniter Klinikum Niederrhein GmbH, Bronchial und Lungenheilkunde, Steinbrinkstrasse 96a, 46145 Oberhausen, Germany.