Isolated Amyloid Tumor in the Mediastinum: Report of a Case

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Abstract
Primary amyloidosis isolated in the mediastinum is rarely encountered in thoracic surgery and few such cases have been reported. We present a case of primary isolated hilar amyloidosis of the mediastinum to illustrate the difficulties in differentiating this disorder preoperatively from central bronchial carcinoma, carcinooid tumor, and mediastinal lymphoma. Usually, a definitive diagnosis can only be made by open biopsy during thoracoscopy or thoracotomy. In conclusion, amyloidosis should be considered in the differential diagnosis of patients when calcifications are found, bearing in mind that radiologic findings are inconclusive and transbronchial biopsy can be negative.

Key words Thoracic surgery · Mediastinal tumor · Amyloidosis

Introduction
Unilateral hilar amyloidosis of the mediastinum is an extremely rare condition1-5 and is therefore not often considered in the differential diagnosis of patients with respiratory symptoms. Despite extensive diagnostic workup, including imaging and biopsy, isolated mediastinal amyloidosis is usually only diagnosed by histologic examination of a specimen obtained by open biopsy during thoracotomy or thoracoscopy. Amyloidosis should be considered in the differential diagnosis of patients with respiratory symptoms but inconclusive radiologic findings such as calcifications, even when a transbronchial biopsy is negative.

Case Report
A 61-year-old man was hospitalized for right chest pain and pneumonia. He was otherwise in good general health and well nourished. Coarse bibasilar rales were heard on auscultation of the lungs but lung function parameters and laboratory values were within the normal range. Chest X-ray showed a right hilar density containing scattered calcifications (Fig. 1), which led us to suspect central bronchial carcinoma in the right lung. Computed tomography (CT) of the chest demonstrated a soft-tissue mass of 8–10 cm in size with calcifications in the mediastinum, but no intrapulmonary nodules were seen. There was also constriction of the pulmonary artery trunk. Virtual bronchoscopy demonstrated an intrabronchial tumor located in the upper lobe bronchus (Fig. 2). Noncontrast CT of the chest showed a calcified mediastinal tumor protruding into the right main bronchus (Fig. 3). Fiberoptic bronchoscopy confirmed an infiltrating mass in the right upper lobe bronchus. Microscopic examination of the biopsy material showed pronounced regressive alterations, which did not allow for morphologic correlation. Although there were no signs of malignancy, a thoracotomy was performed due to a strong suspicion of malignancy in the upper lobe bronchus.

A posterolateral thoracotomy was performed through the fifth intercostal space on the right side. After opening the pleural cavity, a hard and nodular baseball-sized tumor was immediately seen. The tumor surrounded the entire pulmonary hilum with extensive infiltration of the mediastinum. Dissection in the middle posterior mediastinum revealed that the mass extended to around the aortopulmonary window on the left side. Cranially, paratracheal infiltration was seen in the anterior upper mediastinum and around the right pulmonary artery trunk, which was completely encased. The mass was yellow-white and contained calcified structures. A large biopsy specimen was excised. The second
and third segments of the upper lobe were partly carniﬁed as a result of chronic atelectasis, but otherwise, the parenchyma of the right lung unfolded with ventilation. Complete resection of the tumor was not possible due to its extensive growth. The intraoperative findings suggested a T4-bronchial carcinoma. The postoperative course was without complications.

Histology

Examination of frozen and parafﬁn-embedded sections revealed tumor-like, localized deposits of amorphous, faintly eosinophilic material, that were PAS-positive and Congo red-positive, showing an “apple-green” birefringence under polarized light. These deposits were surrounded by histiocytes, multinucleated giant cells, and more or less prominent chronic inﬂammatory inﬁltrate containing polyclonal lymphocytes and plasma cells. There was no evidence of immaturity or cellular atypia. Similar deposits were seen in interspersed walls which were partially and diffusely thickened. Focal microcalcifications and metaplastic bone formation were also identiﬁed. Thus, the diagnosis of an amyloid tumor was conﬁrmed.

Discussion

Pulmonary amyloidosis is not often considered in the differential diagnosis of patients with respiratory symptoms because of its rarity. Furthermore, primary amyloidosis can only be diagnosed after the secondary form of amyloidosis has been excluded. Basically, three types of thoracic amyloidosis have been deﬁned: tracheobronchial, nodular, and diffuse amyloidosis. The tracheobronchial type is characterized by the massive deposition of amyloid in the bronchial wall, which leads to postobstructive pneumonia as a result of bronchial stenosis. According to Glenner, this is the most common type of pulmonary amyloidosis. In the nodular type, amyloid accumulates in the lung parenchyma in