Case Report

Management of Tracheobronchial Amyloidosiis With Therapeutic Bronchoscopic Techniques

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ABSTRACT

Amyloidosis is a systemic disease caused by abnormal deposition of amyloid material that is detected with Congo red staining and is difficult to diagnose. Involvement of the tracheobronchial tree is rare and is a challenge for pulmonologists because of the wide differential diagnosis of this disease. We present two cases where tracheobronchial affection has been observed: in one of them as a primary disease, and in another as secondary affection. The use of bronchoscopic techniques is essential for the diagnosis of tracheobronchial involvement. In the absence of an effective drug therapy, local management of this disease with endoscopic techniques for bronchial repermeabilization is able to provide clinical improvement and expand the treatment options and prognosis in this disease.

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RESUMEN

La amiloidosis es una enfermedad sistémica producida por el depósito anómalo de material amiloido; tiene la peculiaridad de detectarse con la tinción rojo Congo y es de difícil diagnóstico. La afectación del árbol traqueobronquial es muy poco frecuente y constituye un reto para el neumólogo debido al amplio diagnóstico diferencial de esta enfermedad. Se presentan 2 casos en los que se ha objetivado la afectación traqueobronquial: en uno de ellos como enfermedad primaria y en otro como afectación secundaria. El uso de técnicas broncoscópicas es primordial para el diagnóstico de la afectación traqueobronquial. En ausencia de un tratamiento médico eficaz, el manejo local de esta enfermedad con técnicas endoscópicas de repermeabilización bronquial consigue una mejora clínica y amplía las opciones terapéuticas y pronósticas en esta enfermedad.

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Introduction

Amyloidosis is a systemic disease of unknown origin which is characterized by extracellular deposits of proteins with a specific structural conformation that gives them a characteristic apple-green birefringence when stained with Congo red. They are deposited mainly in the heart, kidneys and liver. Diseases caused by abnormal amyloid deposits can be classified into primary or secondary, local or systemic, based on their distribution. Therefore, pulmonary involvement by amyloidosis can be both secondary and primary, systemic or localized. It has an estimated incidence of between 5 and 10 cases per million persons/year. However, primary pulmonary involvement is much rarer,1 and presents a challenge for the pulmonologist, as the use of bronchoscopy techniques is essential for diagnosis and to provide optimal treatment.

Cases

Case 1

A 46-year-old male who attended our clinic due to unproductive cough, moderate effort dyspnea of recent onset and temperature of 38 °C. His only history was that he smoked 45 packs/year, with no symptoms of chronic bronchitis. On physical examination, he had baseline oxygen saturation measured by pulse oximetry (SpO₂) of 93% on room air, and ronchi in both hemithorax on pulmonary...
diagnosis of localized primary tracheobronchial amyloidosis was established.

Case 2

57-Year-old woman with repeated episodes of hemoptysis with no other symptoms. She had a history of hypertension for which she was on treatment with valsartan, chronic nasal obstruction and no toxic habits. The physical examination was normal, as were additional tests performed: laboratory tests and chest X-ray. A bronchoscopy was carried out, detecting multiple friable, bleeding nodules on the anterior and posterior side of the trachea, one of which was ulcerated (Fig. 2). The histopathology study demonstrated the presence of eosinophilic amorphous material that exhibited birefringence with polarized light, corresponding to amyloid material. An extension study of the disease was carried out, in which amyloid material was also detected in the subcutaneous fat and bone marrow biopsy. The patient was diagnosed with primary systemic amyloidosis with lung and bone marrow involvement.

Discussion

Amyloidosis is a rare disease of unknown etiology characterized by abnormal deposition of fibrillar proteins that typically exhibit apple-green birefringence under polarized light after Congo red staining. Involvement is variable, and there may be primary, secondary or senile disease. It may be systemic or well localized in an organ or tissue. Pulmonary involvement by amyloid is exceptional, whether it forms part of a systemic disease or is localized. Five forms of primary pulmonary amyloidosis have been described: tracheobronchial, nodular (solitary or multiple), senile pulmonary, mediastinal-hilar and diffuse interstitial. Tracheobronchial amyloidosis is the most common presentation, and is characterized by amyloid deposits in the tracheobronchial tree only. Amyloidosis affects mainly males (2:1) and in middle age (50–60 years). The clinical presentation, in the case of systemic disease, varies depending on the organs involved. In the case of pulmonary involvement, cough (74%), audible wheezing (70%), dyspnea (60%), hemoptysis (50%) and stridor (30%) are more common. More than half of cases may present as obstructive pneumonia, bronchiectases or atelectases, so its identification poses a major
The patient in case 1 was treated on two occasions with Nd:YAG laser to repermeate the airways (Fig. 1B), owing to the stenosis that he presented due to amyloid infiltration, achieving clinical and functional improvement. With respect to the patient in case 2, she received treatment with a combination of prednisone and melphalan, with a good response and partial resolution of the tracheal lesions, with no functional or symptomatic impairment.

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**Conflict of Interest**

The authors declare that there is no conflict of interest.

**References**