Caso Clínico
Clinical Case

Sandra Saleiro[1]
Venceslau Pinto Hespanhol[2]
Adriana Magalhães[3]

Amiloidose traqueobrônquica primária – A propósito de dois casos clínicos

Primary tracheobronchial amyloidosis – Two case reports

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Resumo

A amiloidose traqueobrônquica primária é uma forma de amiloidose respiratória, caracterizada pela presença de depósitos insolúveis de proteína fibrilar amilóide, ao longo da parede das vias aéreas. É uma doença pouco frequente, sendo necessário uma amostra de tecido para estabelecer o diagnóstico definitivo, com base em características histológicas específicas.

Os autores relatam dois casos clínicos de amiloidose traqueobrônquica, descrevendo os seus sintomas e os procedimentos diagnósticos e terapêuticos que foram efectuados.

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Palavras-chave: Amiloidose, traqueobrônquica.

Abstract

Primary tracheobronchial amyloidosis is a form of respiratory amyloidosis, characterised by insoluble amyloid fibril proteins deposits along the airways wall. It is an uncommon disease, requiring a tissue sample to establish the definite diagnosis based on specific pathological features.

The authors report two cases of tracheobronchial amyloidosis, describing their symptoms and the diagnostic and therapeutic procedures that were performed.

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Key-words: Amyloidosis, tracheobronchial.

[1] Interna Complementar de Pneumologia
[2] Chefe de Serviço de Pneumologia; Director do Serviço de Pneumologia do Hospital de São João; Professor Agregado da Faculdade de Medicina da Universidade do Porto
[3] Assistente Hospitalar Graduada de Pneumologia

Serviço de Pneumologia – Hospital de São João
Director de Serviço: Prof. Dr. Venceslau Pinto Hespanhol

Endereço:
Hospital de São João
Alameda Prof. Hernâni Monteiro
4202-451 – Porto – Portugal

Contacto:
E-mail: sandrasaleiro@portugalmail.pt

Introduction

Amyloidosis includes a group of diseases characterised by the extracellular deposit of fibrillar protein in one or more organs. Amyloidosis can be localised or systemic, according to the anatomic involvement, and primary or secondary, in this last case if associated with chronic inflammatory or infectious diseases or neoplasms. Primary respiratory amyloidosis is uncommon and can involve the tracheobronchial tree (in a focal or diffuse way) or the pulmonary parenchyma (as multiple nodules or diffuse interstitial infiltrate).

In tracheobronchial amyloidosis, symptoms such as dyspnoea, cough, hoarseness, haemoptysis, wheezing or stridor can be present, due to bronchial obstruction.

The authors describe two cases of localised tracheobronchial amyloidosis with distinct clinical features and management approach.

Case 1

A 68 year-old female nonsmoker presented with hoarseness, stridor and progressive dyspnoea for the last four months. The patient had cleft palate and a past history of arterial hypertension and cerebrovascular disease, with no neurological sequelae. She had worked as a dressmaker, and retired 3 years ago. On physical examination, she had respiratory distress, with stridor audible over the trachea. At laryngeal observation, oedema and irregularity of glottic and infra-glottic region were recognised. Full blood count, biochemical and arterial blood gases (while breathing room air) analysis were within normal values. Chest radiography showed no abnormalities. Computed tomography of the chest showed circumferential wall thickening of the trachea and the main bronchi, with no relevant lung parenchyma abnormalities (Fig. 1). Pulmonary function tests revealed the following values (as % predicted): FVC: 101%, FEV1: 87%; FEV1/FVC: 71%; PEF: 48%; FEF 25-75: 58%; FEF 50: 53%; FEF 75: 76%; VC: 96%; TLC: 87%; RV: 86%; RV/TLC: 41%.

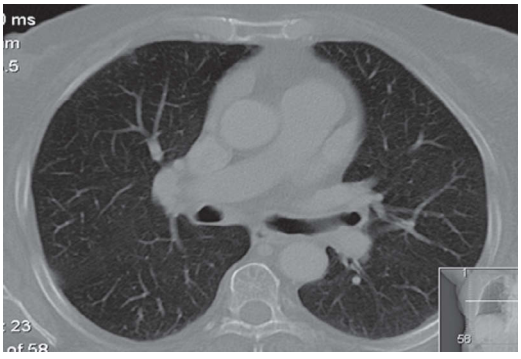

Fig. 1 – Thoracic CT scan: circumferential wall thickening of the main bronchi

Subsequently, the patient underwent fiberopticbronchoscopy, which documented, besides great deformation of ari-epiglottic folds and glottis, mucosal irregularity along the trachea and both proximal bronchial trees. There were several mucosal and submucosal yellowish lesions, some of which were flat and some were projecting from the tracheal wall to the tracheal lumen. Since two mucosal prominences, most evident in infra-glottic region, determined significant obstruction, the patient was submitted immediately to rigid bronchoscopy. During this procedure, mechanical dilatation was performed, followed by insertion of a stent (*Dumon*, 16/30 mm) in the upper tracheal portion. At this time, biopsies from the infra-glottic lesions were obtained. When she