Tracheal amyloidosis

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CASE PRESENTATION

A 62-year-old woman was referred to respiratory for a tracheal mass on computed tomography scan. She experienced cough six months before presentation; however, at the time of assessment, there was no fever, weight loss, cough, hemoptysis or dysphagia.

Her medical history was unremarkable. She worked as a housekeeper and denied any relevant occupational exposures. She was a 20 pack-year smoker and had quit one year previously. Physical examination was unremarkable, with no evidence of cachexia, hypoxemia, clubbing, stridor, dysphonia, heart failure, rash or focal neurological findings.

Computed tomography imaging revealed nodular thickening of the trachea and focal bronchiectasis in the lingula and right middle lobe (Figure 1). There was no evidence of parenchymal lung disease. Spirometry was normal, with no evidence of central or small airway obstruction.

Bronchoscopy revealed sessile masses along the cartilaginous walls of the trachea and mainstem bronchi (Figure 2). The lesions were gelatinous and, while they appeared friable, did not bleed. Biopsy revealed fibrillar material that stained positive for Congo red and demonstrated apple-green birefringence under polarized light consistent with amyloid.

Assessment by hematology ruled out systemic or other organ-specific amyloidosis. Over the past 1.5 years of follow-up, the airway lesions have been intermittently debulked by interventional pulmonology.

Figure 1 Computed tomography thorax images showing endoluminal lesions involving the distal trachea and right main stem bronchus (arrows)

Figure 2 Amyloid plaques before manipulation in the distal trachea (A) and proximal right mainstem bronchus (B) (arrows). The same right mainstem lesion is seen following gold probe cautery (C) (arrows)

KEY LEARNING POINTS

- Amyloidosis can be primary or secondary (ie, multiple myeloma) (1).
- Most cases are sporadic and can be either systemic or organ specific. However, a minority is hereditary in nature and these cases tend to have systemic involvement. Any part of the respiratory tract can be involved (2).
- Tracheobronchial amyloidosis is usually not associated with parenchymal (interstitial) or systemic involvement and, therefore, has a better prognosis (2).
- In proximal airway disease, spirometry is only affected once airway diameter is <8 mm (3).
- If the distal airways are involved, spirometry often reveals airflow obstruction (4).
- Lesions should only be debulked once they cause symptoms (5).

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