Primary tracheobronchial amyloidosis: Commonly uncommon?

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Primary tracheobronchial amyloidosis has been described as a rare condition in which amyloid deposits build up in the airway mucosa. It was first reported more than 50 years ago. Aside from the lungs, the other organs are spared from the disease, in contrast to systemic amyloidosis. Clinical presentation is highly nonspecific and varies in the degrees of wheezing, shortness of breath and cough. With the increased use of flexible bronchoscopy and computed tomography imaging in the past 20 years, the number of reported cases has increased significantly (1). However, we must ask: is the disease more prevalent, or is it simply more frequently diagnosed?

In the current issue of the *Canadian Respiratory Journal*, three different cases of primary tracheobronchial amyloidosis are reported (2-4).

Clinical presentation varies from chronic cough to shortness of breath, hemoptysis and even stridor. The final diagnosis is made during bronchoscopy, which includes bronchial biopsies revealing positive Congo red staining (5). In addition to macroscopic findings on bronchoscopy, involvement of the membranous trachea and calcification of the airways on computed tomography are useful findings that suggest the presence of the disease (6).

The treatment of primary tracheobronchial amyloidosis remains a challenge. The treatment most often reported in the literature is mechanical debulking. Improving airway patency can be performed using rigid or flexible bronchoscopy, in addition to cautery, laser or forceps. It is reserved for symptomatic cases in which there is a high frequency of chronic recurrence. The prognosis remains more favourable than for systemic amyloidosis. Recently, the use of radiation therapy of the airway has shown encouraging results in the treatment of the disease (7). In the current issue of the *Journal*, Numbere and Grayez (4) (pages 273-275) showed significant improvement following 20 Gy of radiation in 10 fractions, and the patient remained asymptomatic 14 months after therapy.

It is possible that tracheobronchial amyloidosis is more prevalent than previously believed because it was underdiagnosed. Clinicians should be aware of it. In patients with refractory asthma or chronic cough of unknown etiology, a more extensive workup, including

flexible bronchoscopy and computed tomography of the chest, would enable the identification of rare diseases including primary tracheobronchial amyloidosis.

It is also important to communicate adequately with the pathologist. If the macroscopic appearance of the mucosa on bronchoscopy and computed tomography is suspicious for amyloid deposits, the pathologist should be made aware so that adequate staining can be performed on the pathology specimen. A multidisciplinary approach, including radio-oncology, oncology, interventional respiratory, radiology and pathology, is necessary to offer optimal therapy. Systemic involvement of other organs must be ruled out.

More research needs to be performed to understand the pathophysiology of the disease to develop a more effective treatment. The first step remains to diagnose the disease more accurately and to report it in the literature so that all clinicians can benefit from learning about these cases. Primary tracheobronchial amyloidosis could become an uncommonly common disease.

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