Case report

Tracheobronchopatia osteochondroplastica (TPO) associated with tracheobronchial amyloidosis (TBA)

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\section*{ABSTRACT}

Tracheobronchopatia osteochondroplastica (TPO) is an idiopathic disease involving the cartilage rings of the large airway, characterized by submucosal calcified nodules. Localized tracheobronchial amyloidosis (TBA) is another rare disease with localized amyloid deposits in the tracheobronchial tree. The two diseases rarely coincide, and only a few case reports and series have been reported.

A patient with dyspnea was referred to our clinic for suspicion of TBA. Chest computed tomography (CT) scan showed marked thickening of the tracheobronchial wall with calcified endobronchial submucosal nodules. The nodules were resected with a Diode Laser under rigid bronchoscopy, and results from the biopsy showed both osteochondroid metaplasia on microscopy in Hematoxylin and Eosin staining and apple-green birefringence on polarized microscopy in Congo red staining. This is a rare case in which microscopic findings of both TPO and TBA were observed on one slide. These findings suggest that localized TBA could be a cause of TPO.

1. Introduction

Tracheobronchopatia osteochondroplastica (TPO) and primary tracheobronchial amyloidosis (TBA) are two rare diseases. Sometimes it is reported that the two diseases are diagnosed in one patient, but tissue confirmation is usually not done for both of the entities. This case report provides a pathology slide with both of the diseases.

2. Case presentation

A 52-year-old patient visited our hospital with dyspnea on exertion which had started three years previously and worsened over the previous three weeks. Three years ago when the symptom started, he had visited a local clinic and undergone bronchoscopy which showed bronchial narrowing. At that time, he received no treatment.

He had type 2 diabetes mellitus and took oral hyperglycemic agents (metformin 1000mg, glimepiride 4mg, and acarbose 100mg twice daily). His hemoglobin A1c level was 7.1%. He was a 17.5-pack-year current smoker. He had undergone a cranial operation due to trauma. His father had a history of tuberculosis, and his mother had diabetes mellitus.

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He was diagnosed with TPO associated with TBA and started taking methotrexate once weekly. His lung function improved: forced expiratory volume in 1 second (FEV1) increased from 2.03 L to 2.24 L after rigid bronchoscopy laser resection. After following up for 3 years with methotrexate maintenance, his symptoms and pulmonary function were stable and the patient was referred back to the local clinic.

3. Discussion

Tracheobronchopathia osteochondroplastica (TPO) with primary tracheobronchial amyloidosis (TBA) is very rare. One study of 41 patients with TPO demonstrated that TBA confirmed by Congo red staining was observed in only 2/16 specimens [1]. The coincidence of these two rare diseases occurring simultaneously was first documented in 1968 [2]. The necropsy of a patient showed extensive amyloid infiltration in the submucosa and islands of bony deposits along cartilaginous rings on microscopy as well as a “cobblestone appearance” of the trachea on bronchoscopy. The authors postulated that the advanced ossified stage of primary TBA may be TPO.

There are only a few reports successfully demonstrated the coexistence of TPO and TBA with Hematoxylin and Eosin (H&E) and Congo red staining [3,4]. In some studies, amyloidosis was confirmed by Congo red staining, but TPO was not proved by pathology because the lesion spared the posterior membrane of the trachea. However, some cases of localized amyloidosis could involve only the anteromedial aspects of trachea. Therefore, pathologic confirmation is needed for diagnosis of TPO and TBA.

Regarding pathogenesis, amyloid fibrils have affinity for calcium, and amyloid deposits frequently accompany calcification and ossification [5]. It could be suggested that TBA is one of the causes of TPO, which is characterized by calcification and ossification in the submucosa.

In conclusion, chest CT and bronchoscopy are helpful for diagnosis of TPO and TBA, although definitive diagnosis is a pathological diagnosis. The present case demonstrated the coexistence of osteochondrosis and amyloidosis in the submucosa, suggesting their association in the pathogenesis.

Declaration of competing interest

The authors have nothing to disclose.
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