An Overlooked Airway Disease

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Explanation:
Therapy for tracheobronchomegaly is essentially supportive1. Other than tobacco abstinence and immunization, there is no specific preventive therapy in asymptomatic individuals. Intensive respiratory rehabilitation and appropriate antibiotics during infectious exacerbations are the mainstay of treatment 2. Inhaled bronchodilators and corticosteroids are ineffective except in the aforementioned exacerbation 2. In acute settings, Continuous positive airway pressure (CPAP) can sustain an open airway and ease secretion drainage. Patients may use intermittent CPAP as longterm therapy. However, CPAP does not appear to have a longterm influence on dyspnea or cough.Mucous plugging removal can be facilitated by bronchoscopy and /or tracheostomy. Surgical therapy rarely has a role due to the diffuse airway involvement. Nevertheless, airway stenting and tracheobronchoplasty have been recently considered in patients with this disorder.
Discussion:
MounierKuhn syndrome (MKS), or tracheobronchomegaly (TBM) is a rare entity, with less than a few hundred reported cases 1, 2. Most of these cases were described in middleaged men (75% >29 years old). However, it is likely that the true prevalence may be higher, as cases have been overlooked due to the overlapped respiratory manifestation with other common airway diseases.
The etiology remains unknown. Atrophy or absence of elastic fibers and thinning of the

muscular mucosa and tracheal cartilage are the main features with subsequent outpouching of the membranous and cartilaginous portions of the trachea and main bronchi. The patient eventually develops broad airways diverticulosis and secondary collapse of the herniate tissues between the cartilaginous rings 2. Ultimately, secretions are retained in dilated airways. Hence, lower respiratory tract infections occur, owing to the insufficient coughing mechanisms and deficient ciliary clearance. Subsequent emphysema, large bronchiectasis and parenchymal fibrosis may complicate the condition. Notably, airways beyond the fourth and fifthorder divisions are commonly spared.

The majority of cases are sporadic. Nevertheless, tracheobronchomegaly can be associated with rare genetic and connective tissue disorders, such as BrachmanndeLange syndrome, EhlersDanlos, Cutis Laxa, Marfan syndrome, ataxiatelengiectasia, relapsing polychondritis, rheumatoid arthritis, generalized elastolysis, ankylosing spondylitis, and KenneyCaffey syndrome. Immune deficiency diseases, such as Bruton–typeagammaglobulinemia have also been associated with tracheobronchomegaly. Other variants can be found in individuals with prolonged intubation and severe pulmonary scarring secondary to advanced sarcoidosis, cystic fibrosis or allergic bronchopulmonaryaspergillosis2 .

Tracheobronchomegaly has been classified in 3 different subtypes based on the anatomical involvement1. Type 1 is described as a symmetrical and diffuse enlargement of trachea and major bronchi. Type 2 presents as airway enlargement with bizarre diverticulosis, whereas in type 3, the airway dilatation extends to the peripheral airways.

Diagnosis of is usually made by imaging and bronchoscopy with dynamic assessment. Specifically, tracheal, right main bronchus, and left main bronchus diameters that exceed 3.0 cm, 2.4 cm, and 2.3 cm, respectively, are consistent with tracheobronchomegaly.

References:

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