htd
hrvatsko torakalno društvo

TORAKS 2019

9. Kongres Hrvatskog torakalnog društva
9th Congress of Croatian Thoracic Society

Hotel Westin Zagreb
10.–13. 4. 2019.

MOUNIER KUHN SYNDROME – TRACHEOBRONCHOMEGALY AS A CAUSE OF BRONCHIECTASIS IN ADULT

NEDELJKOVIĆ V.[1], Markelić I.[2], Petrović Vlahović A.[3], Hećimović A.[2], Vukić Dugac A.[2]

[1] Ordinacija opće medicine, Zagreb, Croatia
Ordinacija opće medicine

[2] KBC Zagreb, Zagreb, Croatia
Klinika za plućne bolesti Jordanovac

[3] Specijalna bolnica za plućne bolesti Rockefellerova, Zagreb, Croatia
Pulmologija

Objective: Mounier-Kuhn syndrome (MKS) or tracheobronchomegaly is a rare disorder of unknown origin characterised by thinning of muscular mucosa and atrophy of longitudinal muscular and elastic fibres of tracheal wall and main bronchi, often with a finding of diverticuli between cartilages. The hallmark of this condition is a dilatation of the main airways. The size of trachea is determined by sex, not by body height and mass. The criteria for tracheobronchomegaly are diameters that exceed 30 mm for trachea, 20 mm for right main bronchi and 23 mm for left main bronchi. Men are mostly affected (8:1 compared to women) and it occurs at the age of 56. This syndrome is often associated with severe bronchiectasis and it can resemble COPD, with which it often overlaps and the diagnosis is set radiologically, usually with a CT, which is a golden standard.
Most common clinical presentation is non-specific and involves recurrent respiratory infections with dry or productive cough, purulent sputum, dyspnoea and haemoptysis.
Our patient is a 53-year-old man, smoker with 66 pack years who has suffered from recurrent pneumonias with the clinical presentation of COPD exacerbation for the last 4 years. With no impressive past medical history, he was admitted to the hospital 2-3 times annually due to breathing difficulties with mucoid expectoration which

became purulent during infectious exacerbations. He had fever with occasional pain in his left hemithorax. Lung auscultation revealed bronchial breathing bilaterally, and other physical findings were unremarkable. His ABS showed partial respiratory insufficiency and CRP and SR were elevated. During patients' treatment, chest X-rays, spirometry, sputum cytology and culture, bronchoscopy and MSCT were done. A chest radiography suggested emphysema with bronchiectasis while the MSCT showed tracheobronchomegaly with the transverse diameter of trachea being 33 mm, right main bronchi 21 mm, and left main bronchi 23 mm.
On bronchoscopy all air pathways seemed dilated. Patients spirometry showed obstruction characteristic for COPD: (forced expiratory volume in one second) FEV1 46.5%, (forced vital capacity) FVC 68%, FEV1/FVC 55.16%, (maximal (mid-)expiratory flow) MEF50 25%, and negative bronchodilator test. Patient was treated with antibiotics, bronchodilators and supportive therapy which accomplished clinical response and improvement of his condition.
MKS can easily be misdiagnosed. In patients with remittent respiratory infections, chronic cough and occasional blood-stained sputum it should be considered as an uncommon cause of bronchiectasis. Therapy is generally supportive, and is directed to minimize the damage from stasis and infections.