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Case Report

Can Intubate but Not Ventilate Nightmare – Mounier-Kuhn Syndrome- Case Report

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ABSTRACT

The Mounier-Kuhn syndrome (MKS) or tracheobronchomegaly (TBM) is a rare condition of unknown frequency, only 300 cases have been reported [1,2]. It is characterized by significant expansion of the trachea and bronchi and recurrent respiratory infections [3,4]. Our patient was a 60-year-old male with pneumonia, admitted to ICU due to the urge for mechanical ventilation. Ventilation by mask was normal, he was intubated without any difficulty, but mechanical ventilation was impossible due to enormous leakage, bag valve ventilation was only option to deliver any tidal volume. Bronchoscopy revealed the collapse of the distal trachea and the left main bronchus, mucous plaque was removed. Tamponade of the larynx reduced the leakage. MSCT of the neck was made, tracheomegaly and bronchomegaly were described; Mounier Kuhn syndrome was suspected. Further bronchoscopy revealed mucoid impactions filling the bronchi of the lower right lobe. He was weaned and extubated, but oxygenation worsened despite of HFNC, so he was reintubated with larger tracheal tube. Further ventilation was carried out without leakage, a percutaneous tracheotomy was performed 15 days later and he was gradually weaned from mechanical ventilation.

KEYWORDS

Tracheomegaly, Tracheobronchomegaly, Mounier-Kuhn syndrome.

Introduction

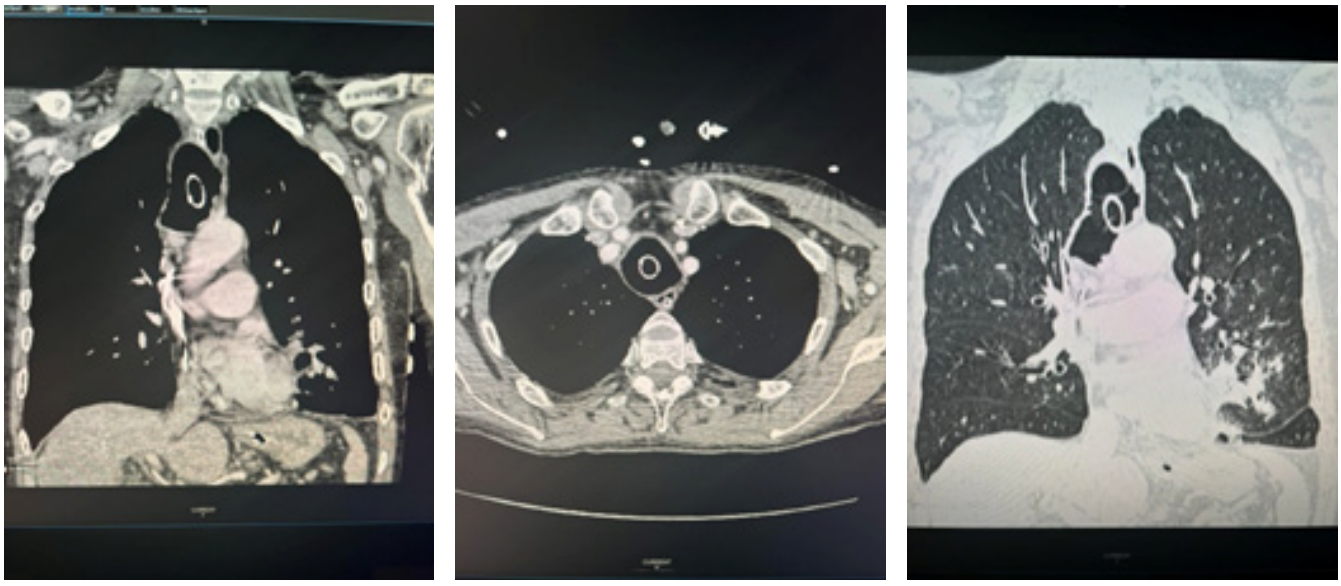
Mounier-Kuhn syndrome or tracheobronchomegaly is a rare disorder characterized by marked dilatation of the trachea and main bronchi, bronchiectasis, and recurrent respiratory tract infections. Its clinical presentation may vary and mimic a variety of disorders.

Case Report

A 60-year-old male presented in hospital with pneumonia, soon upon admission his respiratory status worsened so he was admitted to ICU. He is a former drug addict, HepC positive, 15years ago he had amputation of lower leg as consequence of trauma. He is protégé of a nursing home, a smoker and has a permanent urinary catheter. Upon admission to ICU patient is dyspnoic, SpO_2 rises till 84% despite all measures, mechanical ventilation was initiated. Patient was normally ventilated by mask, intubation procedure was standard, without airway abnormalities (Cormack Lehane 1), but mechanical ventilation was impossible, only way to deliver any tidal volume was by bag valve mask, but with a lot of resistance and the most of the air came out through the mouth. The tube was exchanged but the problem maintained. An urgent bronchoscopy was performed, the collapse of the distal trachea and the left main bronchus was observed. A ENT specialist was called for a consultation, tamponade was put in larynx and afterwards further ventilation became possible, leakage was acceptable. Since the admission was in the middle of the night, MSCT of the neck was made the following morning. It revealed a tracheomegaly with broadest diameter of 38,5cm and endotracheal tube cuff within the

middle third of the trachea whose diameter was up tp 30,5mm. Bronchomegaly of initial parts of bronchi – 20mm in the right and 19mm in the left bronchus.

In the repeated bronchoscopy we found many mucose impactions that obstruct main right bronchus (PHD: food particles of herbal origin). He was weaned and extubated the next day, he was oxygenated over HFNC and he was coughing up, assisted by Cough Assist device. His oxygenation parameters worsened, so he was intubated again, this time tracheal tube 10,0 cuff was used (in preouvious attempts tube 8,5cuff was used) and further ventilation was uneventful. Percutaneous tracheotomy was performed to maintain the airway secure and he was weaned successfully in following days.



Figures showing discrepancy between tracheal wall and tracheal tube, MSCT.

Discussion

Mounier-Kuhn syndrome is rare underdiagnosed condition of unknown prevalence. About 300 cases have been reported to date. The condition is more frequent in men [2]. It results from the atrophy of elastic fibers in the trachea and main bronchi which lead to thinning of the smooth muscle layer and subsequent tracheobroncheal flaccidity, dilatation and collapse. The etiologic mechanism remains unknown. Possibly it is a congenital defect of the smooth muscle of the airway, due to which the cough mechanism is ineffective, which leads to recurrent infections.

This rare clinical and radiologic condition is characterized by marked tracheobronchial dilation and recurrent lower respiratory tract infections [5]. Diagnosis is typically accomplished with the use of computed tomography and bronchoscopy, as well as pulmonary function testing. Patients may be asymptomatic; however, symptoms can range from minimal with preserved lung function to severe respiratory failure. Therapy, if any, is supportive but minimal [5].

Conclusion

Rare conditions as this one is may cause a real nightmare to an intensivist, since inability to ventilate is a real nightmare to a health professional. This syndrome is probably underdiagnosed, so it should be considered as differential diagnosis with recurrent lower respiratory tract infections, specially within a middle aged men group.

References

1. Milić R, Plavec G, Stojisavljević M, Tomić I, Šarac S. The Mounier-Kuhn syndrome. Vojnosanitetski pregled. 2010; 67: 867-870.
2. Search for a rare disease Orpnahet: Mounier-Kühn syndrome. (Accessed on 23 Mar 2024) https://www.orpha.net/en/disease/detail/3347
3. Rjimati M, Serrai M, Elbiaze M, Amara B, Benjelloun MC. Mounier-Kuhn syndrome (Tracheobronchomegaly): Radiological diagnosis. Radiol Case Rep. 2021; 16: 2546-2550.
4. Gaillard F, Elhusseiny A, Saber M, et al. Mounier-Kuhn syndrome. Reference article, Radiopaedia.org (Accessed on 23 Mar 2024) https://doi.org/10.53347/rID-8592
5. Celik B, Bilgin S, Yuksel C. Mounier-Kuhn syndrome: a rare cause of bronchial dilation. Tex Heart Inst J. 2011; 38: 194-196.