Coexistence of Lung Cancer and Mounier-Kuhn Syndrome

Akciğer Kanseri ve Mounier-Kuhn Sendromu Birlikteliği

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Abstract: Mounier-Kuhn Syndrome, characterized by tracheobronchomegaly, is a rare condition marked by the loss of elasticity in the airways, particularly leading to the expansion of the trachea and main bronchi. Mounier-Kuhn syndrome, first defined in 1932, is characterized by recurrent lower respiratory tract infections and bronchiectasis due to difficulty in expelling secretions. Its association with lung cancer has been rarely reported. In our 56-year-old male patient, a mass lesion was detected on a chest CT scan, and further investigations revealed that the mass was a squamous cell carcinoma, accompanied by tracheobronchial dilation. After the tests, the patient was diagnosed with Mounier-Kuhn syndrome. Based on the evaluation, the patient was referred to medical oncology as a case of multiple N2, and in our study, we aimed to present this rare association.

Key Words: Lung cancer, Syndrome, Mounier-Kuhn

Özet: Trakeabronkomegali ile karakterize olan Mounier-Kuhn Sendromu hava yollarının elastikiyetinin bozulması ve özellikle trakea ve ana bronşlarda genişleme ile karakterize ve nadir görülen bir hastalıktır. İlk kez 1932 yılında tanımlanan Mounier-Kuhn sendromu sekresyonların atılmasında zorluğa bağlı tekrarlayan alt solunum yolu enfeksiyonları ve bronşektaziler ile karakterizedir. Akciğer kanseri ile birlikteliği nadir olarak raporlanmıştır. 56 yaşında erkek olgumuzda çekilen toraks bilgisayarlı tomografisinde kitle lezyonu saptanması üzerine yapılan ileri incelemede kitlenin squamoz hücreli karsinom olduğu, trakeobronşial dilatasyonun eşlik ettiği saptandı. Yapılan tetkikleri sonrasında hasta Mounier-Kuhn senromu olarak değerlendirildi. Hastanın değerlendirmelerinde hasta multiple N2 olarak değerlendirilmesi üzerine medikal onkolojiye yönlendirilirken bizde çalışmamızda bu nadir birlikteliği sunmaya çalıştık.

Anahtar Kelimeler: Akciğer kanseri, Sendrom, Mounier Kuhn

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Received: 31 December 2024 **Accepted:** 28 July 2025 **DOI:** 10.33716/bmedj.1610571

INTRODUCTION

Mounier-Kuhn Syndrome (MKS), also known as tracheobronchomegaly, is a rare clinical condition characterized by the dilation of the trachea and bronchi. This syndrome was first described by Mounier-Kuhn in 1932 (Mounier-Kuhn, 1932; Ayup and Saif, 2017). The dilatation associated with MKS was first visualized radiologically using computed tomography in 1988 (Shin et al., 1988; Dunne and Reiner., 1988). The dilatation caused by MKS complicates the clearance of respiratory secretions, leading to conditions such as bronchiectasis, recurrent lower respiratory tract infections, and pulmonary fibrosis. The association of MKS with lung cancer has been rarely reported in the literature (Ayup and Saif, 2017).

In our study, we aim to present a case in which squamous cell carcinoma was detected in the lung, and during further investigations, MKS was diagnosed, alongside a review of the relevant literature.

Case Report

A 56-year-old male patient with no known chronic illnesses and a 40 pack-year history of smoking presented to an external medical center with complaints of persistent, recurrent cough and sputum production. A chest radiograph obtained at the referring center revealed consolidation in the left lung. The patient was referred to our institution for further evaluation and diagnostic workup. We were found to have a mass lesion measuring 37 mm, invading the artery and causing atelectasis in the apical segment of the left upper lobe on chest computed tomography (CT). Additionally, tracheobronchomegaly, particularly affecting the trachea and right main bronchus, was noted (Figure 1).

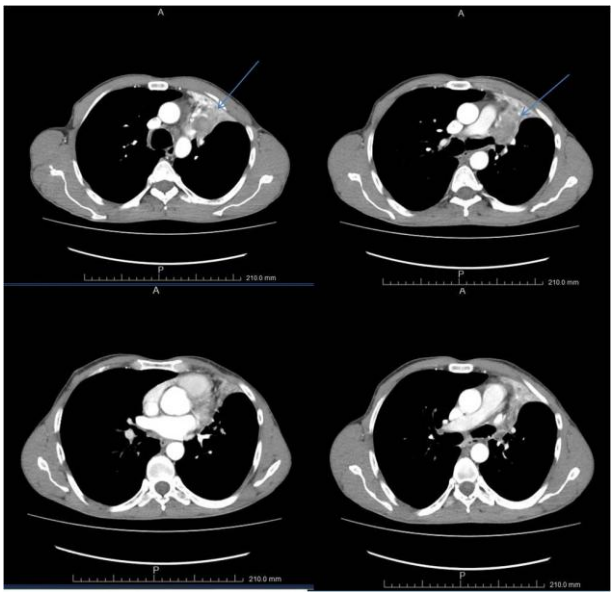

Figure 1: Appearance of the mass located in the left lung on thorax CT.

On chest CT, the tracheal diameter was 34.4 mm in the transverse section and 27.9 mm in the sagittal section, while the right main bronchus diameter was 21.8 mm in the transverse section and 20.8 mm in the sagittal section (Figure 2).

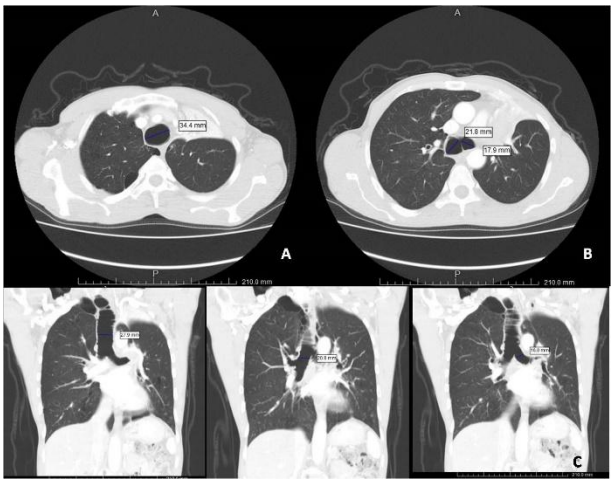

Figure 2: Thorax CT shows tracheobronchomegaly.

Bronchoscopy revealed that the trachea and main bronchi were dilated, with complete obstruction of the left upper lobe bronchus by the mass and widespread mucopurulent secretion in other areas. Based on these findings, the patient was diagnosed with MKS. A biopsy of the mass was reported as squamous cell carcinoma. A positron emission tomography (PET) scan revealed uptake in the lesion in the left upper lobe with an SUVmax of

16.8, as well as involvement of mediastinal lymph nodes, including the largest (1.6 cm, SUVmax 9.9) located in the pre- and subcarinal regions. Based on the current findings, the case was classified as T2aN2M0 stage 3A and referred to medical oncology.

DISCUSSION

Mounier-Kuhn Syndrome is a disease characterized by tracheobronchomegaly, first described in 1932 (Mounier-Kuhn 1932). The diagnosis of this disease was first made through bronchoscopy in 1949 by Lemoine, who recorded the bronchoscopy findings (Simon et al., 2014). The radiological findings of the disease were first revealed by chest CT in 1988 (Shin et al., 1988; Dunne and Reiner., 1988).
The disease is more commonly found in middle-aged men. The pathology of MKS is thought to be due to pathological development of connective tissue or the absence of smooth muscle cells in the trachea, leading to the breakdown of the elastic structure in the trachea and the development of tracheobronchial dilation (Ayup&Saif., 2017; Pacheco et al., 2018). Patients typically present with recurrent infections, shortness of breath, hemoptysis, and purulent sputum (Pacheco et al., 2018).

Krustins reviewed 128 cases of MKS published in the last 25 years (Krustins 2016). Among these cases, 114 were male and 14 were female, with an average age of 53.9. The average tracheal diameter in these patients was measured at 36 mm. Pacheco et al. reported a maximum tracheal diameter of 39 mm in their cases (Pacheco et al., 2018). Simon et al. described a maximum tracheal diameter ranging from 40 to 50 mm in their cases (Simon et al., 2014). In our case, the widest tracheal diameter was measured as 34.4 mm.

In the differential diagnosis of MKS, connective tissue disorders should also be considered, and the diagnostic process must be carried out with careful attention. Rare conditions that may lead to secondary tracheobronchial dilatation—such as Ehlers-Danlos syndrome, Marfan syndrome, Kenny-Caffey syndrome, Brachmann-de Lange syndrome, and cutis laxa—should be kept in mind during evaluation (Celik et al., 2011).

Literature review shows that MKS is commonly associated with clinical findings such as recurrent pneumonias, bronchiectasis, and hemoptysis. Ayup and Saif reported the first case of lung cancer associated with MKS in 2017 (Ayup and Saif, 2017). The patient, a 62-year-old male, had a squamous cell carcinoma mass in the right upper lobe of the lung and metastases to the liver and brain at the time of diagnosis. Pacheco et al. also presented a 40-year-old male patient with MKS who had a mass resembling Lymphangiomyomatosis in the right upper lobe, caused by an Aspergillus ball in 2018 (Pacheco et al., 2018). In our case, a 56-year-old male patient with previously undiagnosed MKS had a mass lesion obstructing the left upper lobe bronchus. The pathology was reported as squamous cell carcinoma, and mediastinal lymph node involvement was present at the time of diagnosis. Due to these findings, the patient was considered inoperable and referred to oncology.

CONCLUSION

In conclusion, MKS is a rare disease that often goes unnoticed unless symptomatic or detected during chest CT imaging for another reason. Patients typically present with lower respiratory tract infections and receive symptomatic treatment; however, it should be kept in mind that there may be an underlying endobronchial mass in these patients.

Authorship Contributions: MK, İA collected the data and wrote the main manuscript. MK, İA analyzed and interpreted the patient data. MK, İA designed the work and substantively revised the article. All authors read and approved the final manuscript.

Funding: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflict of interest: The authors have no conflicts of interest to declare.

Ethical approval: Informed consent was obtained from the patient to publish this report. The study was conducted in line with the principles of the "Helsinki Declaration."

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