Radiology Case Reports 19 (2024) 3962–3965

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journal homepage: www.elsevier.com/locate/radcr

Case Report

An atypical encounter: Mounir-Kuhn syndrome and aspergilloma coexistence: A case report☆,☆☆

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ARTICLE INFO

Article history:
Received 14 May 2024
Revised 31 May 2024
Accepted 3 June 2024

Keywords:
Mounier-Kuhn syndrome
Tracheobronchomegaly
Aspergilloma
COPD

ABSTRACT

We report a coexistence of Mounir-Kuhn syndrome and aspergilloma in a 69-year-old male presented with hemoptysis, cough, shortness of breath and fever. This patient has a history of recurrent hospital admissions for infective exacerbations of chronic obstructive pulmonary disease. Diagnostic imaging revealed the presence of aspergilloma and marked dilatation of the trachea and main bronchi, leading to a diagnosis of Mounier-Kuhn syndrome. Given the frequent association of Mounier-Kuhn syndrome with recurrent respiratory infections, we recommend that clinicians consider this syndrome when evaluating patients with recurrent pneumonia.

Background

Mounier-Kuhn syndrome (MKS), also known as tracheobronchomegaly, is a rare condition that is still the subject of debate over whether it is congenital or acquired [1]. It is characterized by marked widening of the trachea and major bronchi and should be sought in adults with recurrent respiratory tract infections with or without underlying pulmonary diseases [2]. MKS is principally a clinical-radiological diagnosis, and the main diagnostic tests are conventional chest X-rays and chest computed tomography [3]. Due to its generalized nature, the main focus of treatment lies in modifying the risk factors, addressing the recurrent infections, and minimising the potential complications; thereby, room for surgical interventions might not be available [4]. Here, we report a case of MKS associated with a rare encounter of aspergilloma. We aim to raise awareness about this rare and underdiagnosed condition, highlight its diagnostic challenge, and provide some key messages regarding management.

Case presentation

We present a case of a 69-year-old gentleman who smokes, averaging 20 cigarettes per day for 35 years (35 pack years), with a history of recurrent infective exacerbations of chronic

☆ Acknowledgments: This article received no funds.
☆☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.
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https://doi.org/10.1016/j.radcr.2024.06.001

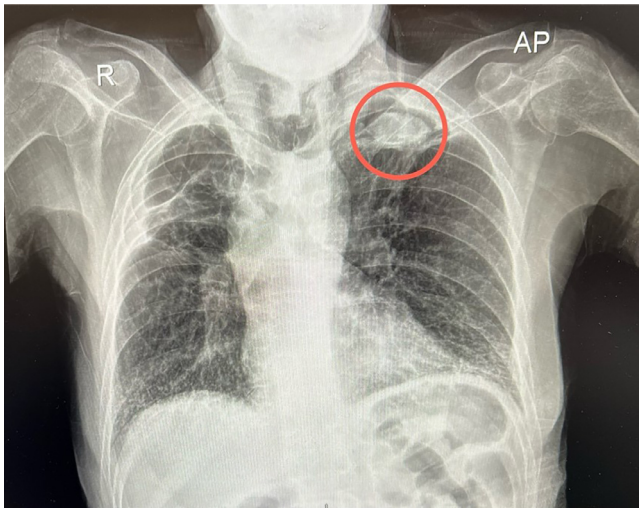
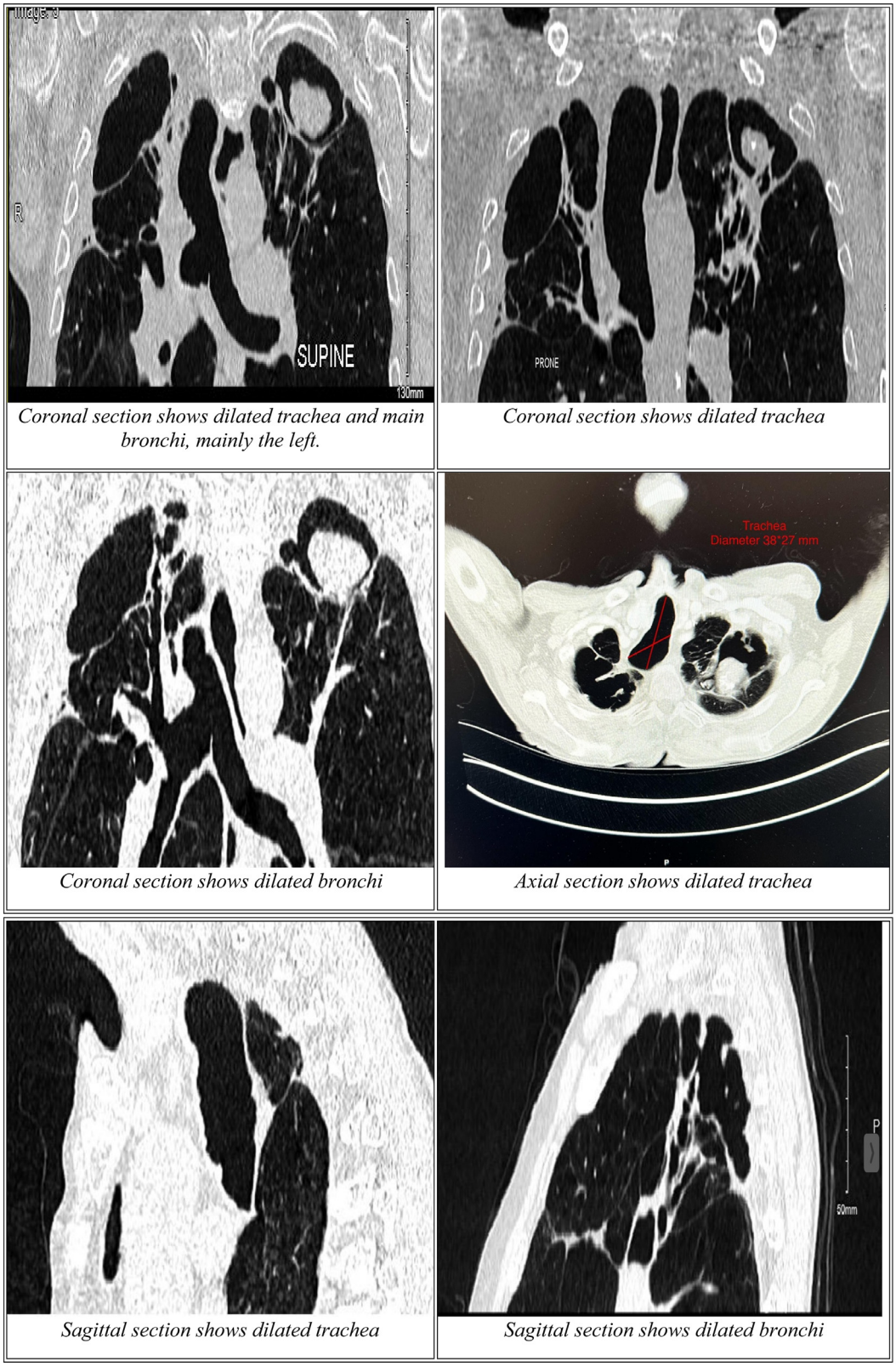


Fig. 1 – Anteroposterior CXR reveals dilatation of the tracheal cavity and aspergilloma (Red circle).

obstructive pulmonary disease (COPD), which necessitated multiple hospitalisations over the past 2 years. Presented to the emergency department with fever, shortness of breath, cough, and hemoptysis, along with significant weight loss over the last 6 months (weight on admission: 39 kg, BMI 12). His family history included COPD related to his father, but no lung cancer or other chronic illnesses. Occupational history did not indicate any significant exposure to industrial dust or chemicals. On the clinical exam, he looked ill, cachectic, dyspneic, and diaphoretic. Vital signs on admission were a pulse rate of 96 beats per minute, a respiratory rate of 38 cycles per minute, a blood pressure of 146/94, and a temperature of 37.9°C. Chest examination revealed signs of chest hyperinflation, bronchial breathing in the left upper chest, and scattered wheezing. Chest X-rays (Fig. 1) demonstrated bulbous emphysema in the upper zones with irregular pleural thickening and a rounded mass-like density in the left apex, raising differential diagnoses of aspergilloma versus pleural mass. Further imaging with chest computed tomography (CT) revealed tracheobronchomegaly involving the entire trachea and main bronchi, with sagittal and transverse tracheal diameters of 38 and 27 mm, respectively. Pulmonary aspergillomas were evident in the left upper lung lobe (Fig. 2). A galactomannan test was done, and it came back positive twice. Fiberoptic bronchoscopy revealed dilated trachea and enlarged main bronchi with prominent cartilage rings and mucosal atrophy (Fig. 3). Pathological examination of bronchial lavage specimens confirmed septate hyphae typical of Aspergillus spp., leading to a diagnosis of MKS with pulmonary aspergilloma. Treatment commenced with oral Voriconazole and tranexamic acid. However, Voriconazole was switched to Itraconazole due to deranged liver functions. Surgery was considered in the multidisciplinary meeting due to the risk of life-threatening massive hemoptysis, but the patient was deemed unfit. However, he experienced rapid recovery from hemoptysis without any recurrence, and he continued taking oral itraconazole for the long term, even though there was only limited improvement after completing the 6-month course. After 8 months, the patient was readmitted with massive diarrhea, diagnosed as a Clostridium difficile infection likely related to itraconazole, and treated with oral vancomycin for 10 days. Discharged in good condition. He undergoes regular follow-ups with the respiratory outpatient clinic.

Discussion

MKS is a rare clinical-radiological diagnosis whose origin, whether congenital or acquired, is still debatable [1]. The condition is characterized by marked widening of the trachea and major bronchi. Czyhlarz initially identified the illness during an autopsy in 1897. It was not until 1932 that Mounier-Kuhn linked the visibly expanded airway seen in endoscopic and radiographic images with repeated respiratory tract infections. MKS is also known as tracheobronchomegaly, tracheobronchiectasis, tracheocele, trachiectasis, megatrachea, tracheomalacia, and tracheobronchopathia malacia [2]. It is characterized by significant dilation of the trachea and main bronchi associated with thinning or atrophy of the elastic tissue and sometimes diverticulum formation due to the increased compliance of the tracheal wall [3,4]. Those significantly enlarged and compromised airways, together with an ineffective cough mechanism, hinder the removal of mucus through the mucociliary system, which eventually results in mucus buildup and the occurrence of repeated pneumonia, emphysema, bronchiectasis, and scarring of lung tissue [4,5]. Therefore, MKS should be sought in instances of recurrent chest infections. The clinical presentation is vague and can vary significantly, ranging from asymptomatic disease with preserved lung functions to recurrent chest infections, wheezing, and exertional dyspnea [2,4,6,7]. However, the majority of patients presented with recurrent bronchopulmonary infection [2,3], frequently interpreted as infective exacerbations of COPD [7]. MKS can co-occur with other congenital connective tissue illnesses such as Ehlers-Danlos syndrome, cutis laxa, and Marfan syndrome [6,7], and respiratory conditions like pulmonary fibrosis and bronchiectasis [1]. In our case, the patient had aspergilloma, which had only been reported a few times in conjugation with MKS [3,8]. In their comprehensive review involving 365 patients, Payandeh et al. delineated four distinct cohorts of MKS patients based on etiologies. The first cohort received the diagnosis following tracheal occlusion, while the second cohort manifests after a respiratory illness, suggesting that inflammation or infection could serve as both a consequence and a potential trigger. The third group presents with cutaneous elastolysis, indicating a possible underlying systemic condition. Finally, the fourth cohort comprises individuals for whom no discernible pattern of traits suggests the aetiology of the disease [3]. MKS is mainly a radiological diagnosis, and the main diagnostic tests are plain chest X-rays and computed tomography. Bronchoscopy can also record the findings [3,4]. An adult is diagnosed with tracheobronchomegaly if the diameter of the trachea, right main bronchus, and left main bronchus exceeds 30, 24, and 23 mm, respectively, on a standard chest radiograph or bronchogram, as these measurements are the upper limits of the means plus three standard deviations [4]. In our case, the



Coronal section shows dilated trachea and main bronchi, mainly the left.

Coronal section shows dilated trachea

Coronal section shows dilated bronchi

Axial section shows dilated trachea

Sagittal section shows dilated trachea

Sagittal section shows dilated bronchi

Fig. 2 – Chest CT reveals dilated trachea and main bronchi, and aspergilloma on coronal, axial and sagittal sections.

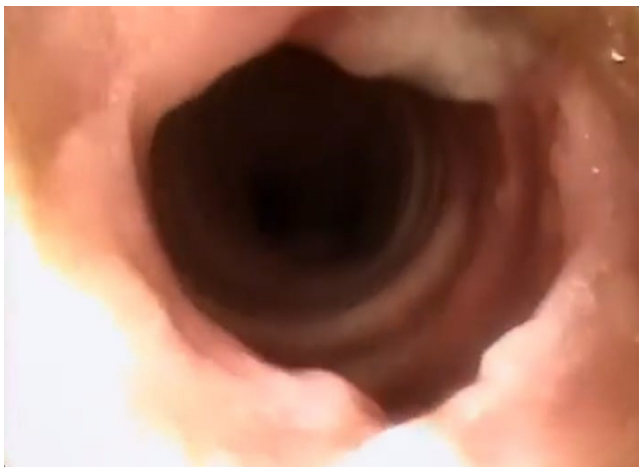


Fig. 3 – Fibreoptic bronchoscopy showed dilated trachea and prominent cartilage rings with mucosal atrophy.

trachea's transverse and sagittal diameters were 27 and 38 mm. Upon bronchoscopy examination, proximal airway dilation, tracheal, and bronchial diverticula, and retained secretion could be observed [6]. In our case, we detected dilated trachea and enlargement of both main bronchi. The cartilage rings of the trachea and main bronchi were notably prominent. Mucosal atrophy of both the trachea and bronchi was also evident. Patients without symptoms do not need any specific treatments except quitting smoking and reducing exposure to occupational pollutants or irritants [6]. Treating the infection and offering chest physiotherapy to encourage mucociliary clearance are feasible options in symptomatic cases. Surgery is not indicated due to the widespread extent of the illness. Bronchoscopy may be necessary for clearing secretions, and tracheostomy may be necessary in difficult situations [2,4].

Learning points

- MKS is frequently overlooked because of its rarity.
- Clinicians should maintain a high index of suspicion for MKS in patients experiencing recurrent pneumonia.
- In the context of COPD, recurrent infective exacerbations should not be automatically attributed solely to the underlying COPD.

Patient consent

We, the authors, certify that we have obtained written informed consent from the patient to publish our case report, including accompanying images. The patient has been encouraged to ask questions and has received satisfactory answers to all his inquiries. He had also been told to feel free to withdraw at any time before publication.

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