

Early manifestation of ARDS in COVID-19 infection in a 51-year-old man affected by Mounier-Kuhn syndrome

Ramezan Jafari^a, Luca Cegolon^{b,c}, Fatemeh Dehghanpoor^a, Mohammad Javanbakht^{d,*},
Morteza Izadi^e, Seyed Hassan Saadat^{d,f}, Babak Otoukesh^{g,h}, Behzad Einollahi^d

^a Department of Radiology, Baqiyatallah University of Medical Sciences, Tehran, Iran and Health Research Center, Baqiyatallah University of Medical Sciences, Tehran, Iran

^b Local Health Unit N. 2 "Marca Trevigiana" Public Health Department, Treviso, Italy

^c IRCCS "Burlo Garofolo", Institute for Maternal & Child Health, Trieste, Italy

^d Nephrology and Urology Research Center, Baqiyatallah University of Medical Sciences, Tehran, Iran

^e Health Research Center, Baqiyatallah University of Medical Sciences, Tehran, Iran

^f Behavioral Science Research Center, Lifestyle Institute, Baqiyatallah University of Medical Sciences, Tehran, Iran

^g Bone and Joint Reconstruction Research Center, Iran University of Medical Sciences, Tehran, Iran

^h Orthopedic Surgery Fellowship in Département Hospitalo-Universitaire MAMUTH «Maladies Musculo-Squelettiques et Innovations Thérapeutiques», Faculté de médecine Pierre et Marie Curie, Sorbonne Université, Paris, France

ARTICLE INFO

Article History:

Accepted 12 May 2020

Keywords:

COVID-19

Mounier-Kuhn syndrome

Computed tomography

Respiratory tract infections

ARDS

ABSTRACT

A 51-year-old man known to be affected by Mounier-Kuhn syndrome (MKS), presented to Accident & Emergency (A&E) with fever, dyspnea and deterioration of his chronic coughs. Increased diameters of his trachea (39 mm), right (30 mm) and left (26 mm) main bronchi were revealed by chest computerized tomography (CT) scan. CT scan showed also ground-glass opacities (GGO) and bronchiectasis in the mid and lower zones of both lungs. COVID-19 infection was eventually confirmed by RT-PCR. A severe form of COVID-19 could occur even in the early stages of the disease in presence of underlying co-morbidities including MKS, which increases the susceptibility to more recurrent and severe respiratory infections.

Introduction

COVID-19 as an emerging infectious disease of the human respiratory system, with a broad clinical pattern ranging from asymptomatic to a critical life threatening condition.^{1–3} A severe form of COVID-19 can develop quickly in a minority of patients, with the onset of acute respiratory distress syndrome (ARDS), septic shock, pleural effusion and other organ dysfunction/failure.^{2,4–6}

Mounier-Kuhn Syndrome (MKS, a form of tracheo-bronchomegaly), is a clinical idiopathic yet extremely rare disorder, featured by abnormal dilatation of the trachea and main bronchi due to atrophy or lack of elastic fibers and smooth muscle cells. Since it is a significant risk factor for recurrent lower respiratory infections, MKS needs to be detected as early as possible to prevent potential complications.^{7–10}

We report a patient affected by MKS admitted to Baqiyatallah hospital in Tehran (Iran) for COVID-19, who subsequently developed a form of the disease with acute respiratory distress syndrome (ARDS).

Case presentation

A 51-year-old man, known to be affected by MKS was admitted to our accident & emergency (A&E) service of the Baqiyatallah hospital in Tehran (Iran), with a new onset of fever, dyspnea and deterioration of his chronic coughs. On physical examination the patient was ill, febrile, tachycardic, hypoxic with O₂ saturation of about 80% and diffuse rhonchi at chest auscultation. The most significant laboratory finding was a notable leukopenia.

A chest CT scan revealed increased diameter of his trachea (39 mm), right (30 mm) and left (26 mm) main bronchi, due to MKS. Furthermore, the CT scan showed bilateral ground-glass opacities (GGO) and bronchiectasis in the mid and lower zones of both lungs, with small air cysts in the lower zone of the right lung at coronal bronchiectasis in the mid and lower zones of both lungs, with small air cysts in the lower zone of the right lung at coronal (Fig 1) and axial (Fig 2) imaging's, compatible with an early stage ARDS.

In a few hours the patient became cyanotic, with altered mental status, shortage of breath and drop in O₂ saturation to about 45%. In compliance with Iran's treatment protocols, the patient was promptly

* Corresponding author.

E-mail address: mhmjvbt81@yahoo.com (M. Javanbakht).

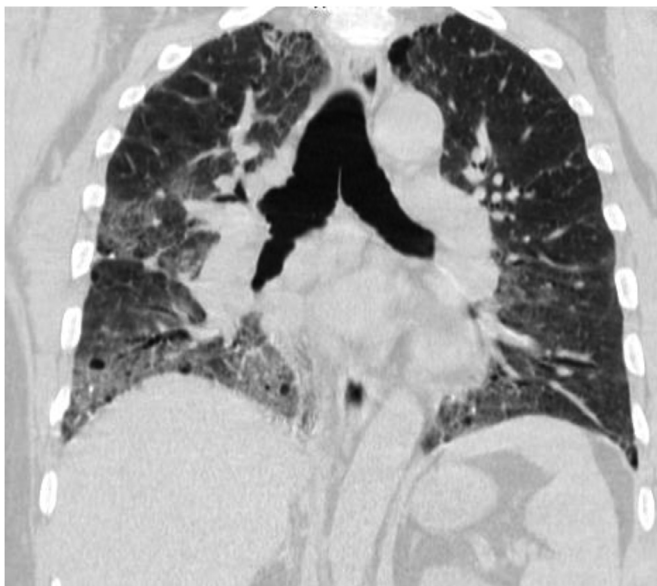


Fig. 1. Coronal reconstructed ct scan shows trachea-bronchomegaly (Mounier-Kuhn syndrome) . The diameter of trachea measures 39 mm, the right main bronchus 30 mm and the left main bronchus 26 mm. Ground-glass opacities (GGO) and bronchiectasis in the mid and lower zone of both lungs, with small air cysts in the lower zone of the right lung due to early stage ARDS in COVID-19 pneumonia can be noted.

intubated and transferred to the intensive care unit (ICU) for further diagnostic and therapeutic procedures, his clinical and radiological pattern being compatible with ARDS. He was placed under O₂ therapy mechanical ventilation, with a tracheostomy also performed. The patient health conditions progressively improved over time and after about two weeks he was weaned and discharged from hospital.

Although the patient conditions was unusual for an early stage of the disease, COVID-19 was still our first differential diagnosis, given we were in the middle of the COVID-19 epidemic. COVID-19 infection was eventually confirmed by real time reversed polymerase chain

reaction (RT-PCR). A severe form of COVID-19 could in fact occur even in the early stages of the disease in presence of underlying comorbidities, including MKS which increases the susceptibility to more recurrent and severe respiratory infections.

Discussion

MKS is a congenital disorder more common in men and manifesting with tracheo-bronchial dilation. Although the exact etiology of MKS is unknown, familial susceptibility and genetic predisposition seem to play a role. MKS is supposedly sustained by lack of musculo-elastic tissue at different levels of the trachea and main bronchi, causing diverticular structures and sacculational outpouchings especially in the posterior membranous trachea.^{7,9-12}

The clinical presentations of MKS are not specific and include a wide spectrum of clinical features ranging from asymptomatic or minimal disease to progressive manifestations leading to respiratory failure.¹³ Common presentations are chronic productive cough, dyspnea and recurrent respiratory infections due to mucociliary dysfunction and accumulation of secretions.^{11,14,15}

MKS' diagnosis relies on detection of enlarged diameter of the trachea, right and left main bronchi by CT scan.¹⁰⁻¹³ In the presented case the diameter of trachea, right and left bronchi were 39 mm, 30 mm and 26 mm, respectively. However, considerable variations in the enlargement of the lower respiratory tract tree have been documented. For instance, Woodring reported the diameter of right and left main bronchi being larger than 21 mm and 18 mm respectively in men affected by MKS.¹⁶ Similar radiological features were confirmed by Nobrega and Menon, with the diameter of trachea, right and left bronchi being larger than 30 mm, 24 mm and 23 mm, respectively.^{17,18} A remarkable trachea larger than 40 mm was reported by Bastos in a 33 year old man with MKS.¹⁹ Likewise, Abdelghani described a 53 old man affected by MKS with a diameter of his trachea, right and left bronchi being of 50 mm, 38 mm and 34 mm respectively.¹⁰

The above variability in the anatomical presentations of MKS can translate into different risk of acquiring respiratory tract infections and should be taken into account when managing COVID-19 patients, as they may influence the course of the disease. COVID-19 can

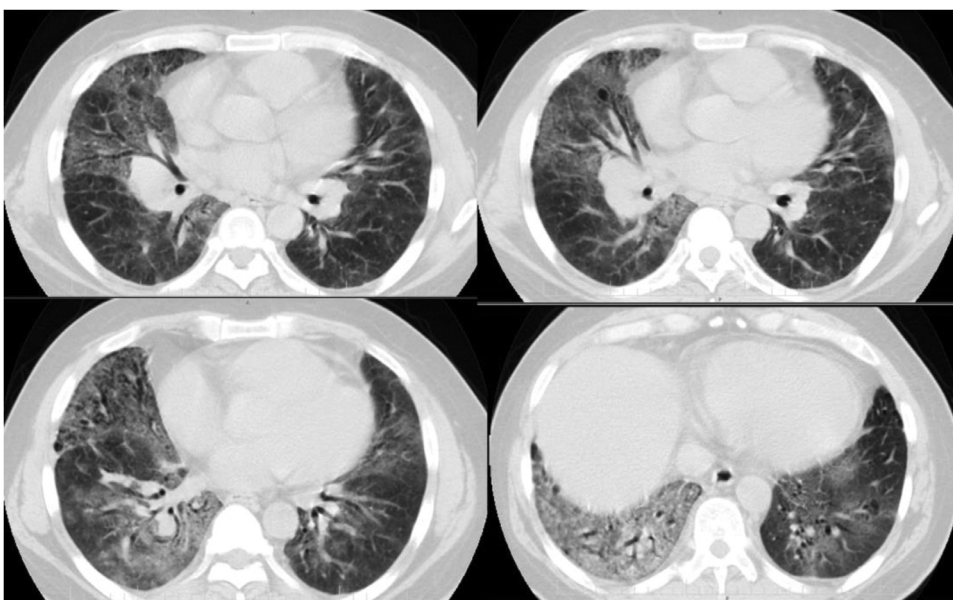


Fig. 2. Four axial CT scan show ill-defined bilateral ground-glass opacities (GGO) and bronchial dilatation within areas of GGO bilaterally and a small air cyst in the right middle lung lobe, suggestive of an early stage ARDS due to COVID-19 pneumonia subsequently confirmed by PCR test.

present with mild, moderate and severe symptoms. Whilst the vast majority of patients develop a mild form of the disease, underlying co-morbidities such as diabetes, hypertension, cardiovascular disorders and malignancies can significantly increase the risk of developing a severe form of COVID-19, especially among patients older than 50.^{20,21}

Conclusion

All possible underlying co-morbidities, even rare conditions as MKS, should be therefore carefully considered when managing a COVID-19 patient, because they can significantly influence the course of the disease.

Authors Contributions

RJ, LC, FD, MJ, MI, SHS, BK, and BE participated in editing throughout the writing process of case report, assessed the images of radiology and prepared the figures, design and draft of the case report. All authors have read and approved the final manuscript.

Acknowledgements

Thanks to guidance and advice from "Clinical Research Development Unit of Baqiyatallah Hospital".

Funding

None

Availability of supporting data

All data are available in the manuscript.

Ethics approval and consent to participate

This case report has been described in accordance with the ethical standards laid down in the "Declaration of Helsinki 1964".

Consent for publication

Written consent for publication was obtained from the patient.

Declaration of Competing Interest

The authors declare that they have no competing interests.

References

1. WHO-China Joint Mission. *Report of the WHO-China Joint Mission on Coronavirus Disease 2019 (COVID-19)*. 2020. <https://www.who.int/docs/default-source/coronavirus/who-china-joint-mission-on-covid-19-final-report.pdf>. (accessed March 1, 2020).
2. Lippi G, Plebani M, Henry BM. Thrombocytopenia is associated with severe coronavirus disease 2019 (COVID-19) infections: a meta-analysis. *Clinica Chimica Acta*. 2020;506:145–148.
3. Wu F, Zhao S, Yu B, Chen YM, Wang W, Song ZG, et al. A new coronavirus associated with human respiratory disease in China. *Nature*. 2020;579(7798):265–269.
4. Li H, Liu SHM, Yu XH, Tang SL, Tang CHK. Coronavirus disease 2019 (COVID-19): current status and future perspective. *Int J Antimicrob Age*. 2020 105951.. Available online 29 March; <https://www.sciencedirect.com/science/article/pii/S0924857920301011>.
5. Mattiuzzi C, Lippi G. Which lessons shall we learn from the 2019 novel coronavirus outbreak? *Ann Transl Med*. 2020;8. accessed March 8, 2020; <http://atm.amegroups.com/post/view/which-lessons-shall-we-learn-from-the-2019-novel-coronavirus-outbreak>.
6. Huang C, Wang Y, Li X, et al. Clinical features of patients infected with 2019 novel coronavirus in Wuhan, China. *Lancet*. 2020;395:497–506.
7. Lazzarini-de-Oliveira LC, Costa de Barros Franco CA, Gomes de Salles CL. A 38-year-old man with tracheomegaly, tracheal diverticulosis, and bronchiectasis. *Chest*. 2001;120:1018.
8. Ghanei M, Peyman MR, Aslani J, Zamel N. Mounier-Kuhn syndrome: a rare cause of severe bronchial dilatation with normal pulmonary function test: a case report. *Respiratory Med*. 2007;101(8):1836–1839.
9. Shah SS, Karnak D, Mason D, et al. Pulmonary transplantation in Mounier-Kuhn syndrome: a case report. *J Thorac Cardiovasc Surg*. 2006;131(3):757–758.
10. Abdelghani A, Bouazra H, Hayouni A, et al. Mounier-Kuhn syndrome: a rare cause of bronchial dilatation: a case report. *Respiratory Med CME*. 2009;2(4):164–166.
11. Gupta P, Gorsl U, Bhalla A, Khandelwal N. Mounier-Kuhn syndrome masquerading pulmonary thromboembolism in an elderly male. *Lung India*. 2014;31:76–78.
12. Sivanmani K. A case report of Mounier-Kuhn syndrome. *J Assoc Chest Physicians*. 2017;5:39–41.
13. Mesiha N, Mogia A., MONA Alipour M., et al. Mounier-Kuhn syndrome: a rare cause of a common disease. *Chest* 154(4):581A–582A.
14. Fletcher A, Stowell J, amoulis S. Congenital Tracheobronchomegaly (Mounier-Kuhn Syndrome) in a Woman with Human Immunodeficiency Virus: a Case Report. *Cureus*. 2017;9(4):e1136.
15. Gouder C, Bilocca D, Fsadni P, Montefort S. A delayed diagnosis of Mounier-Kuhn syndrome. *BMJ Case Rep*. 2014. <https://doi.org/10.1136/bcr-2014-203674>.
16. Woodring JH, Howard 2nd RS, Rehm SR. Congenital tracheobronchomegaly (Mounier-Kuhn syndrome): a report of 10 cases and review of the literature. *J Thorac Imaging*. 1991 Apr;6(2):1–10.
17. Menon B, Aggarwal B, Iqbal A. Mounier-Kuhn syndrome: report of 8 cases of tracheobroncho-megaly with associated complications. *South Med J*. 2008;101:83–87.
18. Nóbrega BB, Figueiredo SS, Cavalcante LP, et al. Traqueobroncomegalia (síndrome de Mounier-Kuhn): relato de caso e revisão da literatura. *Radiol Bras*. 2002;35:187–189.
19. Bastos AL, Brito ILA. Mounier-Kuhn syndrome: radiological findings and clinical presentation. *Radiol Bras*. 2011;44(3):198–200.
20. Cascella M., Rajnik M., Cuomo A., Dulebohn S.C., Napoli R.D. Features, Evaluation and Treatment Coronavirus (COVID-19). Last Update: March 20, 2020. https://www.ncbi.nlm.nih.gov/books/NBK554776/?fbclid=IwAR2b6hvY1reNFrmeQJcznD7wkd5NTj_X19LbhNkRi6hOwhEDmWocU9qeE_k.
21. World Health Organization. *Report of the WHO-China Joint Mission on Coronavirus Disease 2019 (COVID-2019)*. 2020. February 16–24.