RADIOLOGY FORUM

A Triad of Tracheomegaly, Tracheal Diverticula, and Bronchiectasis

Ram Babu Sah[1], RS Pal[2]

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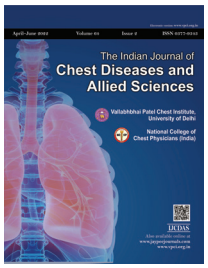

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Abbreviation used in this Article

CT = Computed tomography; CECT = contrast-enhanced computed tomography

Clinical Summary

Mounier-Kuhn syndrome is an uncommon respiratory disorder characterized by tracheomegaly, tracheal diverticula, and bronchiectasis. Patients usually have a history of recurrent respiratory tract infections. We report a case of an elderly male who presented to us for the evaluation of recurrent respiratory tract infection, affected by Mounier-Kuhn syndrome.

Investigations

A 61-year-old, non-smoker, male presented with complaints of productive cough and exertional dyspnea since childhood. He had a history of multiple episodes of recurrent respiratory tract infections in the past. There was no other comorbidity or significant past history. General physical examination was unremarkable. Examination of the respiratory system revealed bilateral coarse crackles and rhonchi. Other system examination was unremarkable. Routine hematological and biochemical tests were within normal limits. Chest radiograph (postero-anterior view) revealed tracheomegaly (Fig. 1). Further evaluation with contrast-enhanced computed tomography (CECT) of the thorax showed tracheomegaly, tracheal diverticula, and bilateral extensive bronchiectasis. This triad is characteristic of Mounier-Kuhn syndrome (Fig. 2).

[1,2]Department of Pulmonary Medicine, ESI-PGIMSR, Basaidarapur, New Delhi, India

Corresponding Author: Ram Babu Sah, Department of Pulmonary Medicine, ESI-PGIMSR, Basaidarapur, New Delhi, India, Phone: 01125970955, e-mail: rambabusah1008@gmail.com

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Diagnosis

Mounier-Kuhn syndrome.

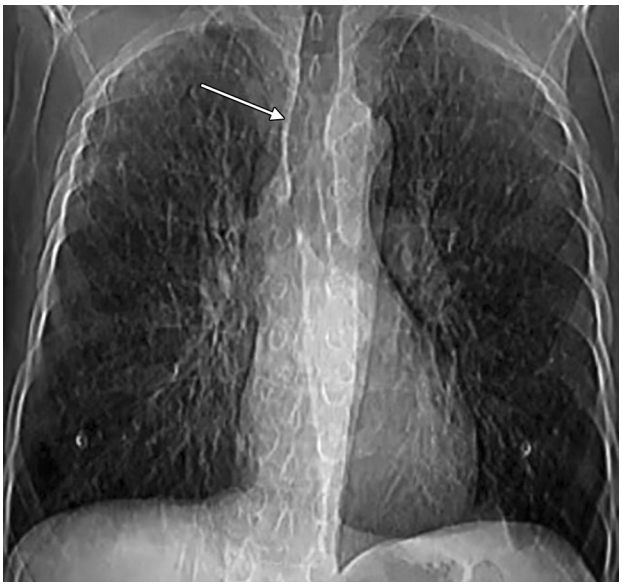
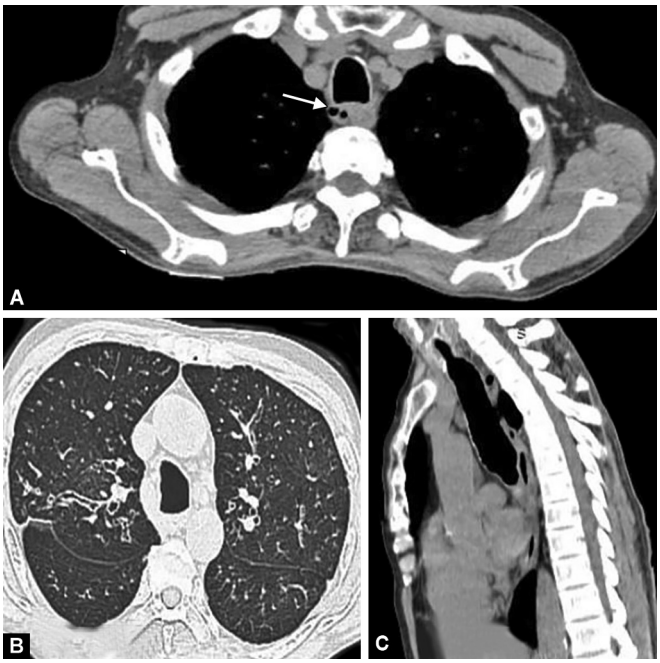


Fig. 1: Chest radiograph (postero-anterior view) showing tracheomegaly



Figs 2A to C: Contrast-enhanced computed tomography (CT) of thorax showing (A) Tracheal diverticula, (B and C) Tracheomegaly, and bilateral extensive bronchiectasis

Discussion

Mounier-Kuhn syndrome is a rare entity with abnormal tracheobronchial dilatation due to atrophy of the muscular and elastic tissues in the tracheal and the bronchial walls. Patients can develop tracheal diverticulosis and bronchiectasis. These patients usually present in the 3rd or 4th decade of life with non-specific respiratory symptoms including recurrent respiratory tract infection and subsequently end up being misdiagnosed with chronic obstructive pulmonary disease. Three subtypes of this syndrome had been described, (subtype I) symmetric dilation of the trachea and mainstem bronchi, (subtype II) tracheal dilation and tracheal diverticula, and (subtype III) has diverticular and saccular structures extending to the level of the distal bronchi.[1] Our patient likely fits into subtype II of this syndrome.

Tracheomegaly should be considered in males, when the transversal and sagittal diameter of the trachea is larger than 25 mm and 27 mm, respectively, and the diameter of the left-main bronchus and the right-main bronchus is larger than 18.4 mm and 21 mm, respectively, and for females, the transversal and sagittal diameter of the trachea is larger than 21 mm and 23 mm, respectively, and the diameter of the left-main bronchus and the right-main bronchus is larger than 17.4 mm and 19.8 mm, respectively.[2] In our case the transversal and sagittal diameter of the trachea was 28 mm and 31 mm, respectively.

Overall, treatment is supportive, usually with antibiotics, physiotherapy, and postural drainage. In rare instances, tracheal stenting has been used. It can be challenging in post-intubation patients due to cuff leaks.[3]

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