

CASE REPORT

A UNIQUE CASE REPORT OF TRACHEOBRONCHOMALACIA IN A PATIENT WITH HIV INFECTION

Ermias Merkebu, MD¹, Beza Leulseged, MD¹, Charles Sherman, MD², Dawit Kebede, MD^{1*}

ABSTRACT

Tracheobronchomalacia is an uncommon disease characterized by marked dilation of the trachea and mainstem bronchi, occurring as a result of atrophy or absence of elastic fibers and muscle tissues. Furthermore, atrophy of the connective tissue between the cartilage rings may result in the formation of diverticula, contributing to the development of recurrent respiratory infections. Tracheobronchomalacia may be congenital (Mounier-Kuhn Syndrome) or acquired. Patients can be asymptomatic or present with mild to severe disease with respiratory failure. We report the first case of Tracheobronchomalacia from East Africa, in a 45-year-old woman with HIV infection.

Key words: Tracheobronchomalacia, HIV.

INTRODUCTION

Tracheobronchomalacia (TBM) is an uncommon disease characterized by marked dilation of the trachea and main stem bronchi, occurring as a result of atrophy or absence of elastic fibers and muscle tissues, and fragmentation of the tracheal cartilage. These abnormalities result in proximal airway dilation during inhalation and collapse during exhalation. Furthermore, atrophy of the connective tissue between the cartilage rings can result in the formation of tracheal diverticula leading to the development of recurrent respiratory infections and further airway damage. TBM may be congenital (Mounier-Kuhn Syndrome) or acquired. Patients can be asymptomatic or present with mild to severe disease with respiratory failure (1). A case of Mounier- Kuhn Syndrome was previously reported from South Africa (2). We report the first case of TBM from East Africa in a 45-year-old woman with HIV who presented with recurrent respiratory infections.

CASE REPORT

A 45-year-old female patient presented to Tikur Anbessa Specialized Hospital, located in Addis Ababa, Ethiopia, for evaluation of two months of cough, productive of white sputum, and progressive dyspnea. She had a 12-year history of recurrent bronchitis. She had undergone tracheostomy 10 years ago after thyroglossal cyst resection. In addition, she had HIV disease for 10 years. Her recent CD4 count was 370 on antiretroviral treatment with Tenofovir, Lamivudine and Efavirenze and preventive therapy with Cotrimoxazole.

On presentation, she was in moderate respiratory distress. Her vital signs were PR- 112 /minute, BP- 80/50 mmHg, RR- 38 breaths/minute, T- 38.1^oC, and SaO₂ - 70-80% breathing room air. She had central cyanosis, finger clubbing, bibasilar crackles, a prominent P₂, and 1+ ankle edema.

Initial investigations included WBC-14.600 (N-79%), L-16%), Hemoglobin- 15g/dl, Platelets 614,000, ESR- 60mm/hr., a negative ANA, and a rheumatoid factor titer of less than 10. Sputum was negative for routine culture and negative for mycobacterium tuberculosis using Gene Xpert. Echocardiography showed an EF 60%, moderate pulmonary hypertension (PAP-57mmHg) and a dilated right atrium and ventricle. Chest x-ray showed dilated trachea and mainstem bronchi and bronchiectasis in both lower lobes (Figure 1). Chest CT showed similar findings but also diverticula in the distal trachea (Figures 2 and 3). Bronchoscopy revealed dilation of the trachea and mainstem bronchi with severe proximal collapsibility on expiration. Extensive diverticula were seen on the posterior wall of the distal third of the trachea (Figure 4).

The patient was admitted to the wards and started on IV Vancomycin and Ceftazidime. In addition, she received furosemide and supplemental oxygen at two liters/min. She clinically improved and was discharged on the fifth hospital day.

¹Addis Ababa University, College of Health Sciences, Addis Ababa, Ethiopia.

²Warren Alpert Medical School of Brown University, Providence, Rhode Island, USA.

*Corresponding Author e-mail: dndrda97@gmail.com



Figure 1: Chest x-ray showing tracheobronchomalacia

Figures 2: Chest CT showing tracheomalacia (transverse diameter 35.4 mm and bronchiectatic Changes).



Figure 3: Chest CT showing dilated main bronchi (transverse diameter of the right mainstem bronchus 25.3 mm and of the left mainstem bronchus 24.3 mm), tracheal diverticulum (arrow), and bronchiectatic changes.

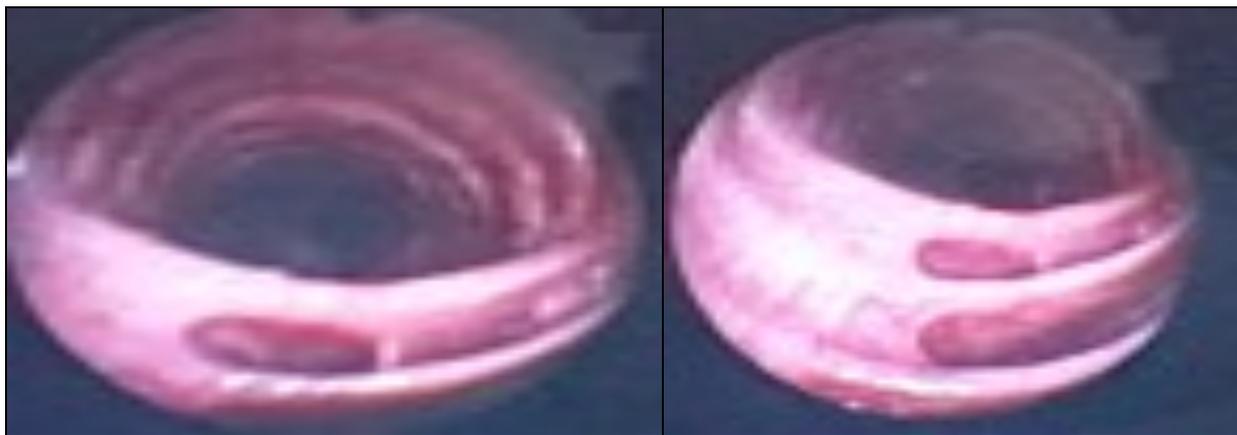


Figure 4: Chest CT showing dilated main bronchi (transverse diameter of the right mainstem bronchus 25.3 mm and of the left mainstem bronchus 24.3 mm), tracheal diverticulum (arrow), and bronchiectatic changes.

The patient was admitted to the wards and started on IV Vancomycin and Cefazidime. In addition, she received furosemide and supplemental oxygen at two liters/min. She clinically improved and was discharged on the fifth hospital day.

DISCUSSION

TBM is an uncommon disease that can be classified as congenital or acquired. Mounier and Kuhn first described the congenital syndrome in 1932 (3). The acquired form may result from airway trauma (i.e., post intubation, post tracheostomy), chronic airway infections, chronic inflammation (i.e. relapsing polychondritis), or chronic external compression of the trachea (i.e. malignancy, mediastinal goiter). Patients can be asymptomatic but most present with recurrent respiratory infections in the 3rd or 4th decade of their life, as did our patient (4,5). HIV, previous tracheostomy, and tracheal diverticula all contributed to our patient's airway abnormalities and recurrent bronchitis, pulmonary hypertension, and cor pulmonale. Prognosis varies but many patients die prematurely from sepsis and respiratory failure (6).

The diagnosis of TBM is often overlooked on chest x-ray. It is best made from Chest CT images where a transverse diameter of the trachea >30mm, of the right mainstem bronchus >24mm, and of the left mainstem bronchus >23mm confirm the diagnosis (6). The diagnosis can also be made based on demonstration of airway collapsibility on dynamic airway Chest CT imaging or during bronchoscopic examination. In our patient, the diameters of the trachea, right and left mainstem bronchi were 35.4 mm, 25.3 mm,

REFERENCES

1. Celik F, Bilgin S, Yuksel C. Mounier-Kuhn syndrome: a rare cause of bronchial dilation. *Tex Heart Inst J* 2011;38(2):194-6.
2. Bass EM. Tracheobronchomegaly: The Mounier-Kuhn Syndrome. *South Afr Med J* 1974;48(40):1718-1720.
3. Mounier-Kuhn P. Dilatation de la trachee: constatations radiographiques et bronchoscopiques. *Lyon Med* 1932;150:106-9.
4. Marques A, Felix M, Barata F, Pires J, Helena E, Martinho do Bispo Coimbra S. Mounier-Kuhn syndrome: a rare aetiology of recurrent respiratory infections. *Rev Port Pneumol* 2007;13(5):721-7.
5. Sivanmani K. A case report of Mounier-Kuhn syndrome. *J Assoc Chest Physicians* 2017; 5(1):39-41.
6. Bastos AL, Brito ILA. Mounier-Kuhn syndrome: radiological findings and clinical presentation. *Radiol Bras* 201;44(3):198-200.
7. Haages JR, Boll D. Congenital Tracheobronchomegaly (Mounier-Kuhn syndrome): A case report of 10 cases and review of the literature. *J Thorac Imaging* 1991;6:1-10.
8. Kheiralla OAM, Babikr WG, Globawe, MMA. Mounier-Kuhn syndrome: a case report and literature review. *WJPMR* 2016;2(3):13-16.
9. Lee JK. *Computed Body Tomography with MRI Correlation*. Vol 1. 2006. Philadelphia. Lippincott, Williams, and Wilkins.
10. Jain P, Dave M, Singh DP, Kumawat DC, Babel CS. Mounier-Kuhn Syndrome. *Indian J Chest Dis Allied Sci* 2002;44:195-198.

TBM is usually managed conservatively with postural drainage of respiratory secretions and antibiotics during episodes of infections (7,8). Supplemental oxygen is often required, which can be challenging to provide in low-income countries where significant number of patients receive health care service in primary health care system with limited access to oxygen therapy.

CONCLUSION

We report the first case of TBM in East Africa. We suspect that the disease is more common than previously described due to the high prevalence of HIV and respiratory infections in much of the continent. Clinicians should consider TBM as one diagnostic possibility in patients with chronic respiratory symptoms and recurrent respiratory infections, especially in those with HIV disease.

ACKNOWLEDGEMENTS

The authors would like to thank Vital Strategies and the Swiss Lung Foundation for their sponsorship of the East African Training Initiative, from which this work originates.