Original Research Paper

Volume - 14 | Issue - 04 | April - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Radiology

TRACHEOBRONCHOMEGALY (MOUNIER-KUHN SYNDROME): A DIAGNOSIS NOT SO COMMON

Dr Mukesh Chandra — MD (Radiology), Lead Radiology, Burjeel Speciality Hospital, Sharjah, UAE.

Dr Samaresh Sahu* — MD (Radiology), DNB(Radiology), FRCR (UK), Specialist Radiology, Burjeel Speciality Hospital, Sharjah, UAE. *Corresponding Author

ABSTRACT Tracheobronchomegaly is a rare entity and often diagnosed in 3rd and 4th decade. It is more common in male vis a vis female. Its transmission is autosomal recessive and sporadic in few cases. It is often associated with multiple diverticulae. Clinically these patients present with poor cough reflex and impaired mucociliary activity. An early diagnosis using CT scan with specific criteria can be helpful for the successful patient management.

KEYWORDS : Tracheal dilatation, Tracheal diverticula, Bronchiectasis

INTRODUCTION:

Tracheobronchomegaly also called as Mounier-Kuhn Syndrome is an extremely rare disease and often its diagnosis is delayed. It is characterised by dilatation of the trachea and main stem bronchi. The aetiology is congenital or acquired remains controversial. First described by Mounier-Kuhn in 1932 till now only fewer than 100 cases have been described in literature. [1]

Here we describe a case of Mounier-Kuhn syndrome in an adult female who presented to our respiratory clinic with history of repeated episodes of lower respiratory tract infection.

Case Report:

32 Years old lady presented to our respiratory clinic with history of repeated episodes of lower respiratory infections.

A High-resolution Computerised Scan (HRCT) of Thorax was suggested for the patient.

HRCT scan of thorax of patient obtained in inspiratory and expiratory manuover revealed the trachea was dilated with maximum diameter of 26.2mm in anteroposterior and 22.5mm in transverse diameters. The diameters of right and left main stem bronchi were 24.6mm anteroposterior and 19 mm in transverse diameters respectively. There were multiple diverticular outpouchings noted arising from the posterior aspect of the trachea and along the tracheal rings. Further multiple diverticular outpouchings also noted arising from the mainstem bronchi and extending along the segmental bronchi bilaterally. Few of these outpouchings showed air-fluid levels within it. None of these bronchi revealed mucous plugging.

Based on these findings a diagnosis of Tracheobronchomegaly (Mounier-Kuhn Syndrome) with multiple tracheobronchial diverticula was made.

DISCUSSION:

Tracheobronchomegaly (Mounier-Kuhn Syndrome) was first described by Mounier-Kuhn in the year 1932 [1]. It is characterised by dilatation of trachea and bronchi and a predominant disease of males. It often presents in the 3rd to 4th decade with history of repeated episodes of chest infections[2].

The genetic transmission pattern of this disease is controversial in literature as some authors depicting it as a autosomal recessive disease as its association with Ehlers-Danlos Syndrome and Cutis Laxa in children [1,2]. While some authors describe this disease as sporadic. Acquired forms have described in neonates post ventilation [3]. Our patient possibly will fall into the sporadic case as no other association could be found.

This entity is characterised by atrophy of the longitudinal elastic fibres, which causes thinning of the muscularis leading to the unusual dilatation of the membranous and cartilaginous rings of trachea. In addition, it leads to formation of the diverticula along the redundant tissue along the tracheal and bronchial rings. This tissue is mostly along the posterior trachealis muscle [4].

Some authors propose smoking as a chronic irritant and a contributing factor for this entity. Though the history of smoking was not present in our patient [4].

The tracheal diverticulae can be congenital or acquired in nature. Congenital diverticulae of trachea, their pathogenesis remains uncertain. The acquired diverticulae of trachea often is found at the level of junction of intra and extrathoracic trachea. It is mostly posterior in location. Chronic cough possibly contributes to the acquired condition as it causes increased intraluminal pressure and the diverticulae formation happens through the weak spots [5].

Clinical symptoms are non-specific and often these patients present with history of recurrent lower respiratory tract infection. Rarely these patients may preset with spontaneous pneumothorax or haemoptysis [6]. Our patient presented with repeated episodes of lower respiratory tract of infection.

There can be various component of this syndrome beyond the pulmonary system. It may include ptosis, epicanthus, microganthism, excess upper lip skin. Our patient on clinical examination did not have any of these findings [7]. It also supports our diagnosis of sporadic aetiology in our case.

The diagnostic criteria for Mounier-Kuhn syndrome is based on the measurements of tracheal diameter on High resolution CT scans. The tracheal diameter in transverse and sagittal diameter increases beyond 25 and 27mm. The diameter of right and left main bronchus characteristically should increase beyond 18 and 21mm respectively in males. The same definition applies to females with measurements being respectively 21,23,17.4 and 19.8mm [8,9]. Few literature calculate the increase in the area of the trachea beyond 371 sq mm in male and 299 sq mm in females as the diagnostic criteria for this disease [10]. Though availability of multidetector CT scan has made this diagnosis possible bronchoscopy has a distinct role in terms of diagnosis and therapy.

Most common complication of Mounier-Kuhn syndrome includes bronchiectasis, fungal infection including aspergillosis. Our patient had cystic bronchiectasis. There can be infection with typical and typical mycobacteria further complicating the disease process [11,12].

Treatment is often symptomatic and involves control of infection. Bronchodilators and steroids are used often used to control symptoms. Protective vaccines including that of influenza and pneumococcal are suggested as per the age group [13]. Cessation of smoking is beneficial. Bronchoscopic stent placement, surgical options like tracheobronchoplasty may be last options tried in few patients when the conservative options fail [14]. Our patient was managed on conservative therapy with antibiotics, bronchodilators and she responded well.

CONCLUSION:

To summarise, the authors wanted to discuss a rare case of tracheobronchomegaly (Mounier-Kuhn Syndrome), its characteristic features, diagnostic features, complications.

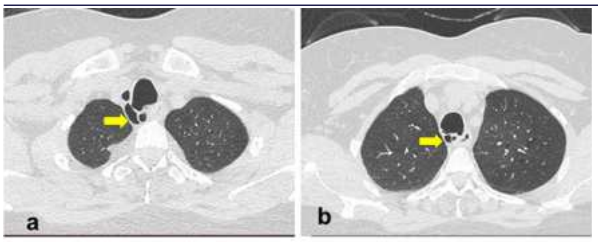

Fig 1 a,b: Axial lung window showing dilated trachea with multiple tracheal diverticula (yellow arrow) arising from the posterior aspect.

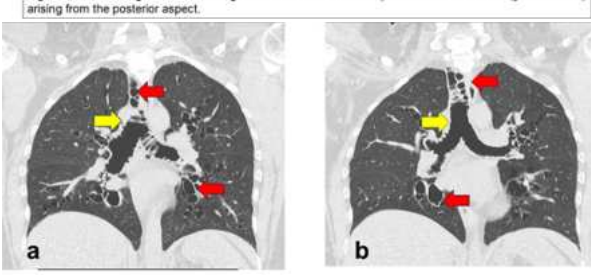

Fig 2 a,b: Coronal lung window showing dilated trachea (yellow arrow) with multiple tracheal diverticula(red arrow) arising from the main and segmental bronchi.

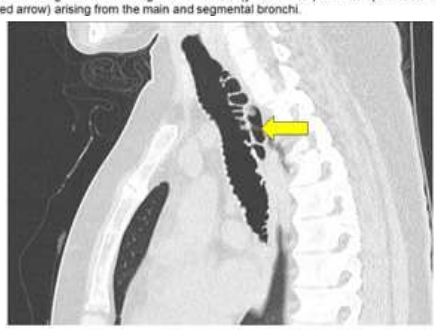

Fig 3: Sagittal lung window showing dilated trachea (yellow arrow) with multiple tracheal diverticula arising from the posterior aspect.

REFERENCES:

1. Mounier-Kunh P. Dilatation de la trachee. Constatations radiographiques et bronchoscopiques. Lyon Medical 1932; 150:160-9.
2. Celik B, Bilgin S, Yuksel C. Mounier-Kuhn syndrome: a rare cause of bronchial dilation. Tex Heart Inst J 2011;38(2):194–6.
3. Ghaneia M, Peymanb M, Aslanic J, Zamel N. Mounier-Kuhn syndrome: a rare cause of severe bronchial dilatation with normal pulmonary function test: a case report. Respir Med 2007; 101:1836–9.
4. Abdelghani A ,Bouazra H ,Hayouni A ,Slama S ,Garrouche A ,Mezghani S ,Klabi N ,Benzarti M . Mounier–Kuhn syndrome: a rare cause of bronchialdilatation: a case report. Respir Med CME 2009; 2:164–6.
5. Mondoni M, Carlucci P, Parazzini EM, Busatto P, Centanni S. Huge tracheal diverticulum in a patient with mounierkuhn syndrome. Europen J Case Rep Intern Med 2016; 3:413–19.
6. Kuwal A, Dutt N, Chauhan N, Kumar S, Purohit S, Saini LK. An atypical case of Mounier-Kuhn syndrome: time to change the diagnostic approach. Journal of Broncho Interventional Pulmonology 2017; 24:84–7.
7. Woodring JH, Howard RS, Rehm SR. Congenital tracheobronchomegaly (Mounier-Kuhn syndrome): a report of 10 cases and review of the literature. Journal of Thoracic Imaging 1991; 6:1–10.
8. Jaubert F., De Blic J. Malformations de l'appareil respiratoire. Poumon in Encycl Med Chir, Paris, France, 6025.
9. Fraser RS, Muller NL, Colman N, Pare PD. Diagnosis of diseases of the chest. 4th ed. Philadelphia, PA: Saunders; 1999.
10. Collard P, Freitag L, Reynaert MS, Rodenstein DO, Francis C. Respiratory failure due to tracheobronchomalacia. Thorax 1996; 51:224–6.
11. Krustins E. Mounier-Kuhn syndrome: a systematic analysis of 128 cases published within last 25 years. Clin Respir J 2016; 10:3–10.
12. Akgedik R, Karamanli H, Kizilirmak D. Mounier-Kuhn syndrome (tracheobronchomegaly): an analysis of eleven cases. Clin Respir J 2016; 12:1–5.
13. Krustins E, Kravale Z, Buls A. Mounier-Kuhn syndrome or congenital tracheobronchomegaly: a literature review. Respir Med 2013; 107:1822–8.
14. Schmitt P, Dalar L, Jouneau S, Toublanc B, Camuset J, Chatte G. Respiratory conditions associated with tracheobronchomegaly (Mounier-Kuhn Syndrome): a study of seventeen cases. Respiration 2016; 91:281–7