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Original Research Paper

Pulmonary Medicine

MOUNIER KUHN SYNDROME - A CASE REPORT OF 41 YEAR OLD MALE WITH TRACHEOMEGALY

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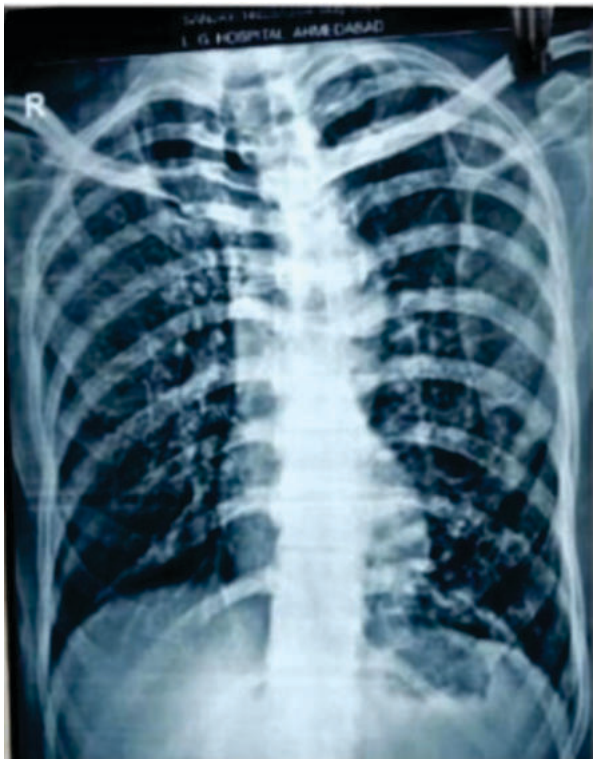
ABSTRACT

Introduction: Mounier Kuhn Syndrome(MKS) is a rare Autosomal recessive clinical and radiological entity characterized by marked tracheobronchial ditatation. To date, fewer than 400 cases of MKS have been reported.[1-1] While no epidemiological study has been published, MKS has been found to disproportionally affect men with 8:1 male to female ratio.[1-2] There is also a prevalence of MKS in both smokers and African Americans, and patients typically present in the third to sixth decades of life. The diagnosis is made through the synthesis of clinical and radiological data cases present in third or later decades with recurrent respiratory tract infections. Although the etiology is uncertain,it is believed to be due to lack of smooth muscle and elastic connective tissue in the trachea and main Bronchus leading to Sacculations and diverticula formation between cartilaginous rings. Disorders such as sarcoidosis , UIP and cystic fibrosis which cause severe fibrosis of upper lobe may also exert sufficient tracheal enlargment.

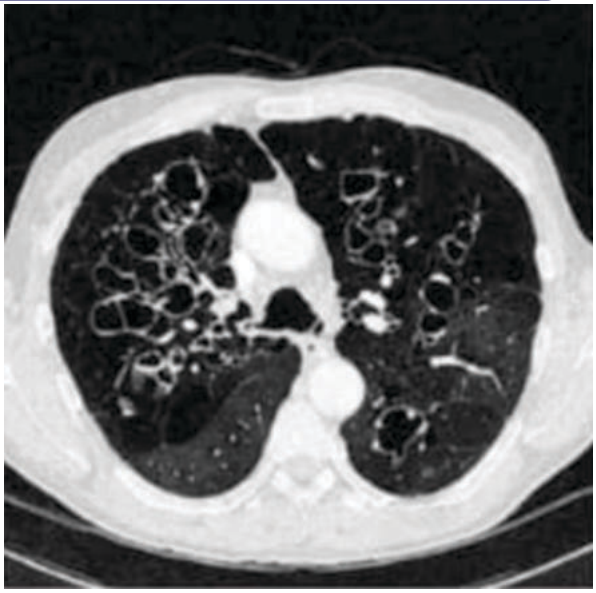
KEYWORDS : Tracheobronchomegaly, Lower Respiratory Tract Infections.

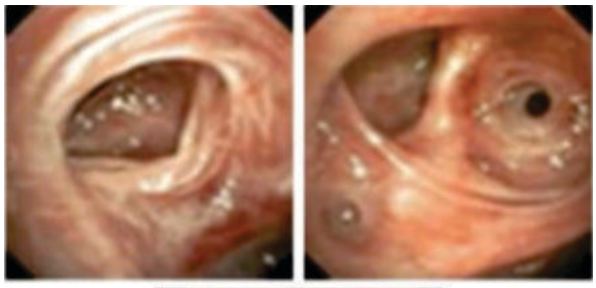
Case Report

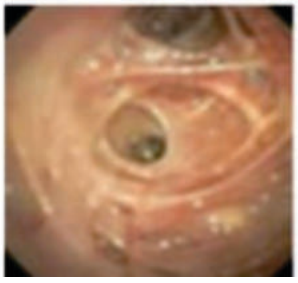
- A 41 year old male presented with complaint of recurrent symptoms of cough with whitish expectoration, fever, dyspnea on exertion since 6 years. Past history of pulmonary Tuberculosis 6 months ago. Allergy to smoke present, working as shoepolish worker.
- Patient undergone chest Xray, routine blood investigations, HRCT Thorax followed by Bronchoscopy .
- CBC and all other blood investigations within normal range .
- CXR shows Bilateral calcified spots in Bronchovascular markings, Emphysematous changes and left lower lobe cystic fibrosis as well as nodular lesions present .



- HRCT Thorax Suggestive of tracheobronchomegaly with cystic bronchoiectesis in bilateral lung fields and multiple diverticuli suggestive of Mounier Kuhn Syndrome. Ildefined Ground Glass opacities in both lung fields suggestive of superadded infection .







- Bronchoscopy suggestive of Tracheobronchomegaly with multiple tracheal and bronchial diverticula.
- Biopsy from secondary carina which shows smooth

muscles with degeneration progressive transition to fibrosis.

- While hospitalized, his symptoms improved after antibiotics, oral steroids, respiratory support, and chest physiotherapy .
- As an outpatient, he is currently well managed with mucolytics, chest physiotherapy, prophylactic vaccinations, and antibiotics during infectious exacerbations.

DISCUSSION

Mounier Kuhn syndrome is most likely due to loss of elastic fibers. Clinical signs are not specific and diagnosis is made by radiological and bronchoscopic findings. Treatment aims to treat broncho- pulmonary infections or better prevent them and to free the airways by physiotherapy. A wide range of Non pulmonary and pulmonary clinical consequences associated with mounier Kuhn syndrome like Bronchiectasis, Tracheobronchomalacia and emphysema. While most common non respiratory comorbidity is Gastroesophageal refulx disease. Patients are managed with Prophylactic vaccination and antibiotics .

REFERENCES

1. Texas heart institute journal
2. Menon B in B. Aggansal 8, Ital A. Mounier Kun syn report of 8 cases of tracheobronchomegaly with associated complications. Southmed J2008 101(1)83-7.