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A Rare Cause of Congenital Bronchiectasis – Mounier Kuhn Syndrome

Authors
Dr T.Bindu.MBBS.MD[1], Dr A.D.Nageswari.MBBS.DTCD.MD[2], Dr H.Peer Mohaideen.MBBS[3]
[1]Assistant Professor, Department of Respiratory Medicine, Tagore Medical College And Hospital Rathinamangalam-Chennai
[2]Professor/Head, Department of Respiratory Medicine, Tagore Medical College And Hospital Rathinamangalam-Chennai
[3]Junior Resident, Department of Respiratory Medicine, Tagore Medical College And Hospital Rathinamangalam-Chennai
Corresponding Author
Dr T.Bindu
No 65,West Mada Street, Thiruvottiyur Chennai-600019
Email: *k_sathish123@yahoo.com, Mobile No: 9841596047*

ABSTRACT
Tracheobronchomegaly also called "Mounier Kuhn Syndrome" is a rare congenital disorder of the lung characterised by an abnormal widening of the trachea and main stem bronchi.It is usually associated with recurrent lower respiratory tract infection, bronchiectasis and tracheal diverticulitis with or without respiratory complications The disease was first described in 1932 and since then, only few hundreds of cases have been reported. Symptoms usually appear in fourth to sixth decade of life and is usually similar to chronic bronchiectasis due to any other cause. Diagnosis depends on enlarged trachea and proximal bronchi on plain radiograph and CT-Chest. Treatment is mainly supportive. We report a case with this condition for its rarity.
Keywords: *Mounier Kuhn, bronchiectasis, trachealdiverticulosis.*

CASE REPORT

A 65 year old male, farmer by occupation, nonsmoker, presented to our hospital with complaints of productive cough (75ml/day) and breathlessness for 3 weeks. Sputum was mucopur-ulent and occasionally blood tinged. He had similar complaints in the past (6 episodes in 2 years) all of which resolved with a short course of antibiotics and none required hospitalization. Family history was irrelevant.

On examination patient was well nourished and well built,febrileand no clubbing was seen.SP02 was 96% in room air. Auscultation revealed bilateral diffuse coarse persistant crackles more over bases and diffuse polyphonic wheeze.0ther systems examination was normal.

CBC showed neutrophilia and ESR was 42mm/hr. Sputum AFB was negative and culture showed growth of streptococcal pneumonia. Plain radiograph showed dilated trachea with bilateral proximal cystic bronchiectasis with air fluid level (Fig: 1). Diagnosis was confirmed by CT Chest which revealed the presence of tracheobronch-omegaly with tracheal transverse and saggital

diameter of 40mm and 33mm respectively. Bilateral proximal cystic bronchiectasis was also noted. (Fig:2,3).

Bronchoscopy showed abnormally dilated trachea and main stem bronchi with increased mobility of the posterior tracheal wall on respiration. The bronchi contained copious mucopurulent secretions which was sent for microbiological examination. Endobronchialbiopsy and BAL showed features of chronic inflammatory changes.Spirometryshowed obstructive pattern with partialreversibility. Patient was treated initially with parenteral antibiotics followed by oral antibiotics, postural drainage therapy and bronchodilators.He symptomatically improved and is currently under regular follow up.

DISCUSSION

HISTORY

Tracheobronchomegaly was first recognised on post-mortemexamination by Czyhlarz in 1897 and later described by Mounier in 1932.[1] The term tracheobronchomegaly was derived from the review of Katz in 1962[2] This is a congenital disorder and principally affects the large airways[3]causing abnormally dilated trachea and mainstern bronchi.

PATHOLOGY

The primary pathology is a congenital defect or atrophy of the elastic and smooth muscle tissue of the trachea and principal bronchus.[1,4] This leads to dilatation of membranous and cartilaginous portion of the airway,mucosalherniation between tracheal rings and subsequent tracheal diverticulosis. lnspirationcauses ballooning and expiration causes collapse of the airways.[1,5]

ETIOLOGY

Etiology remains uncertain in many cases.[1,4] May be associated with Ehler Danlos syndrome, skeletal dysplasias, cutislaxa and ankylosingsp-ondylosis.[1,4] Inhalation of irritants like cigarette smoke and air pollution were raised as possible risk factors in the progression of this condition.[1,6]

CLINICAL PRESENTATION

Affected individuals are commonly males of middle age.[1,7]Clinical features range from asymptomatic presentation detected on screening to recurrent lower respiratory tract infection, bronchorrea, spontaneous neumothorax and hemoptysis. Clubbing may or may not be present.

DIAGNOSIS

Diagnosis depends on demonstration of characteristically large trachea and bronchi on plain radiograph and CT Chest.[1,8] Diagnostic criteria described by woodring et all(1999)[1,8] on X-Ray chest to detect tracheobronchomegaly includes the following:

MALES: Transverse tracheal diameter >25mm and saggital diameter >27mm.

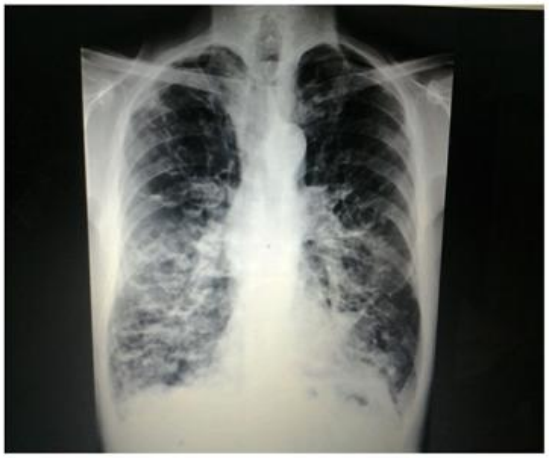
FEMALES: Transverse tracheal diameter >21mm and saggital diameter >23mm Associated central bronchiectasis, emphysema and pulmonary fibrosis can also be seen in advanced cases.

CT Chest shows dilated trachea usually >27mm, right main bronchus >21.1mm and left main bronchus >18.4mm.[1,7,9] Proximal bronchiectasis usually of cystic type is also commonly seen. Bronchoscopy may reveal dilated larger airways withsecretions.Alteration in antero-posterior diameter of the airway lumen during respiration is also observed on bronchoscopy.[1,10] 0ne third cases have tracheal diverculosis seen on scopy.[1] Spirometryusually shows obstructive pattern.

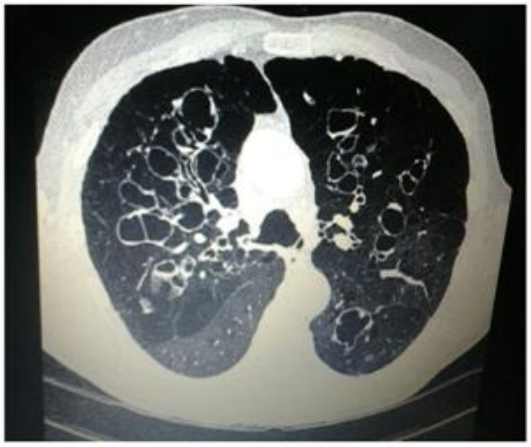
TREATMENT

Supportive therapy with antibiotics,postural drainage and bronchodilators are usually recommended.There is no role of surgery since the disease diffusely involves the airways.[1] Dynamic tracheal stent and tracheal endoprosthesis are of limited success.[1,6,7]

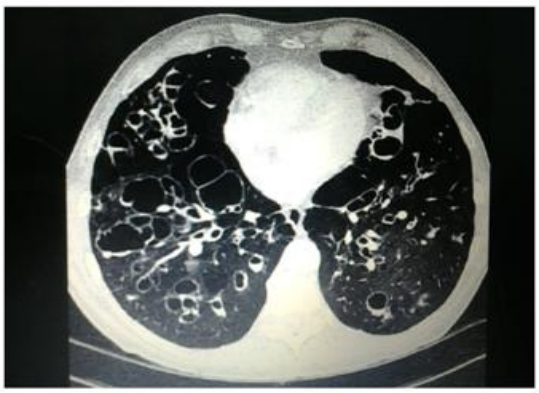
(FIG:1)Plain radiograph showing enlarged trachea and bronchi with bilateral proximal cystic lesions with air fluid level.



(FIG:2)CT Chest showing tracheobronchomegaly with bilateral proximal cystic bronchiectasis.



(FIG:3)



CONCLUSION

Due to its rarity this disease is generally overlooked even in symptomatic patients on plain radiograph. Hence a high index of suspicion is required for its diagnosis and should be considered as a differential diagnosis of proximal bronchiectasis.

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