Medico Research Chronicles

ISSN No. 2394-3971

Case Report

THE RARE CASE OF MONIER KUHN SYNDROME OR THE TRACHEOBRONCHOMEGALY SYNDROME

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Submitted on: December 2014
Accepted on: January 2015
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Abstract:
Tracheobronchomegaly is a rare disease and is essentially characterised by moderate to severe dilatation of the trachea as well as segments of the bronchial tree. The result is recurrent infections .There may be associated anomalies of the bronchial tree.
Key words: Bronchial, Trachea, Infections, syndrome

Case report:
We present the case of a 50 year old male who presented with infection of the chest several times in the last two years. He presented with dyspnea, cough. Despite repeated doses of antibiotics his condition did not improve. Sputum and cultures were negative. A CT Scan demonstrated Trachebronchomegaly.

Text:
This 50 year old male presented with infection of the chest several times in the last two years. For the previous ten years he was having recurrent chest infections and was tasking erratic antibiotics. This time he presented with dyspnea, cough. Despite repeated doses of antibiotics his condition did not improve. Sputum and cultures were negative. A CT Scan demonstrated Trachebronchomegaly. Lab features suggest

- Temp: 98.6^{0}F
- BP: 126/78
- RR: 12/Min
- Pulse 82 bpm
- HB: 11.7 gm/dl
- WBC : 7200 / microlitre
- Platelets : 2,30,000/microlitre (n 150000-400,000)
- Anti CCP antibody Positive
- Sodium: 144meq/L (n 135-145)
- Potassium : 4 meq/L(n 3.5-5)

Normal Histology:
The Walls of the Trachea are formed of four layers:

1. Mucosa
2. Submucosa
3. Fibro cartilaginous coat
4. Fibrosa

Mucosa: is formed of Epithelium lining the trachea is typical respiratory epithelium (ciliated pseudo stratified columnar), which, like the nasal epithelium, contains numerous

cells. Loose connective tissue (the lamina propria) make up the tracheal mucosa containing blood vessels, nerves and Lymphatics. Submucosa: There are seromucous glands or tracheal glands plus solitary lymphatic follicles, blood vessels and nerves. The mucosa is separated from the submucosa by a layer of longitudinal elastic fibers. Fibro cartilage coat is made of C-shaped rings of **hyaline cartilage** which help to keep the lumen of the trachea from collapsing. Fibrosa: Outermost is a layer of connective tissue is the adventitia or fibrosa. The trachea ends by dividing into two main brochi. Extra pulmonary bronchus similar to trachea.

Intra pulmonary Bronchus differs from trachea in:

- Having narrow lumen
- Highly folded mucosa
- Less goblet cells
- Complete smooth muscle coat
- Irregular cartilage plates instead of C shaped cartilage

The Lung consists of Bronchi and Bronchioles. Plus Respiratory bronchioles, alveolar ducts, alveolar sacs and alveoli

Features of Bronchi are:

1. Bronchi are two in number with open lumen.
2. Mucosa is folded and lined by pseudo stratified columnar ciliated epithelium with goblet cells.
3. The adventitia contains lymphoid follicles
4. Irregular cartilaginous plates are present
5. Mucoserous glands are present

Features of Bronchioles are:

1. Bronchioles are MANY in numbers.
2. Mucosa is folded and lined by simple columnar ciliated epithelium with NO goblet cells.
3. The adventitia contains NO lymphoid follicles
4. Irregular cartilaginous plates are NOT present
5. Mucoserous glands are NOT present

Respiratory Bronchiole is lined by simple cuboidal epithelium

Alveolar duct is lined by simple cubical epithelium.

The alveoli and alveolar sacs are lined by simple squamous epithelium separated by CT septae called Inter alveolar septum.

Histopathology:

The Histopathology of Tracheobronchomegaly involves lack of smooth muscle and elastic connective tissue in trachea and bronchial tree with sacculations in between the cartilaginous parts. There is subsequent dilatation of the trachea[1, 2, 3, and 4] as well as main stem bronchi. The Condition is seen to be associated with other conditions like Ehler Danhlos syndrome, Marfans Syndrome, Ankylosing spondylitis, cutis laxata. However sporadiac cases are also seen. The Clinical symptoms range from severe and recurrent chest infections to asymptomatic clinical condition not detected until late.

Histopathological examination can reveal either Atrophy and/or absence of longitudinal elastic fibers in the airway wall or Thinning of muscularis mucosa or absent myenteric plexus or Absence of cartilaginous elements or variations in all these changes.

Conclusion:

The Rare condition can be a cause of recurrent chest infections and can be a differential diagnosis of recurrent chest infections and should be borne in mind.

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Figures:

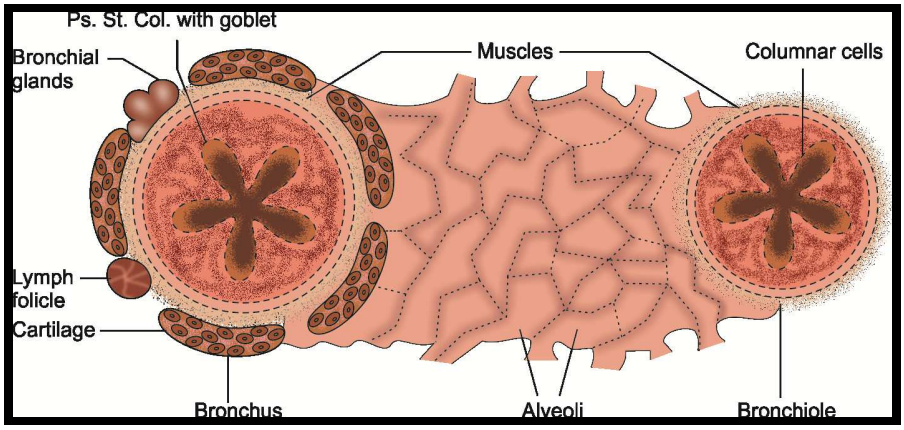

Fig 1: Normal Histology of Tracheobronchial tree

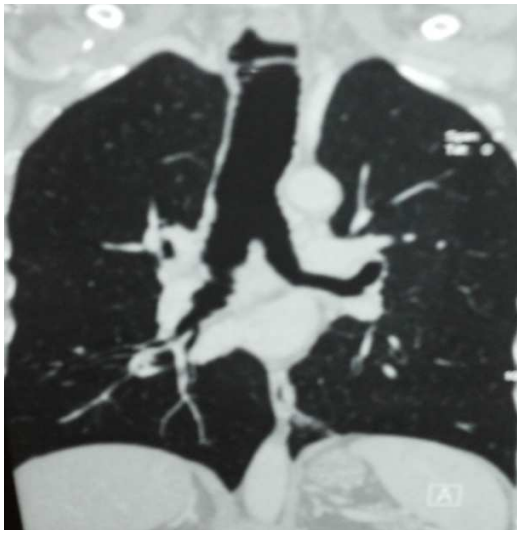

Fig 2: CT Scannogram Showing TracheoBronchomegaly

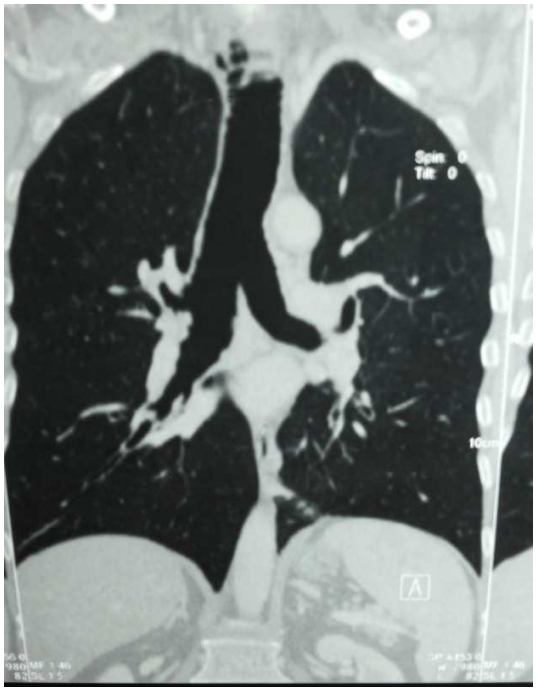

Fig 3: CT Scannogram Showing TracheoBronchomegaly