

## Image in Medicine

**Bhoomi Angirish<sup>1</sup>, Bhavin Jankharia<sup>2</sup>**

### Quiz 1

**CT scan images of a 54 year old woman who presented with sudden onset breathlessness. She had no past history of tuberculosis.**

**Questions :**

- (1) What is the diagnosis?
- (2) What is the etiology of this condition?
- (3) What are the differential diagnosis?

**Answers :**

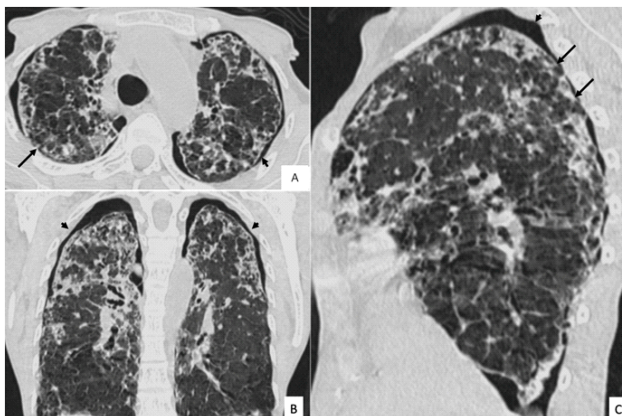
(1) Subpleural reticular opacities associated with mild traction bronchiectasis are seen in both upper lobes. Bilateral pneumothorax is also seen. Biopsy showed

fibrosis with elastin, confirming the diagnosis of pleuroparenchymal fibroelastosis (PPFE).

(2) Pleuroparenchymal fibroelastosis (PPFE) is rare pulmonary fibrosis that is clinically characterized by upper-lobe predominant fibrosis. As compared to idiopathic pulmonary fibrosis (IPF) where the fibrosis is subpleural basal, PPFE presents with upper-lobe-predominant subpleural fibrosis.

PPFE may be idiopathic or may be secondary to a variety of conditions such as connective tissue diseases, following chemotherapeutic drugs or autologous stem cell transplantation.

(3) In our country, tuberculosis is the most common differential diagnosis of upper lobe predominant fibrosis. The other common differentials are interstitial lung disease due to connective tissue disorders, sarcoidosis, drug induced lung injury.



### Quiz 2

**CT scan images of a 19 year old boy who presented with recurrent history of cough with expectoration.**

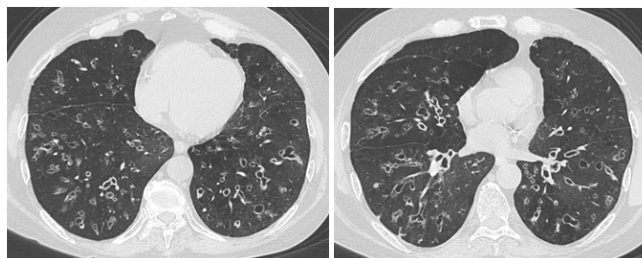
**Questions :**

- (1) What is the diagnosis?
- (2) What are the other conditions associated with this disorder?

**Answers :**

(1) Thick walled tubular and cystic bronchiectasis is seen in both the lungs. Further work up was done, which confirmed the diagnosis of cystic fibrosis.

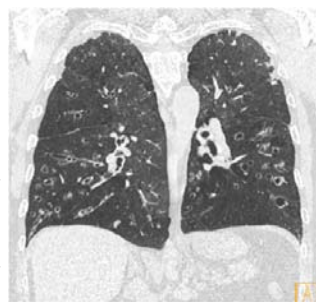
(2) Cystic fibrosis is an autosomal recessive genetic disorder due to homozygous defect of the CFTR gene that encodes for a transmembrane protein (CFTR) which is responsible for regulating chloride passage across cell membranes.



It affects the exocrine function of the lungs, liver, pancreas, small bowel, sweat glands and genital system.

**P u l m o n a y** manifestations are bronchiectasis, pneumothorax, pulmonary artery hypertension.

Abdominal manifestations are fatty replacement of pancreas, pancreatitis, pancreatic cysts, hepatic steatosis, cholelithiasis, sclerosing cholangitis. Gastrointestinal tract manifestations are distal intestinal obstruction, meconium ileus. Urogenital track manifestations are seminal vesicle agenesis, testicular microlithiasis.



Department of Radiology, Picture This by Jankharia, Mumbai, Maharashtra 400004

<sup>1</sup>MD, DNB (Radiology)

<sup>2</sup>MD, DMRD (Radiology)