

## Image in Medicine

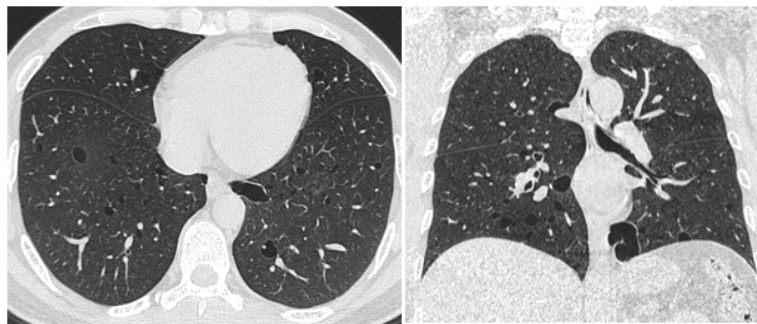
Bhoomi Angirish<sup>1</sup>, Bhavin Jankharia<sup>2</sup>

### Quiz 1

CT scan images of a 46 year old man presenting with occasional cough.

#### Questions :

- (1) What is the diagnosis?
- (2) What is Birt-Hogg-Dube syndrome (BHD)?
- (3) How to differentiate BHD from Lymphangiomyomatosis (LAM)?



#### Answers :

(1) Multiple lenticular shape cysts are seen in both the lungs, larger and more numerous in the lower lobes. These findings are in favour of Birt-Hogg-Dube syndrome (BHD).

(2) BHD is an autosomal dominant syndrome characterized by cutaneous fibrofolliculomas, multiple lung cysts, spontaneous pneumothorax, renal cysts and renal tumours, but the BHD syndrome may occur without other systemic manifestations. This is more common in men and patients are typically folliculin gene (FLCN) positive. BHD being a rare disease, is potentially underdiagnosed or misdiagnosed entity. However accurate and

timely diagnosis is important because such patients have a high risk for renal cell carcinoma and spontaneous pneumothoraces.

(3) BHD and LAM have similar clinical manifestations and hence it is important to differentiate the two conditions as their prognosis and management is quite different. Compared to LAM, cysts in BHD have a more irregular shape with internal septations and had a greater maximum size. In BHD, the cysts are fewer in number and distributed more in lower lobes and laterally. The larger cysts in BHD (>2cm) are subpleural and have concave indentation of mediastinal fat. Air cuff sign was also observed in BHD.

### Quiz 2

A 65 year old lady presented with swelling around middle phalanx of 2<sup>nd</sup> finger.

#### Questions:

- (1) What is the diagnosis ?
- (2) What are the commonly involved locations?
- (3) What is the role of dual energy CT?



#### Answers :

(1) Well defined punched out erosion with sclerotic margins is seen involving head of 1<sup>st</sup> metatarsal (Figure A). These imaging findings are in favour of gout.

(2) Gout has an asymmetrical polyarticular distribution. The most commonly involved joint is the 1<sup>st</sup> metatarsophalangeal joint.

Other joints can also be involved. Monosodium urate crystal deposition can also occur in tendons and bursae.

(3) Dual-energy CT may be used to differentiate uric acid from calcium in musculoskeletal tissue, allowing gouty urate crystals to be distinguished from bone or dystrophic calcifications. Postprocessing of dual-energy CT data yields color-coded cross-sectional images (Fig B). DECT also provides sensitive and specific volumetric quantification of tophi which is important in monitoring treatment response.

Picture This by Jankharia, Mumbai, Maharashtra

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

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