Progression of a rare multiple cystic lung disease

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A 59-year-old male, a reformed smoker, presented with complaints of exertional breathlessness, dry cough and easy fatigability for 7–8 years. He had no other comorbidities. His physical examination was unremarkable. Complete blood counts, ECG, urine analysis, renal as well as hepatic functions were also within normal limits. He was earlier evaluated with chest radiographs and computed tomography (CT) thorax in 2009, 2012, and 2015. Chest X-ray in 2009 showed bilateral lower zone reticulonodular shadows [Figure 1a] and subsequent chest X-rays in 2012 [Figure 1b] and 2015 [Figure 1c] showed an increase in these shadows. CT thorax in 2009 [Figure 2a] showed diffuse centrilobular nodules and cystic areas. CT thorax done in 2015 [Figure 2b] revealed replacement of these centrilobular nodules by cystic spaces and enlargement of existing cysts with coalescence.

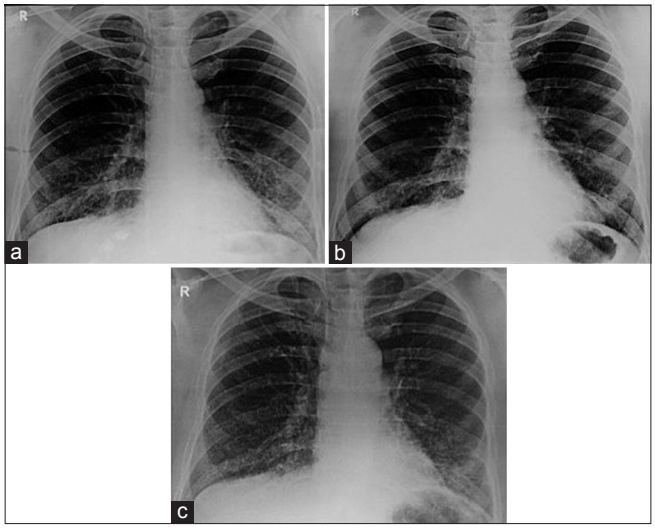

Figure 1: (a) Chest X-ray in 2009 showed bilateral lower zone reticulonodular shadows, (b) chest X-ray in 2012, and (c) chest X-ray in 2015 showed an increase in these shadows

QUESTIONS

1. What is the radiological diagnosis?
2. What is the differential diagnosis?
3. What is the most probable final diagnosis in this patient?
4. What are the radiological features of this condition?

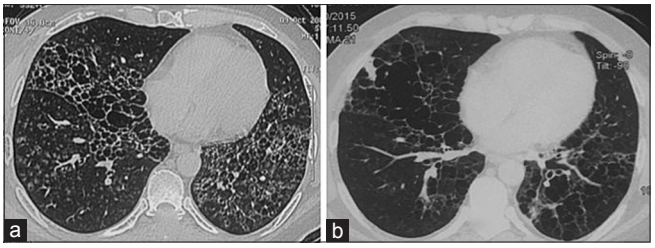

Figure 2: (a) Computed tomography thorax in 2009 showed diffuse centrilobular nodules and cystic areas, (b) computed tomography thorax in 2015 revealed replacement of these centrilobular nodules by cystic spaces and enlargement of existing cysts with coalescence

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ANSWERS

Answer 1: Multiple cystic lung disease.

Answer 2: Differential diagnosis of multiple cystic lung disease:[1]

A. Lymphangioleiomyomatosis
B. Pulmonary Langerhans cell histiocytosis (PLCH)
C. Folliculin gene-associated syndrome:
    1. Brit–Hogg–Dube syndrome
    2. Familial spontaneous pneumothorax
D. Lymphocytic interstitial pneumonia
E. Pneumocystis jiroveci pneumonia
F. Amyloidosis
G. Light chain deposition disease
H. Neoplastic disease
I. Other rare causes:
    1. Desquamative interstitial pneumonia, hypersensitivity pneumonitis, bronchiolitis
    2. Hereditary: Marfan syndrome, Ehlers–Danlos syndrome, neurofibromatosis 1 and Proteus syndrome
    3. Infectious causes: resolving paracoccidioidomycosis/coccidiomycosis
    4. Lung parenchymal involvement in tracheobronchial papillomatosis
    5. Idiopathic.

Answer 3: PLCH.

This is an uncommon clinical entity, usually occurring in the age group of 20–40 years with history of smoking. Its exact incidence and prevalence is not known. One-third of these patients are asymptomatic while symptomatic patients usually present with nonspecific respiratory symptoms such as dyspnea on exertion, cough, and fatigability. However, acute presentation in the form of pneumothorax can occur in 10%–20% of these patients. Extrapulmonary symptoms can occur in 10%–15% of these patients with skin rashes, lymphadenopathy, bone pains, or diabetes insipidus (hypothalamic involvement).[2]

Our patient was an ex-smoker with the complaints compatible with the diagnosis of PLCH.

Answer 4: Chest X-ray is often abnormal with nodular or reticulonodular shadows in early disease while advanced disease is manifested as prominent cystic appearance.

High-resolution computed tomography should be obtained in suspected patients. In the early disease, it shows multiple nodules in the size range of several mm to 2 cm with cavitation in some. In later stages, the nodules are less common, bizarre shaped and thin-walled cysts predominate. These cysts and nodules follow an apicobasal gradient of severity. They are larger and more numerous in the upper lobes than in the lower lobes with sparing of the costophrenic angles, lung bases, and anterior tips of the right middle lobe and lingula.[1-3]

The radiographic features in our patient are compatible with the diagnosis.

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Conflicts of interest
There are no conflicts of interest.

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