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Cardiopulmonary Imaging *Pictorial Essay*

Pictorial review of computed tomography features of diffuse cystic lung disease

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ABSTRACT

Diffuse cystic lung disease represents a diverse group of uncommon disorders that has been increasingly diagnosed due to the increasing use of computed tomography. It poses a frequent diagnostic challenge to radiologists due to the similar clinical and radiological features these diseases share. This pictorial review includes selected cases from the Hospital Authority New Territories West Cluster (NTWC) in Hong Kong from the past 3 years. It will illustrate the spectrum of diffuse cystic lung diseases, including some rarer entities in our locality, and identify the pertinent differentiating imaging features on CT. A flowchart to summarize these features is provided at the end to aid in diagnosis.

Keywords: Diffuse cystic lung disease, Diffuse parenchymal lung disease, Computed tomography

INTRODUCTION

Diffuse cystic lung disease represents a diverse group of uncommon disorders that has been increasingly diagnosed due to the increasing use of computed tomography (including high-resolution CT) in recent decades. It affects patients from different age groups and can be associated with syndromal disorders. CT remains the main investigation of choice for diffuse cystic lung disease. It poses a frequent diagnostic challenge to pulmonologists, pathologists, and radiologists due to the similar clinical and radiological features these diseases share. Some of these patients are asymptomatic, with the diseases discovered only incidentally, further increasing the difficulty of diagnosis and formulation of the subsequent management plans.

This pictorial review includes selected cases from the Hospital Authority New Territories West Cluster (NTWC) in Hong Kong from the past 3 years. It will illustrate the spectrum of diffuse cystic lung diseases, including some rarer entities in our locality, and identify the pertinent differentiating imaging features on CT. This study was conducted in accordance with the Declaration of Helsinki and received local Institutional Review Board (IRB) approval (NTWC/REC/19132).

Lymphangioleiomyomatosis (LAM)

Lymphangioleiomyomatosis is a well-described disorder that occurs almost exclusively in women of child-bearing age. It can be sporadic or in association with tuberous sclerosis complex. The cysts seen on imaging are thought to be caused by peribronchiolar proliferation, leading to air trapping.[1]

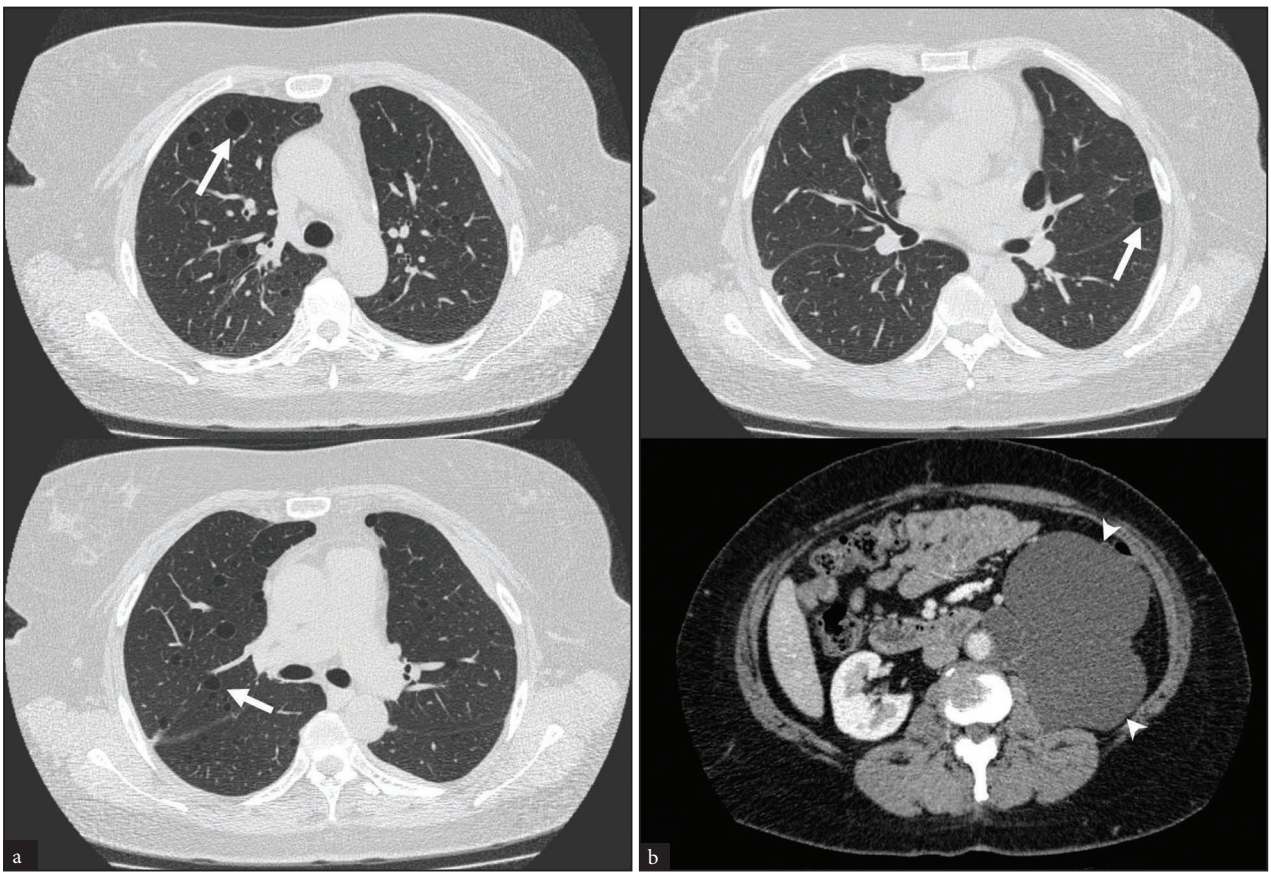

Figure 1a-b: A 43-year-old woman, who presented with hypovolemic shock from a bleeding retroperitoneal lesion, underwent a post-operative follow-up CT study. (1a) Axial CT images of the thorax revealed thin-walled cysts in a diffuse distribution, without lobar or zonal predominance, involving both central and subpleural regions (arrows). Subsequent wedge resection of blebs in the right lung confirmed diagnosis of LAM. (1b) Large lobulated cystic lesions had also recurred in the left retroperitoneum (arrowheads). They were suspected to be lymphangiomas.

CT features include relatively homogenous thin-walled rounded cysts (usually 2–5 mm) involving the entire lung diffusely without zonal predominance. The intervening lung is typically normal. Chylous pleural effusion or ascites can also be present due to lymphatic obstruction.[2]

Patients typically present with recurrent pneumothoraces and shortness of breath. In patients with tuberous sclerosis complex, other features of the disease, such as renal angiomyolipoma, may be detected [Figure 1].

Pulmonary Langerhans Cell Histiocytosis (PLCH)

Pulmonary Langerhans Cell Histiocytosis is a smoking-related lung disease, with over 90% of cases seen in patients who are young active smokers or have a past history of smoking. Peribronchiolar infiltration of the Langerhans inflammatory cells results in stellate centrilobular nodules, which thicken and then cavitate, subsequently forming cysts with variable wall thickness. Both nodules and cysts can coexist at the same time, depending on the stage of the disease. CT features include centrilobular nodules which can be cavitary and bizarre-looking lung cysts with irregularly thickened walls. These nodules and cysts have an upper lobe predilection and typically spare the costophrenic sulci [Figure 2]. Spontaneous and recurrent pneumothoraces are known complications.[3,4]

Lymphocytic Interstitial Pneumonia (LIP)

Lymphocytic Interstitial Pneumonia is a benign lymphoproliferative disorder and interstitial lung disease, commonly associated with autoimmune diseases, especially Sjögren syndrome.[5]

CT features include ground-glass opacities, poorly defined centrilobular nodules, and thin-walled cysts with a basal predominance (ranging from 1–30 mm). The perilymphatic interstitium, including the interlobular septa and pleura, are the most affected. The cysts are usually fewer in number when compared to LAM or PLCH.[6] Additional findings include interlobular septal thickening and areas of ground-glass

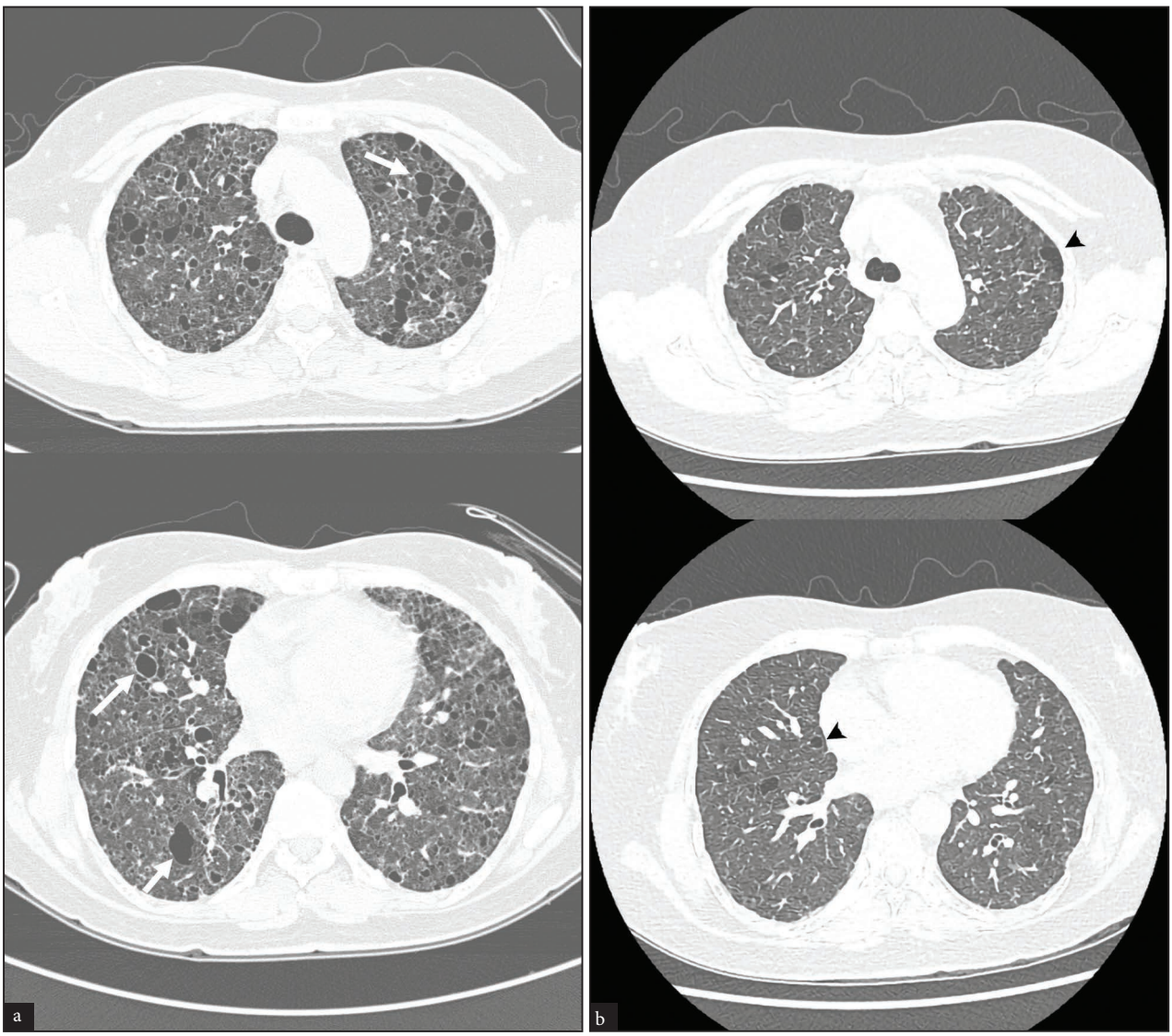

Figure 2a-b: A 56-year-old woman who was a smoker when she presented with chronic cough complicated with episodes of spontaneous pneumothorax 15 years ago was found to have cystic lung disease. (2a) Axial CT images of the thorax demonstrated diffuse cysts involving bilateral lungs without zonal predilection (arrows). There were mild subpleural fibrotic changes. No centrilobular nodules or cavitation were seen. Open lung biopsy confirmed the diagnosis of pulmonary LCH. The patient subsequently stopped smoking. (2b) Follow-up CT showed significant interval reduction of cysts with relative sparing of the costophrenic angle sulci (arrowhead), likely attributed to smoking cessation.

opacities [Figure 3]. Important differential diagnoses include other pulmonary lymphoproliferative diseases such as follicular bronchiolitis, lymphoma, and focal lymphoid hyperplasia (previously known as pseudolymphoma).

Birt-Hogg-Dubé Syndrome (BHD)

Birt-Hogg-Dubé Syndrome is a rare autosomal-dominant multisystemic disorder, with most of the clinical findings observed in the skin, lungs, and kidneys. Some patients can have only pulmonary manifestations with no other findings. CT of the thorax reveals bullous emphysema and thin-walled cysts. Cysts are bilateral and subpleural, showing a wide range of sizes, from a few millimeters to several centimeters in size (maximum diameter 2–8 cm). The cysts can be septated and lentiform or irregular in shape. Larger cysts are often distributed in the lower zone and have a lobulated, multi-septated appearance.[7] The intervening lung parenchyma is generally normal [Figure 4]. Patients can develop spontaneous pneumothoraces.

Pneumocystis Jirovecii Pneumonia (PJP)

Pneumocystis jirovecii pneumonia (previously *Pneumocystis carinii*, PCP) is a fungal infection that is strongly associated with immunocompromised diseases, such as human immunodeficiency virus (HIV) infection.

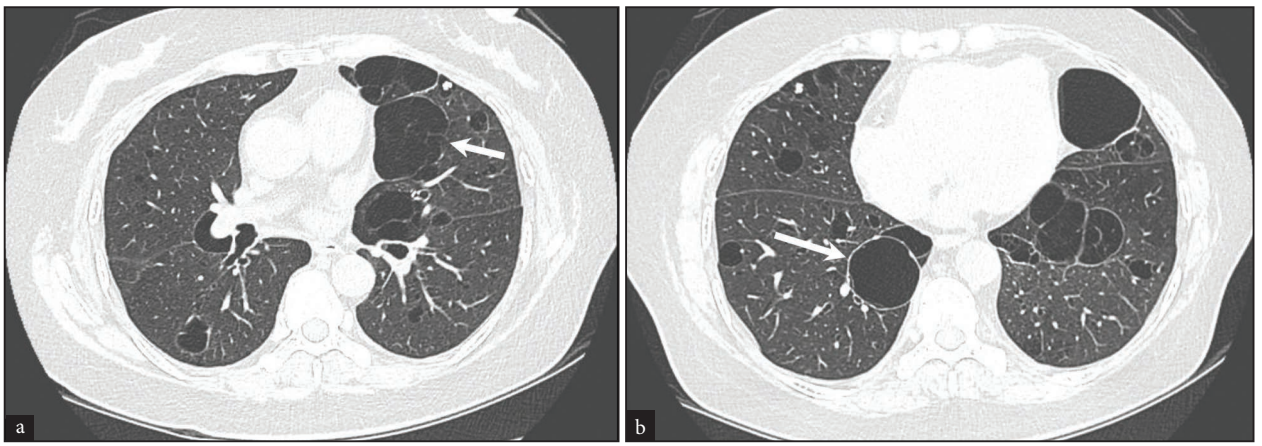

Figure 3a-b: A 52-year-old woman with history of SLE and secondary Sjögren syndrome had incidental findings of cystic lung disease on CT coronary angiography for suspected ischemic heart disease. (3a, b) Axial CT images of the thorax revealed thin-walled cysts of variable size (up to 5.4 cm), mostly seen in the perifissural and subpleural aspects, some containing thin septa or calcifications (arrows). The radiological pattern was suggestive of underlying LIP in this patient given the significant past history.

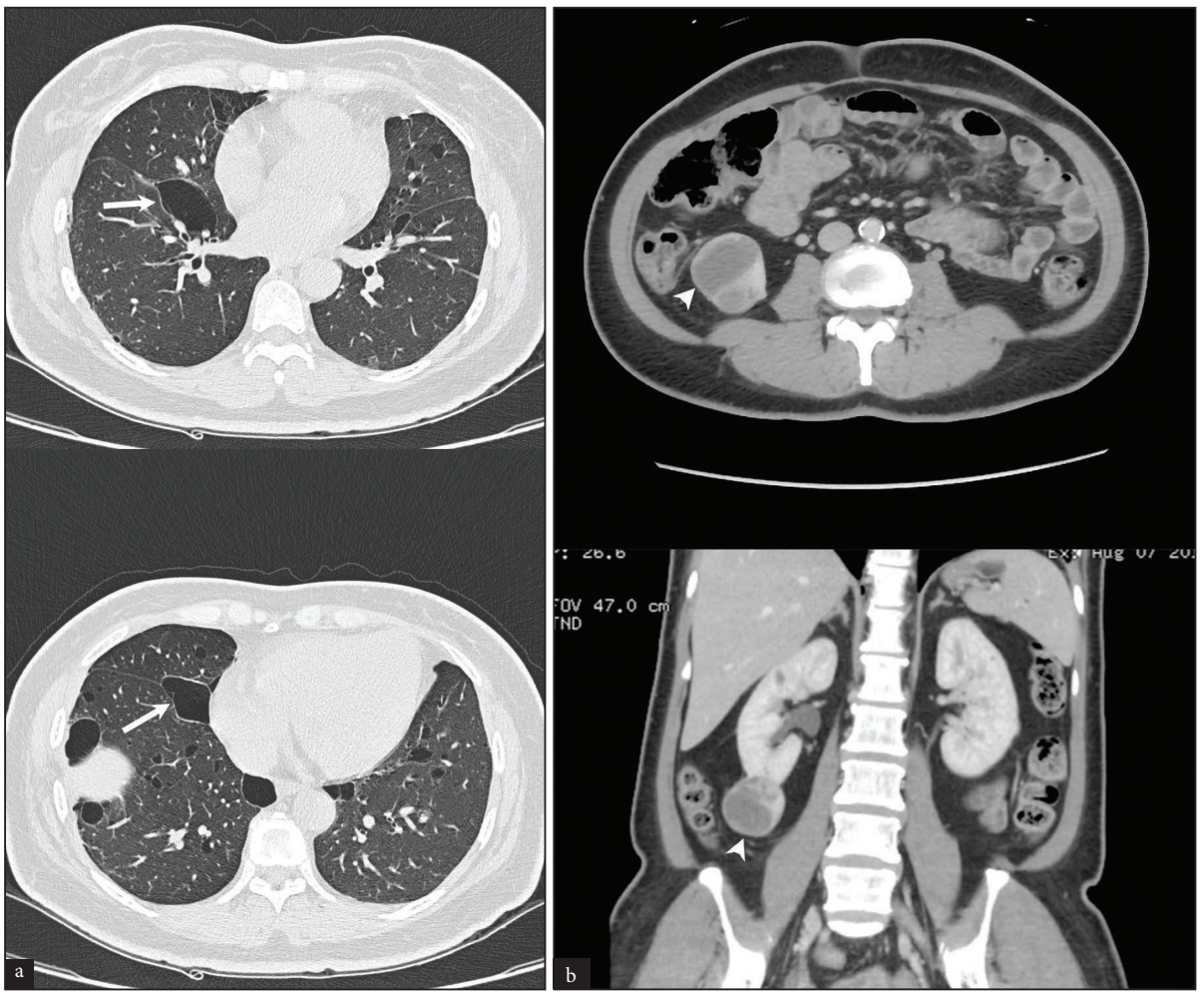

Figure 4a-b: A 44-year-old woman who initially presented with recurrent episodes of pneumothoraces had a chest radiograph showing presence of lung cysts. (4a) Axial CT images of the thorax revealed thin-walled cysts of variable size with lower zonal predominance, some of them being lentiform in shape (arrows). (4b) Subsequent renal ultrasound (not shown) and CT of the abdomen showed right renal mass which was heterogeneously enhancing on CT (arrowheads). There was no fat component detected. It was eventually proven to be an oncocytoma on histology.

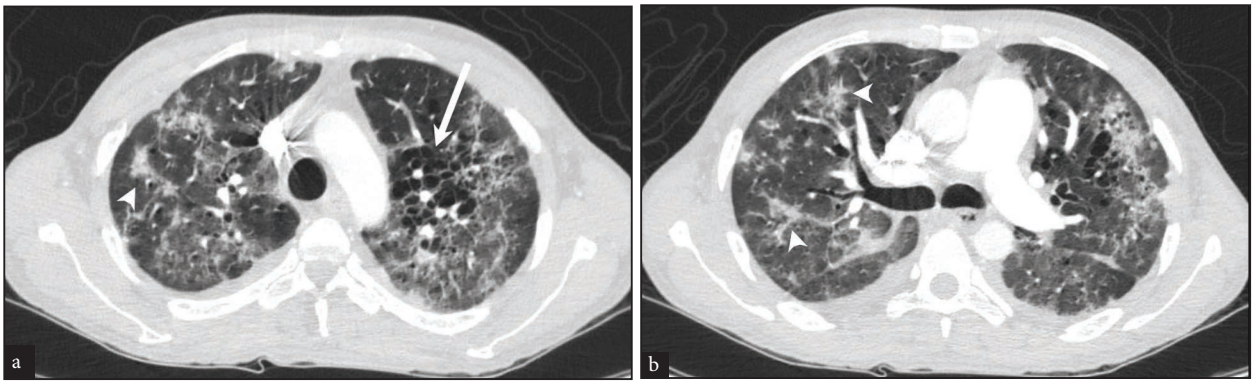

Figure 5a-b: A 34-year-old man with known HIV and repeated PCP infection presented with shortness of breath and cough. (5a, b) Axial CT images of the thorax showed multiple cysts with upper zonal predominance in the central aspects of the lungs (arrow), associated with patchy ground-glass opacities and intralobular septal thickening (arrowheads).

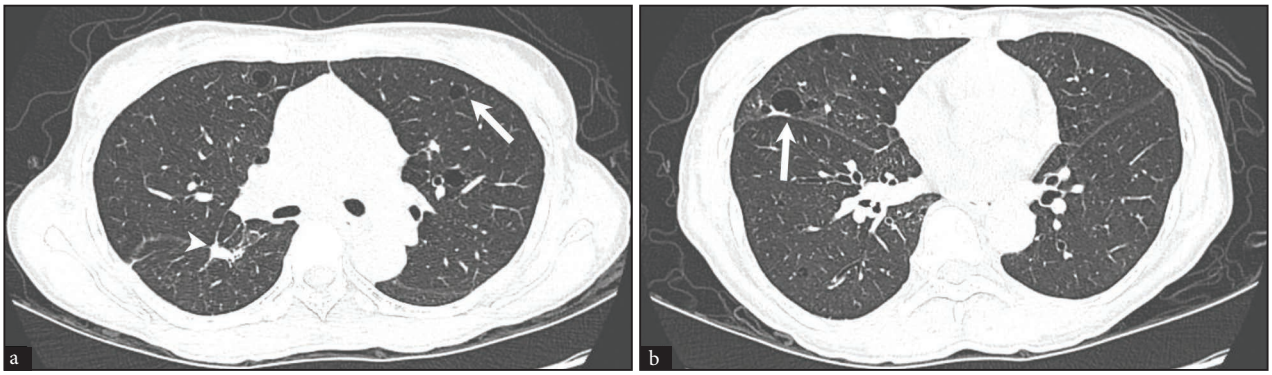

Figure 6 a-b: A 58-year-old woman, with a medical history of SLE and thyroid cancer with thyroidectomy done, presented with presumed chest infection. (6a, b) Chest CT found multiple small bilateral lung nodules (arrowhead) and small lung cysts (arrows) of variable size in random distribution. Mild bronchiectasis was also seen in the upper lobes. Bronchoscopic biopsy of the lung nodules confirmed pulmonary amyloidosis.

Table 1: Table summarizing the radiological features of mimickers of cystic lung disease on CT.

Mimickers of cystic lung disease	Radiological features on CT
Pulmonary emphysema	- Imperceptible wall, paucity of vascular markings - Expanded lung volumes - Presence of bullae
Cavitating pulmonary metastasis	- Irregular, thick-walled+/- air-fluid level - Basal predominance due to vascularity - Accompanied with solid nodules and ground-glass opacities
Interstitial lung disease	- Subpleural cystic changes, honeycomb formation in late stage - Subpleural fibrosis, tractional bronchiectasis
Pneumatocele	- Smooth inner margin - Solitary rather than diffuse depending on location of insult - Tend to resolve after initial insult treated

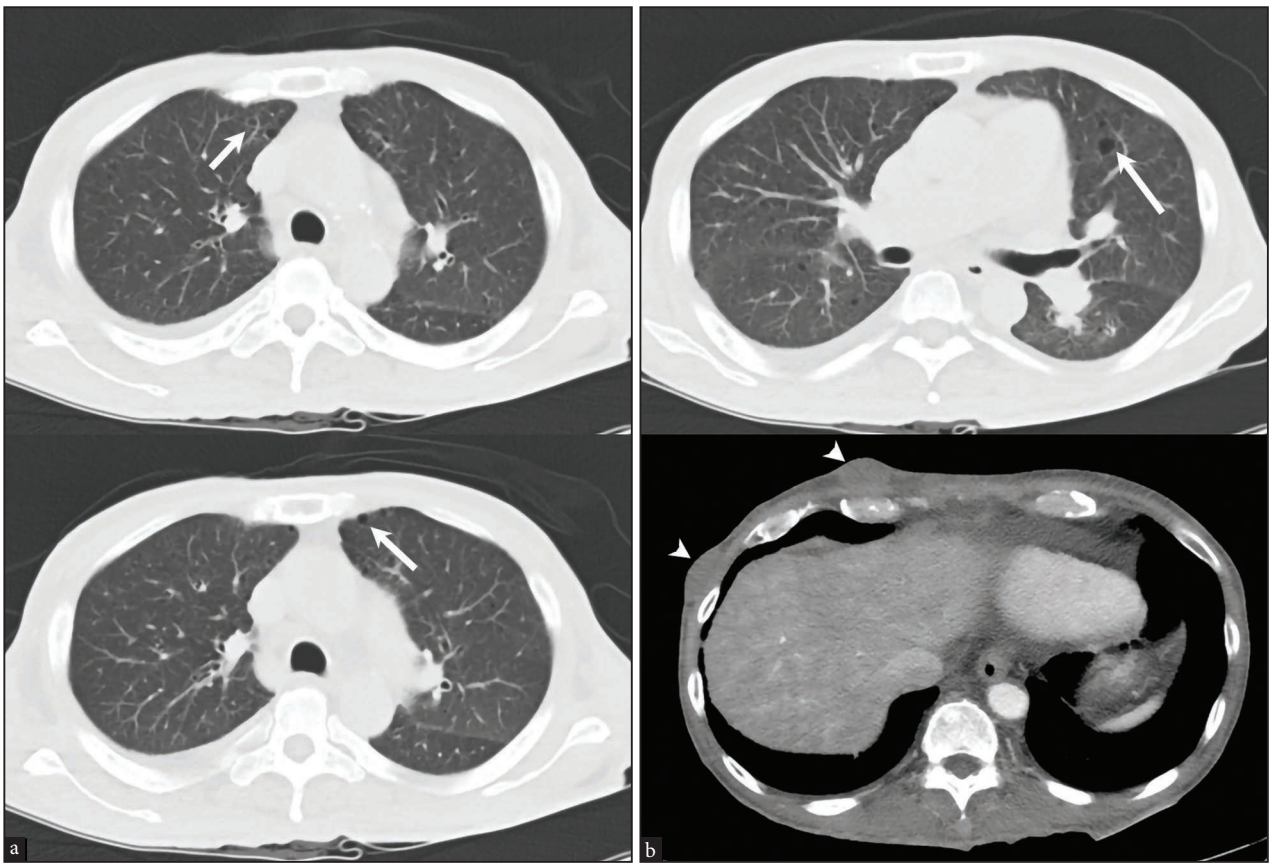

Figure 7a-b: A 59-year-old man with a past history of NF-1 diagnosed by skin café-au-lait spots and eye Lisch nodules presented with the hospital with metabolic acidosis. He was a never-smoker and had a history of chronic pulmonary embolism with right heart failure on warfarin. (7a) Urgent CT thorax, abdomen and pelvis arranged showed multiple small thin-walled cysts ranging from 2mm to 1cm in size with mid-zone and subpleural distribution (arrows). (7b) Skin nodules noted on the chest wall in the soft tissue window were compatible with clinical findings of multiple café-au-lait spots (arrowheads). He later developed refractory shock from right heart failure and succumbed.

CT features include bilateral multifocal, mainly symmetrical, ground-glass opacities distributed in the central portions of the lungs, classically at the perihilar regions as the dominant feature. Intralobular septal thickening seen together with these ground-glass opacities can give rise to a "crazy-paving" pattern.[8] Numerous pulmonary cysts are seen diffusely distributed in both lungs, or predominantly in the upper lobes. They can be of variable sizes and wall thickness [Figure 5]. These cysts can shrink or resolve upon successful treatment. The disease is also associated with an increased incidence of spontaneous pneumothoraces due to the rupture of cysts, although it can also occur without the presence of cysts.[9]

Amyloidosis

Amyloidosis is a disease characterized by the extracellular deposition of amyloid proteins, which can be localized or systemic. Pulmonary amyloidosis can rarely present as a cystic disease. When it does, these cysts tend to be multiple in number, showing around or lobulated configuration with thin walls on CT. They can have both peribronchovascular and subpleural distributions. Other imaging findings include nodules, which can be calcified, interlobular septal thickening, and lymphadenopathy [Figure 6].[10,11]

Neurofibromatosis type 1 (NF-1)

Neurofibromatosis type 1, also known as von Recklinghausen disease, is a multisystem neurocutaneous disorder and one of the most common phakomatosis. Pulmonary manifestations include diffuse cystic lung disease, emphysema and ground-glass opacities with micronodules. The thin-walled cysts seen in NF-1 show upper lobe predominance and are usually subpleural in distribution. They can also have extrapulmonary thoracic manifestations, including skin nodules and peripheral nerve sheath tumors [Figure 7].[12]

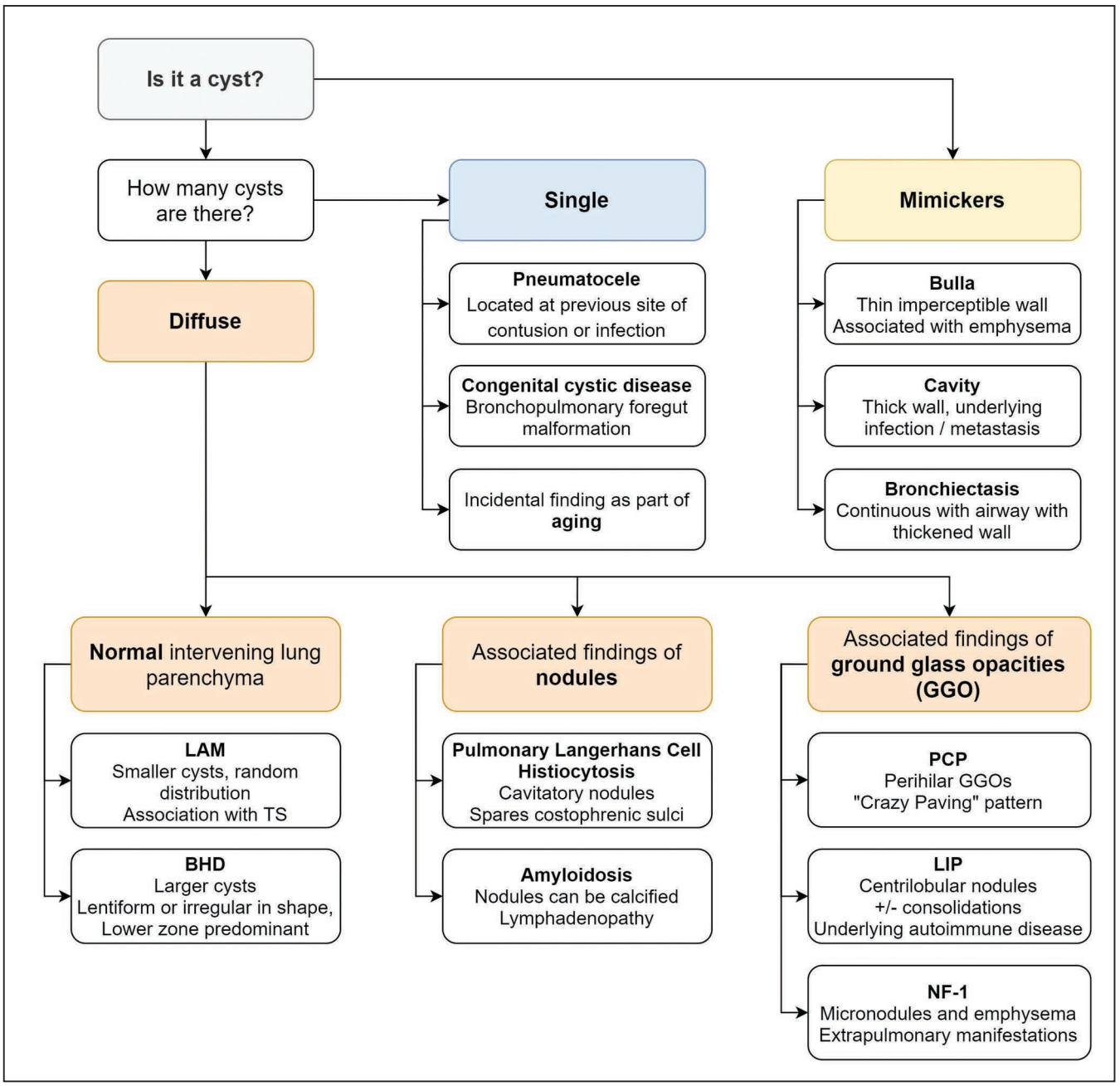

Figure 8: Flowchart for stepwise diagnostic approach on cystic lung disease on CT.

Potential mimickers of cystic lung disease

Many other diseases can also demonstrate cyst like lesions in the lungs. It is sometimes difficult to differentiate them from the diffuse cystic lung diseases mentioned above, but they can be distinguished based on certain radiological features and clinical grounds. A few of these conditions are mentioned below.

These diseases include pulmonary emphysema, cavitating pulmonary metastasis, interstitial lung disease, and pneumatocele. We have highlighted and summarized the radiological features of these diseases in Table 1.

We have created a flowchart [Figure 8] listing out the radiologic distinction of different diseases demonstrating cystic patterns and the stepwise diagnostic approach. This can

help narrow down the possible differential diagnoses, which would aid clinical management.

CONCLUSION

It is important to recognize the differential diagnoses of diffuse cystic lung disease and their distinguishing features to enable timely diagnosis and management of these patients. While the diagnosis may be challenging and the radiological features may appear nonspecific, this process can be simplified by taking a systematic approach such as the one proposed in the flowchart above.

Declaration of patient consent

Institutional Review Board (IRB) permission was obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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