2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM

History

- 23 Y/F with MMRC Grade III breathlessness, Intermittent dry cough
- She is on Long term oxygen therapy since 1 month.
- H/o childhood LRTI when 5 years old
- She had moderate COVID in 2020, When patient was diagnosed with ILD and started on Oxygen therapy
- There is no clinical history pertaining to any systemic disease

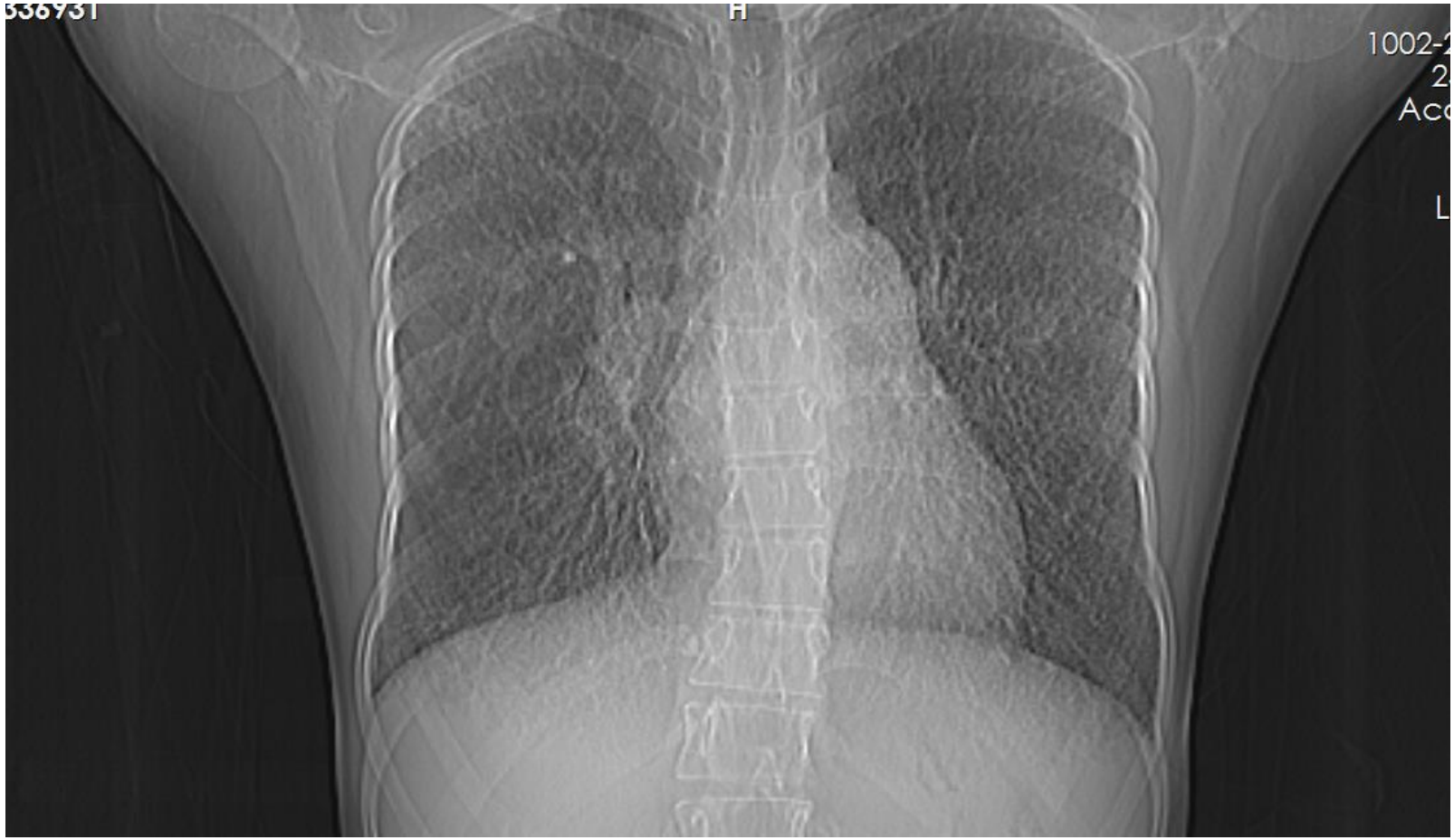
On Examination

- No cyanosis
- No tachypnea
- On auscultation: B/L BS+, B/L basal velcro crepitations
- SpO2: 82 % on room air and 96 % with 2L O2.

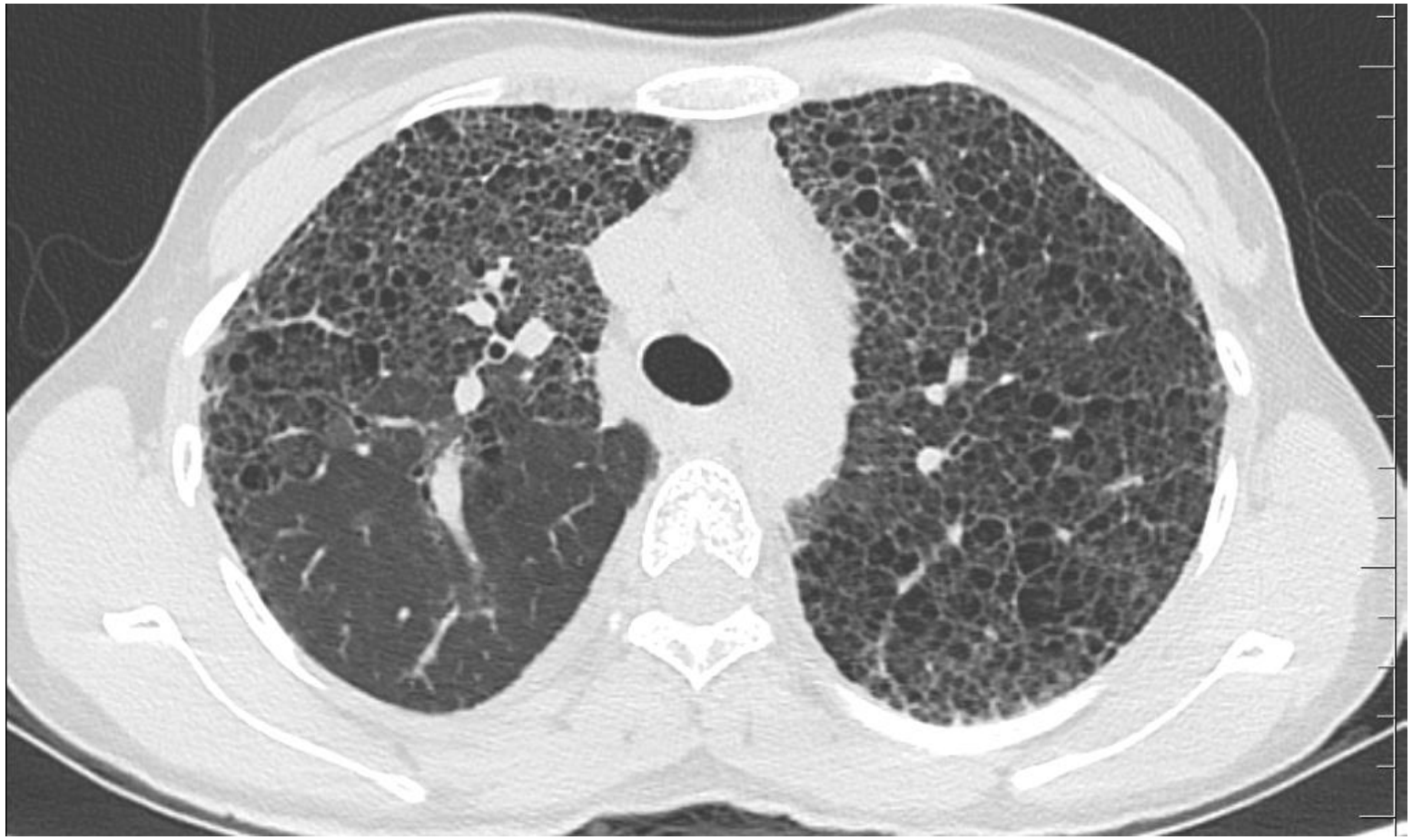
ECHO

- Dilated RA/RV
- PASP: High (76 mmHg)

Imaging: Chest X ray



Chest CT(Plain) Lung window



Summary

- Diffuse cystic changes are seen in both lungs with near total involvement of the left lung and some sparing of the right lung.
- The CP angles and medial aspect of the right middle lobe are involved.
- Diffuse increase in density of the right lung with patchy ground glass opacities of spared lung segments
- No air trapping
- No soft tissue nodule or mass lesion
- Dilated main pulmonary artery and prominent right heart chambers
- No evidence of pneumothorax or pleural effusion

→ Features are consistent with cystic lung disease(Lymphangioleiomyomatosis) with ground glass opacities representing intercurrent infection (LIP is another possibility in view of GGO)

Additional investigations

- USG- Abdomen.
- No evidence of renal masses/ ascites.
- No significant abnormalities detected
- Anti Rho, Anti La, ANAs were negative

Differential diagnosis of cystic lung disease:

-Lymphangioleiomyomatosis.
-Lymphoid interstitial pneumonia.
-Langerhans cell histocytosis.
-Pneumocystis pneumonia.
-Birt Hogge Dube syndrome

Lymphangioleiomyomatosis

Lymphangioleiomyomatosis (LAM) is a rare multi-system disorder that can occur either sporadically or in association with the tuberous sclerosis complex(TSC) and is often considered a form of TSC.

Epidemiology

It almost exclusively affects women of child-bearing age .

Clinical presentation

Patients usually present with exertional dyspnoea and recurrent episode pneumothorax are common.

Pathology

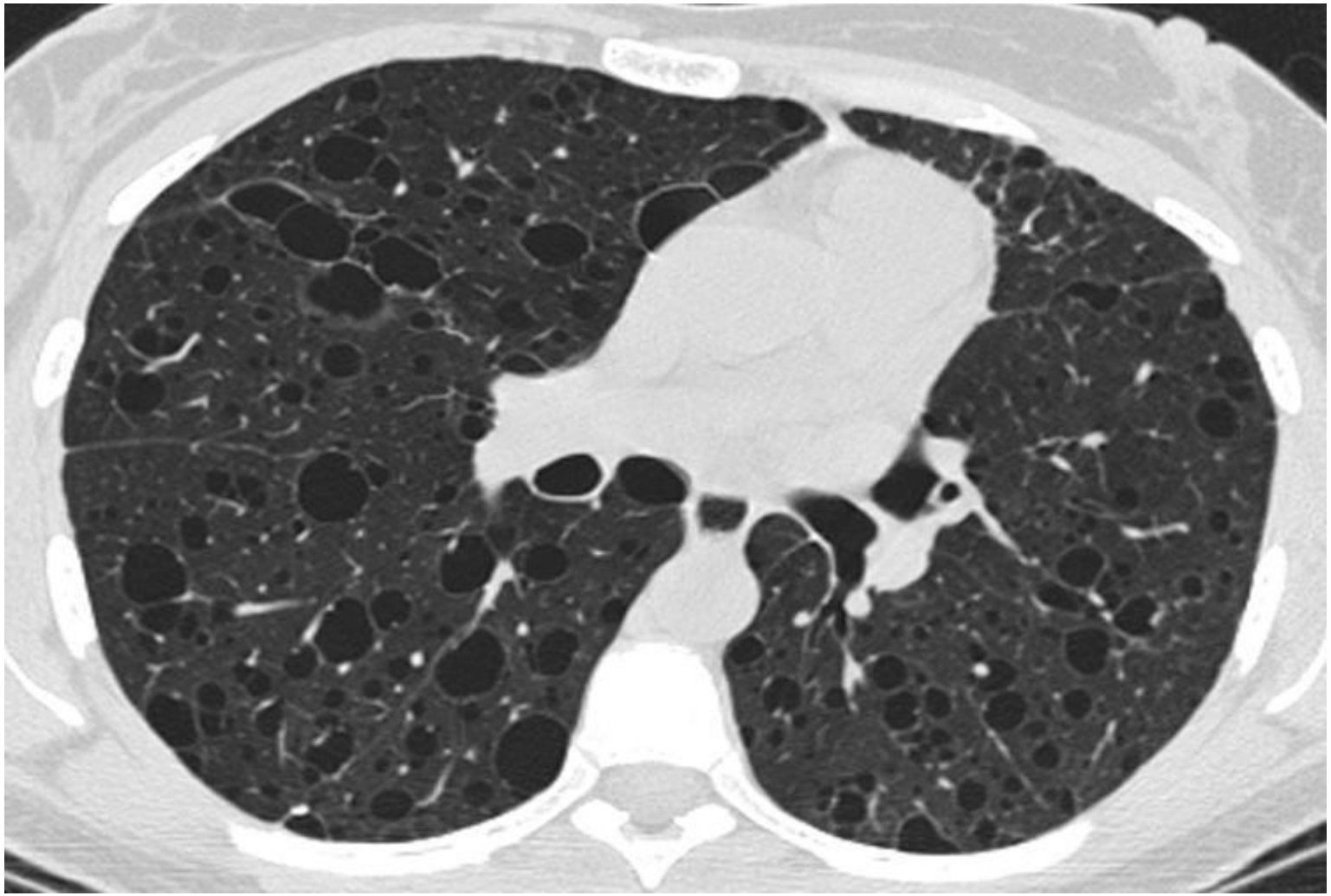
The disease is characterised by the persistence of dilated lymphatics and interstitial proliferation of abnormal smooth muscle that in turn can obstruct venules, lymphatics, and small airways.

Radiographic features

- **X ray**
  - chylothorax: chylous pleural effusion
  - evidence of hyperinflation
  - diffuse bilateral reticulonodular densities
  - recurrent pneumothoraces in complicated cases

- **HRCT**
  - Thin walled and round cysts of variable sizes surrounded by normal lung parenchyma can be seen throughout the lung- Profusely and evenly distributed throughout the lung.
  - Display limited variability in size and shape
  - May show a dilated thoracic duct
  - Haemorrhages may be seen as areas of increased attenuation

CT features



Pulmonary and associated extrapulmonary manifestations

- Prevalence of spontaneous pneumothorax is highest in LAM patients among other cystic lung diseases
- Chylous pleural effusions occur in 10 % of patients with LAM and it can be unilateral or bilateral
- Renal angiomyolipomas are demonstrated in approximately 30 % of the individuals with sporadic LAM and in 80 % pf the individuals with TSC- LAM
- LAMs of the retroperitoneum.
- Cutaneous manifestations of TSC- Malar angiofibromas, Ash leaf spots, Shagreen patches and periungual fibromas
- Neurological manifestations of TSC- Cognitive impairment , Seizures, SEGA, and subependymal nodules

Treatment and prognosis

- mTOR inhibitors- Sirolimus, Everolimus are the main treatment options
- Bronchodilators
- VATS pleurodesis for recurrent pneumothorax
- Lung transplantation for respiratory failure.

Pulmonary Langerhans cell histiocytosis

It can be seen as part of widespread involvement in patients with disseminated LCH or more frequently as a distinct entity in young adult smokers.

Epidemiology

PLCH is usually identified in young adults (20-40 years of age). A history of current or previous cigarette smoking is identified in up to 95% of cases .

Clinical presentation

Presentation is usually with dyspnoea or a non-productive cough.
Other symptoms include constitutional symptoms (fatigue and weight loss), pleuritic chest pain.
Pneumothorax and pleural effusion are less frequently associated

Pathology

Langerhans cells proliferate in the bronchiolar and bronchial epithelium, forming granulomas. It is postulated that as these cellular granulomas evolve, peripheral fibrosis forms resulting in traction on the central bronchiole which becomes cyst-like. Electron microscopy may reveal characteristic Birbeck granules.
More recent evidence suggest that PLCH represents a myeloid neoplasm with inflammatory properties.

Radiographic features

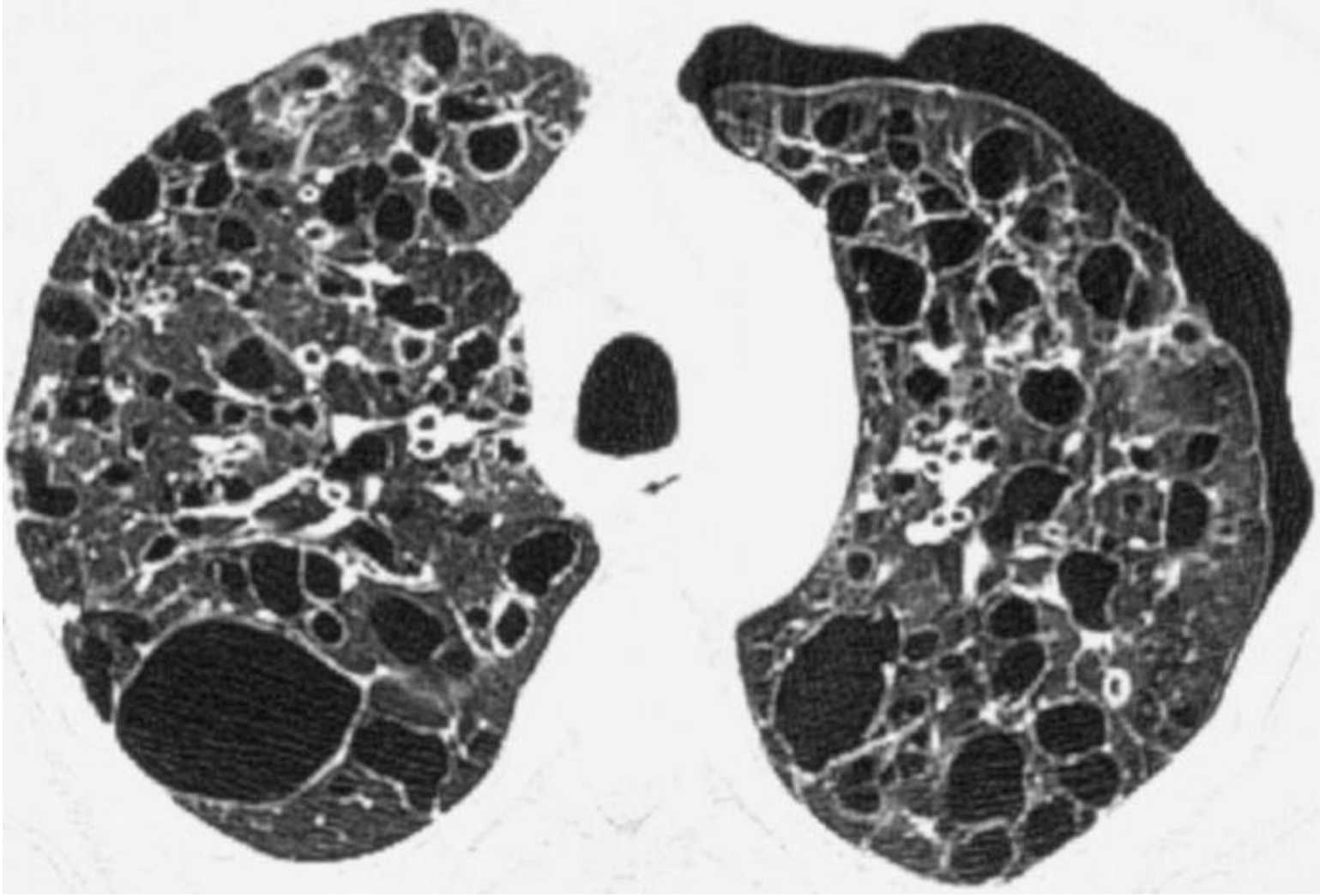
Plain radiograph

- The earliest change is a diffuse bilateral symmetrical reticulonodular pattern with a predilection for the mid and upper zones ranging from 1-10 mm in size.
- Later, cyst formation may be seen or may mimic a honeycomb appearance due to a summation of air-filled cysts.
- There is a preservation of lung volumes or even hyperinflation. Reduced lung volumes are uncommon and only seen in end-stage fibrotic cases . Lymph node enlargement visible on chest x-rays is rare .

CT

- HRCT is superior to plain chest radiography in identifying both the reticulonodular opacities and cysts .
- Distribution is the key in differentiating PLCH from other cystic lung diseases with predilection for the mid and upper zones and regional sparing of the costophrenic recesses, anterior right middle lobe and lingula left upper lobe.
- The cystic lesions of PLCH is irregular and bizarrely shaped
- The combination of nodules and cystic lesions permits a diagnosis with high degree of confidence

CT features



Nodules

- more pronounced early in the disease
- centrilobular distribution, may also be peribronchial or peribronchiolar
- usually have irregular margins
- may be cavitary nodules with thick walls, later becoming cysts
- surrounding lung parenchyma appears normal

Cysts

- more pronounced later in the disease
- may measure up to 2-3 centimetres in size
- usually thin-walled, but on occasion may be up to a few millimetres thick
- confluence of 2 or more cysts results in bizarre shapes, bilobed, cloverleaf, branching, internal septations

Other common findings include :

- ground-glass and/or reticular opacities
- mosaic attenuation
- Interlobular septal line thickening
- Emphysema

In late disease, other findings include:

- cyst coalescing
- fibrosis
- Honeycombing

The appearance of new nodules later in the disease (when cystic change is established) indicates disease progression but is a rare finding .

Associations

- Hematopoietic neoplasms:
- ALL, AML
- Cystic bone lesions
- Diabetes insipidus
- Skin lesions

Treatment and prognosis

- Overall prognosis is generally good with over 50% of patients demonstrating spontaneous resolution or stabilisation even without treatment with smoking cessation
- In those who continue to smoke, the disease is progressive with deterioration in respiratory function and eventual end-stage pulmonary fibrosis .
- Corticosteroids is mainstay of treatment
- In some selected patients lung transplantation may be an option, provided smoking has ceased. Recurrence in the transplanted lung has been described .

Complications

- cyst rupture
- spontaneous pneumothorax: may be the first presentation
- pneumomediastinum
- interstitial fibrosis
- pulmonary arterial hypertension and cor pulmonale
- end-stage pulmonary fibrosis and respiratory failure

Lymphocytic interstitial pneumonitis

Lymphocytic interstitial pneumonitis (LIP) is a benign lymphoproliferative disorder characterised by lymphocyte predominant infiltration of the lungs. It is classified as a subtype of interstitial lung disease.

Epidemiology
LIP can occur at any age. If a child presents with LIP, can be indicative of AIDS.

Clinical presentation
Gradual onset of dyspnea and cough with approximately six months duration.

Pathology
There is diffuse infiltration of the interstitium and alveolar spaces by lymphocytes and plasma cells.

Associations

- Sjogren syndrome: can occur in up to 25% of those with LIP
- AIDS: particularly if it occurs in the young
- Autoimmune thyroid disease
- Systemic lupus erythematosus (SLE)
- Castleman disease
- Common variable immune deficiency (CVID)
- Rheumatoid arthritis
- Pulmonary amyloidosis

Markers

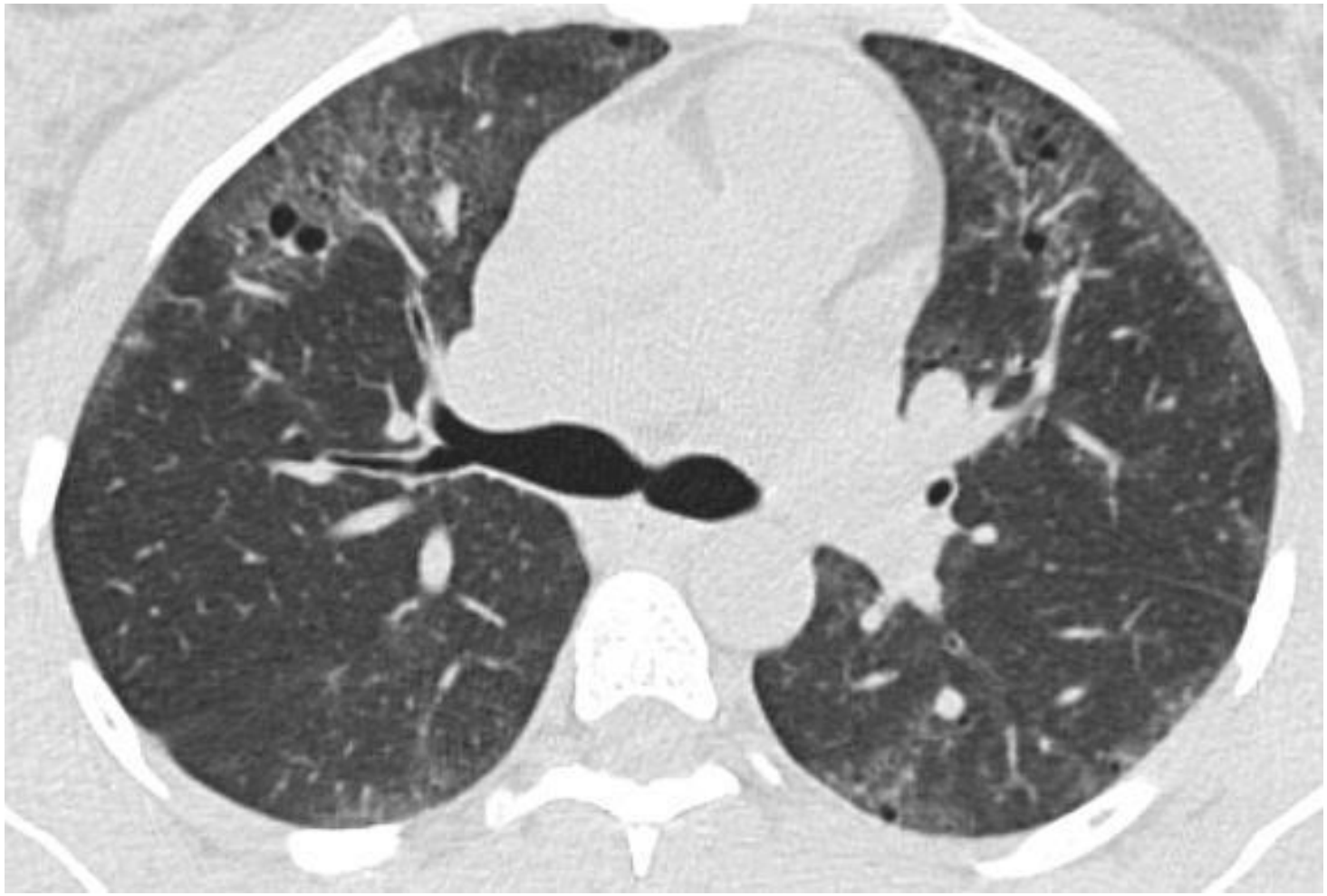
In about 80% of patients polyclonal or IgM monoclonal gammopathy is found .

Plain radiograph

- Features can be non-specific, but may include:
- lower-zone predominant bilateral reticular opacification
- chronic bilateral airspace opacification

CT

- Features tend to be diffuse with mid to lower lobe predominance
- Thickening of bronchovascular bundles
- Interstitial thickening along lymph channels
- Small but variable sized pulmonary nodules (can be centrilobular or subpleural, and often ill-defined)
- Ground-glass change
- Scattered thin walled cysts , typically few in number and seen along areas of ground glass
    - usually deep within the lung parenchyma
    - typically abuts vessels (i.e. is perivascular or subpleural)
    - size range from 1-30 mm (useful for differentiation between lymphoma of the lung)
- Mediastinal lymphadenopathy



Birt Hogge Dube syndrome

- Multisystem disease characterized by:
- Cutaneous manifestations: Fibrofollliculomas
- Multiple lung cysts and spontaneous pneumothorax
- Renal tumours, such as chromophobe oncocytomas and chromophobe RCCs

HRCT findings

- Lung cyst typically develop in adulthood
- Multiple lower zone predominant and bilateral
- Predilection for subpleural, paramediastinal and perifissural lung
- Thin walled, elongated, variable in size, Multilobulated or multiseptate, often lentiform in shape