RADIOGRAPHIC QUIZ From page 15

■ BRIEF OVERVIEW OF THE DISEASE

Usual interstitial pneumonia (UIP) is a histopathologic and radiologic pattern of interstitial lung disease, which is the hallmark pattern for idiopathic pulmonary fibrosis (IPF). On imaging, UIP usually presents with a lung volume loss and a craniocaudal gradient of peripheral septal thickening, bronchiectasis, and honeycombing.

The histological diagnosis of UIP is based on temporal and spatial heterogeneity, which is the identification of fibrotic lesions at different stages (fibroblastic infiltrates, mature fibrosis, and honeycombing) within the same biopsy specimen and architectural distortion. Honeycombing, particularly if it involves more than 5% of the lung volume, is an almost 100% specific finding. On a typical biopsy, there are areas of normal lung alternating with interstitial fibrosis and honeycombing. The distribution of UIP characteristically is with an apicobasal gradient with basal and peripheral (subpleural) predominance, although it is often patchy.

The idiopathic interstitial pneumonias (IIPs) are a group of diffuse parenchymal lung diseases that share many features but are sufficiently different from one another to be designated as separate disease entities. The general term idiopathic interstitial pneumonia includes usual interstitial pneumonia (UIP), nonspecific interstitial pneumonia (NSIP), respiratory bronchiolitis–associated interstitial lung disease (RB-ILD), desquamative interstitial pneumonia (DIP), cryptogenic organizing pneumonia (COP), acute interstitial pneumonia (AIP), lymphoid interstitial pneumonia (LIP), and idiopathic pleuroparenchymal fibroelastosis (IPPFE).

Combining patient history, physical examination, laboratory studies, imaging, and pathologic analysis allows for these entities to be distinguished from other forms of diffuse parenchymal lung disease. However, these computed tomography (CT) and histologic patterns of lung injury are frequently similar or identical to those seen in many other conditions, including connective tissue disease, drug reactions, asbestosis, and chronic hypersensitivity pneumonitis. The term idiopathic is reserved for those conditions in which the cause of the lung injury pattern is unknown.

Previous classifications of the IIPs have been replaced with a unified classification composed by the American Thoracic Society and European Respiratory Society emphasizing the

Idiopathic interstitial pneumonia (IIP)

Morphological pattern	Histologic features	Imaging features	Imaging differential diagnosis
UIP	Spatial and temporal heterogeneity, dense fibrosis, fibroblastic fosi, honeycombing	Basal, peripheral predominance, often patchy, reticular abnormality, honeycombing	Collagen vascular disease, asbestosis, chronic hypersensitivity pneumonitis
NSIP	Spatiallly and temporally homogeneous lung fibrosis or inflammation	Basal predominance, ground-glass abnormality, reticular abnormality	Collagen vascular disease, chronic hypersensitivity pneumonitis, DIP
Respiratory bronchiolitis	Peribronchiolar macrophage accumulation, bronchiolar fibrosis; macrophages have dusty brown cytoplasm	Centrilobular nodules, ground-glass attenuation	Hypersensitivity pneumonitis
DIP	Diffuse macrophage accumulation in alveoli	Basal, peripheral predominance, ground-glass attenuation; sometimes cyst	Hypersensitivity pneumonitis, NSIP
Organizing pneumonia	Patchy distribution of intraluminal organizing fibrosis in distal airspaces; preservation of lung architecture; uniform temporal appearance; mild interstitial chronic inflammation	Ground-glass attenuation; consolidation basal, peripheral predominance	Collagen vascular disease, infection, vasculitis, sarcoidosis, lymphoma, adenocarcinoma
Diffuse alveolar damage	Diffuse distribution, uniform temporal appearance, alveolar septal thickening caused by organizing fibrosis, airspace organizing, hyaline membranes	Diffuse, ground-glass attenuation, consolidation	Acute respiratory distress syndrome, infection, hydrostatic edema, hemorrhage
LIP	Diffuse lymphoplasmatic infiltration of alveolar septa	Ground-glass attenuation, cyst	DIP, NSIP, hypersensitivity pneumonitis
IPPFE	Elastotic fibrosis with intraalveolar fibrosis	Dense subpleural upper-lobe consolidation	Familial pulmonary fibrosis, chronic hypersensitivity pneumonitis

complementary roles of the pathologist, radiologist, and clinician in diagnosis. These societies convened an international committee of pulmonary clinicians, thoracic radiologists, and pulmonary pathologists to clarify disease nomenclature and patterns.
Although the classification of IIPs is rooted in histologic criteria, there is a definite recognition that the pattern at thin-section CT is important in delineating the morphology of the IIPs

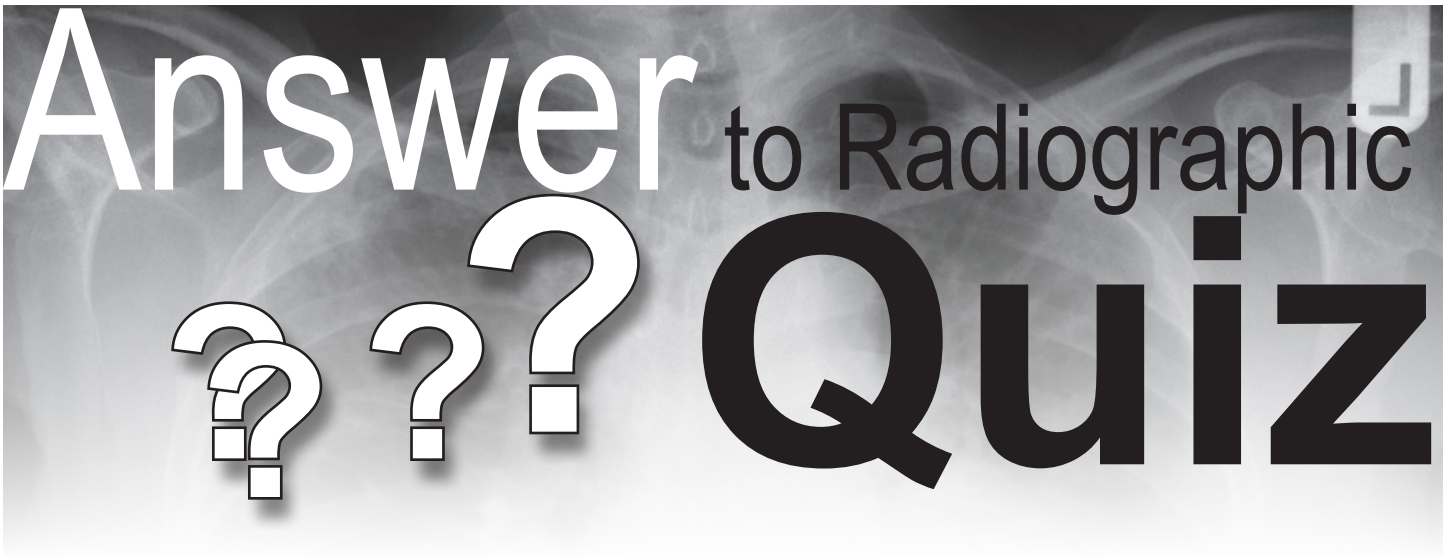
UIP is one of the most common interstitial lung diseases, and high-resolution CT (HRCT) features prominently in the American Thoracic Society (ATS) diagnostic algorithm for IPF. The diagnosis of UIP is frequently based on clinical and imaging features, without the need for surgical biopsy, because of the high accuracy of thin-section CT diagnosis in many cases of UIP
Typical CT-based morphologic patterns are associated with the IIPs, and radiologists play an important role in diagnosis and characterization. Basal and peripheral predominant reticular pattern with honeycombing and traction bronchiectasis characterizes UIP. Basal and peripheral or peribronchovascular ground-glass opacity with or without reticular pattern and traction bronchiectasis characterizes NSIP. The smoking-related lung diseases RB-ILD and DIP demonstrate centrilobular nodules and lower-lobe predominant ground-glass opacity (frequently with cysts), respectively. Patchy peripheral or peribronchovascular consolidation and ground-glass typifies COP. Diffuse lung consolidation and ground-glass opacity characterizes AIP. Ground-glass opacity and perivascular cysts typify LIP. Dense upper-lobe pleuroparenchymal fibrosis characterizes IPPFE. All IIPs should be classified using an interdisciplinary approach. Plain film features are non-specific. While chest radiographs can be even normal in patients with very early disease, in advanced disease, it may show decreased lung volumes and basal fine to coarse reticulation. Usually, due to the more extensive involvement of the lower lobes, the major fissure is shifted inferiorly which is best seen on the lateral chest radiograph.

■ REFERENCES

1. Weerakkody Y, et al. Usual interstitial pneumonia Available in: https://radiopaedia.org/articles/usual-interstitial-pneumonia.

2. Hobbs S, Lynch D. The Idiopathic Interstitial Pneumonias. An Update and Review. Radiol Clin N Am 52 (2014) 105–120

3. Sverzellati N, Lynch D, Hansell DM, Johkoh T, King TE, Traveis WD. American Thoracic Society–European Respiratory Society Classification of the Idiopathic Interstitial Pneumonias: Advances in Knowledge since 2002. Radiographics. 2015;35(7):1849–1871. doi:10.1148/rg.2015140334



Description of the images
The chest x ray reveals subtle peripheral patchy opacities and elevation of the right hemidiaphragm as consequence of lung volume loss.
The CT images show peripheral bibasilar honeycombing images associated with retraction bronchiectasis and reticular opacities.
Diagnosis: Usual interstitial pneumonia (UIP)

Answer to the Quiz

1. A, C and D

2. A, B and E