1. A 48-year-old woman presents with slowly progressive respiratory insufficiency. Chest x-rays show a diffuse infiltrative process most prominent at the lung base. An open lung biopsy shows extensive fibrosis of the interalveolar septa, dilatation of alveolar ducts, and chronic inflammatory infiltration of the septa. Histopathologic changes are patchy, with areas of advanced fibrosis alternating with areas of active inflammation and new collagen deposition. These findings are most consistent with:

A. Desquamative interstitial pneumonia  
B. Giant cell interstitial pneumonia  
C. Lymphocytic interstitial pneumonia  
D. Nonspecific interstitial pneumonia  
E. Usual interstitial pneumonia

2. A 38-year-old woman with a history of heavy smoking presents with slowly progressive respiratory insufficiency. Chest x-rays show a bilateral ground-glass interstitial infiltrate involving the lung base. An open lung biopsy reveals the following histopathologic changes: alveolar spaces filled with histiocytes, hyperplasia of alveolar type II cells, and lymphomonocytic, plasma cell, and neutrophilic interstitial infiltrate. These changes are relatively homogeneous. The most likely diagnosis is:

A. Centriacinar emphysema with chronic bronchitis  
B. Desquamative interstitial pneumonia  
C. Nonspecific interstitial pneumonia  
D. Respiratory bronchiolitis  
E. Usual interstitial pneumonia

3. Extrinsic allergic alveolitis (also called hypersensitivity pneumonitis) is a reaction to long-term exposure to various antigens of animal or plant origin. The inflammatory infiltrate present in the lung is predominantly composed of:

A. Cytotoxic T lymphocytes and plasma cells  
B. Eosinophils and neutrophils  
C. Helper T lymphocytes and plasma cells
D. Histiocytes and multinucleated giant cells
E. Lymphocytes and histiocytes

4. Pulmonary conditions such as Löfgren syndrome, tropical eosinophilia, allergic bronchopulmonary aspergillosis, and bronchocentric granulomatosis usually result from hypersensitivity reactions to:
   A. Allergens of plant origin
   B. Bacterial organisms
   C. Fungi, parasites, and drugs
   D. Nitrogen dioxide (NO₂)
   E. Viruses

5. An adult patient presents with a chest x-ray picture of bilateral areas of diffuse consolidation. Biopsies of involved areas demonstrate multiple necrotizing granulomas in close proximity to small bronchi. Bronchial spaces are filled with mucus, neutrophils, and eosinophils. No histologic evidence of arteritis is seen. Peripheral eosinophilia is mild, and antineutrophil cytoplasmic antibodies (ANCA) are undetectable. Which of the following is the most likely diagnosis?
   A. Allergic bronchopulmonary aspergillosis (ABPA)
   B. Allergic granulomatosis (Churg-Strauss syndrome)
   C. Bronchiolitis obliterans–organizing pneumonia
   D. Bronchocentric granulomatosis
   E. Wegener granulomatosis limited to the lungs

6. Which of the following histopathologic findings is considered most characteristic of bronchiolitis obliterans–organizing pneumonia?
   A. Hyaline membranes
   B. Interstitial fibrosis
   C. Intrabronchiolar fibroblastic plugs
   D. Macrophages within alveolar spaces
   E. Peribronchial granulomas

7. A lymphoma originating from BALT (bronchial-associated lymphoid tissue) is usually:
   A. An aggressive high-grade lymphoma
   B. Associated with lymphatic tracking
   C. Composed of T lymphocytes, similar to lymphocytic interstitial pneumonia (LIP)
   D. Not associated with lymphoepithelial lesions
   E. Seen in young patients

8. The entity known as lymphomatoid granulomatosis is a/an:
   A. Infectious condition due to mycobacteria
   B. Leukemic infiltration
   C. Malignant lymphoid proliferation
   D. Process akin to mycobacterial pseudotumor
   E. Reactive lymphoid process

9. A 20-year-old man undergoes surgery for resection of a well-circumscribed mass in the right lung. Histologically, it is composed of spindle cells arranged in fascicles variably admixed with a plasma cell population. The spindle cells react with antibodies to vimentin and actin and are negative for CD68. The plasma cell component is polyclonal as evaluated by light chain immunohistochemistry. The diagnosis most compatible with these features is:
   A. Bronchiolitis obliterans–organizing pneumonia
   B. Lymphomatoid granulomatosis
   C. Mycobacterial pseudotumor
   D. Plasma cell granuloma
   E. Pulmonary hyalinizing granuloma

10. A 13-year-old child presents with recurrent hemoptysis and iron deficiency anemia. Granular perihilar infiltrates are present on chest x-rays. Which of the following histologic findings supports a diagnosis of idiopathic hemosiderosis instead of Goodpasture syndrome?
    A. Absence of IgG deposition along basement membranes
    B. Accumulation of hemosiderin-laden macrophages
    C. Hyperplasia of alveolar cells
    D. Presence of interstitial fibrosis
    E. Presence of interstitial lymphocytic infiltration

11. A 35-year-old woman manifests increasingly severe respiratory distress and chest pain. Cardiac catheterization leads to a clinical diagnosis of pulmonary hypertension of unknown origin. In the earliest stages of this condition, an open lung biopsy would reveal predominantly:
    A. Fibrin thrombi
    B. Intimal hyperplasia
    C. Medial thickening
    D. Plexogenic lesions
    E. Reduplication of elastic lamina
12. The condition known as lymphangiomyomatosis is seen exclusively in:
   A. Children
   B. Young men
   C. Women of childbearing age
   D. Postmenopausal women
   E. Elderly persons

13. Which of the following pulmonary conditions is due to the accumulation of surfactant apoprotein within alveoli?
   A. Alveolar proteinosis
   B. Amyloidosis, diffuse alveolar septal type
   C. Goodpasture syndrome
   D. Idiopathic hemosiderosis
   E. Pulmonary alveolar microlithiasis

14. The illustrated lesion is most likely:
   A. Usual interstitial pneumonitis (UIP)
   B. Desquamative interstitial pneumonia (DIP)
   C. Cryptogenic organizing pneumonia (COP)
   D. Diffuse alveolar damage (DAD)
   E. Respiratory bronchiolitis (RB)

15. The illustrated lesion is most likely:
   A. Bronchopneumonia
   B. Diffuse alveolar damage

16. The illustrated lesion is most likely:
   A. Desquamative interstitial pneumonia
   B. Bronchiocentric granulomatosis
   C. IgG4-related disease
   D. Respiratory bronchiolitis
   E. Lymphangioleiomyomatosis

17. The illustrated lesion is most likely:
   A. Giant cell pneumonia
   B. Usual interstitial pneumonia
   C. IgG4-related disease
   D. Nonspecific interstitial pneumonia
   E. Pulmonary Langerhans histiocytosis
18. The illustrated lesion is most likely:
   A. Cryptogenic organizing pneumonia
   B. Extrinsic allergic alveolitis
   C. Nonspecific interstitial pneumonitis
   D. IgG4-related disease
   E. Bronchocentric granulomatosis

20. The illustrated lesion is most likely:
   A. Usual interstitial pneumonia
   B. Cryptogenic organizing pneumonia
   C. Lymphoid interstitial pneumonia
   D. IgG4-related lung disease
   E. Pulmonary Langerhans histiocytosis

19. The illustrated lesion is most likely:
   A. Centrilobular emphysema
   B. Pulmonary Langerhans cell histiocytosis
   C. Lymphangioleiomyomatosis
   D. Metastatic leiomyosarcoma
   E. Honeycomb lung

21. The illustrated lesion is most likely:
   A. Granulomatous pulmonary arteritis (Wegener granulomatosis)
   B. Lymphomatoid granulomatosis
   C. Churg-Strauss arteritis
23. Cryptogenic organizing pneumonia is characterized by all the following features, except:
   A. Elevated erythrocyte sedimentation rate
   B. Restrictive pulmonary function tests
   C. Destruction of pulmonary architecture
   D. Hyperplasia of type II pneumocytes
   E. Uniformity of stage of organization in any given biopsy

24. Diffuse alveolar damage (DAD):
   A. May result in UIP
   B. May be focal
   C. Contains numerous neutrophils in the alveolar spaces
   D. Progresses gradually frequently after an upper respiratory infection
   E. Has markedly reactive type II pneumocytes

25. Tobacco-related lesions of the lung include all the following EXCEPT for:
   A. Respiratory bronchiolitis
   B. Respiratory bronchiolitis with interstitial lung disease
   C. Diffuse alveolar damage
   D. Desquamative interstitial pneumonia
   E. Pulmonary Langerhans cell histiocytosis

26. Pulmonary Langerhans cell histiocytosis (eosinophilic granuloma) is characterized by all EXCEPT for which of the following features?
   A. Bronchiolocentricity
   B. Interstitial location
   C. Composed of histiocytes with groove nuclei and Birbeck granules
   D. Enlarges and cavitates to simulate malignancy
   E. Often lacks eosinophils
1. Correct choice: E

USUAL INTERSTITIAL PNEUMONIA—UIP may be associated with auto-immune disease such as rheumatoid arthritis or scleroderma, but most cases are idiopathic and are often preceded by viral-like symptoms. UIP shows interstitial fibrosis with an uneven distribution of recent and old fibrotic changes, leading to honeycombing in late stage. Choice A is characterized by prominent alveolar macrophage accumulations, evenly distributed, with less fibrosis, Choice B has numerous multinucleated macrophages, Choice C has prominent lymphocytic interstitial infiltrate, and Choice D has an even distribution of temporally similar fibrotic changes.

2. Correct choice: B

DESQUAMATIVE INTERSTITIAL PNEUMONIA—This form of pneumonia has the same predisposing conditions as UIP, but occurs on average one decade before UIP. The histopathologic hallmark is an intra-alveolar accumulation of macrophages, with hyperplasia of type II pneumocytes. DIP may progress to end-stage (honeycomb) lung, but usually responds to steroid treatment, hence the importance of differentiating DIP in lung biopsies from other forms of interstitial lung disease. UIP leads to homogeneous pathologic changes, as opposed to the patchy involvement of UIP. Respiratory bronchiolitis may mimic DIP, affects smokers preferentially, but is associated with a predominantly peribronchiolar pattern of inflammatory changes.

3. Correct choice: A

EXTRINSIC ALLERGIC ALVEOLITIS—This entity is important because progression of lung disease can be halted by preventing further exposure to the inciting agent. Other synonyms for EAA are farmer’s lung, bird fancier’s lung, maple bark stripper’s lung, humidifier lung, and so on, depending on the agent. EAA is a form of hypersensitivity pneumonia leading to inflammatory infiltration of the interstitium, alveoli, and terminal airways. The distribution of inflammatory changes is uneven. Cytotoxic T cells and plasma cells are the predominant inflammatory elements, but scattered histiocytes and multinucleated giant cells are present. Eosinophils and neutrophils are infrequent, whereas granulomas may be found in terminal airways.

4. Correct choice: C

PULMONARY EOSINOPHILIC REACTIONS—This pattern of injury is common to different conditions, all of which result from a hypersensitivity IgE-mediated reaction to fungi, parasites, and occasionally drugs. Aspergillus is the most commonly involved agent, for example, in allergic bronchopulmonary aspergillosis, bronchocentric granulomatosis (BCG), and chronic eosinophilic pneumonia. Wuchereria bancrofti is responsible for tropical eosinophilia. Tissue and blood eosinophilia represent the unifying theme. Pulmonary changes are described as eosinophilic pneumonia, because the intra-alveolar protein-rich fluid, along with polymorphonuclear infiltration and edema, imparts an exudative character to the histologic picture. Chronic eosinophilic pneumonia is associated with bronchiolitis obliterans and few eosinophils. NO₂ causes acute noncardiogenic edema known as silo-filler’s disease.

5. Correct choice: D

BRONCHOCENTRIC GRANULOMATOSIS—This pulmonary condition is akin to other conditions covered under the umbrella of pulmonary eosinophilic reactions. It is characterized by necrotizing granulomas centered on the walls of small bronchi and bronchioles. Histologic changes may bear a striking similarity to tuberculosis. There is no necrotizing vasculitis or ANCA, but peripheral eosinophilia may not be present. As with other forms of pulmonary eosinophilic syndromes, asthma is a frequent manifestation. Wegener granulomatosis limited to the lungs is histologically identical to the more common systemic form.

6. Correct choice: C

ORGANIZING PNEUMONIA OR CRYPTOGENIC ORGANIZING PNEUMONIA—COP is considered a clinicopathologic syndrome resulting from a number of inciting events, including infectious agents, toxic inhalants, drugs, and immune-related collagenopathies. Clinical presentation may be acute (dyspnea, cough, fever) or insidious. Histologically,
intrapulmonary fibrous plugs (Masson plugs), interstitial inflammatory infiltration, and endoluminal macrophages lead to consolidation of pulmonary parenchyma. Masson plugs have a characteristic serpiginous shape. A classification of COP with respect to underlying etiologies is proposed in the table that follows.

The angioinvasive nature of this process leads to large areas of coagulation necrosis. Lymphomatoid granulomatosis eventually evolves to aggressive lymphoma with a poor prognosis. The EBV genome has been found in up to 70% of cases. Lungs, skin, and the central nervous system are the most commonly involved sites.

### Cases in which an underlying cause is apparent
- Extrinsic allergic alveolitis
- Chronic eosinophilic pneumonia
- Organizing bacterial pneumonia

### Cases in which COP constitutes the predominant morphologic pattern
- Mycoplasma pneumonia
- Viral pneumonia
- Toxic fumes (NO₂) inhalation
- Collagen vascular diseases

### Cases in which the underlying condition remains unknown
- Approximately 50%

#### 7. Correct choice: B

**BALT-ASSOCIATED LYMPHOMA** (p. 1135)—This low-grade lymphoma is usually discovered in asymptomatic middle-aged or elderly patients. It is sometimes difficult to differentiate a pulmonary lymphoma from benign lymphoid infiltrates such as LIP or pulmonary pseudolymphoma (PL), and probably, pulmonary lymphoma arises from progressive degrees of BALT hyperplasia. Features favoring BALT lymphoma include sheets or confluent nodules of uniform small lymphocytes, B-cell composition (in contrast to LIP), characteristic lymphoepithelial lesions (similar to MALT lymphoma of the GI tract), and spread along lymphatics and blood vessels. The latter feature is referred to as lymphatic tracking. The prognosis is excellent.

#### 8. Correct choice: C

**LYMPHOMATOID GRANULOMATOSIS**—This condition is a lymphoproliferative disorder characterized by an angiocentric/angiodestructive proliferation of a polymorphic cell population. It can be thought of as a T-cell–rich B-cell lymphoma. Infiltrates consist of mixtures of atypical lymphocytes, histiocytes, plasma cells, and so on, but B lymphocytes are the true neoplastic element. T lymphocytes and other inflammatory cells are reactive.

### 9. Correct choice: D

**PLASMA CELL GRANULOMA**—This lesion consists of variable mixtures of plasma cells (polyclonal, nonneoplastic) and spindly cells with features of myofibroblasts. The latter cell component, which may generate confusion with spindle cell neoplasms, expresses vimentin and actin but not CD68 (a histiocytic marker). PCG is usually observed in young people, most of whom are asymptomatic. The lesion that most closely mimics PCG is *mycobacterial pseudotumor*, which can be diagnosed by CD68 immunoreactivity for histiocytes and, most important, stains for acid-fast bacilli. *Pulmonary hyalinizing granuloma* is composed of collagen bands arranged in a storiform pattern and contains scanty lymphocytes and plasma cells.

#### 10. Correct choice: A

**IDIOPATHIC HEMOSIDEROSIS**—This case highlights the principal clinical and histopathologic findings of idiopathic hemosiderosis, which manifests in children and young adults with hemoptysis and anemia. Histologically, hemosiderin-laden macrophages, alveolar cell hyperplasia, varying degrees of interstitial inflammatory infiltration, and fibrosis are the main findings. Vasculitis and linear IgG deposits along the alveolar basement membrane are absent. The latter negative findings, as well as absence of renal involvement, distinguish idiopathic hemosiderosis from Goodpasture syndrome.

#### 11. Correct choice: C

**PRIMARY PLEXOGENIC HYPERTENSION**—This form of primary pulmonary hypertension affects young women. Although plexogenic lesions are the most characteristic, early stages are associated with hypertrophy of vascular media. Intimal hyperplasia and reduplication of the elastic lamina follow, and arteriolar dilatation, plexogenic lesions, and fibrin thrombi occur in late phases. This sequence of events, however, has been questioned. Morphologic changes of PPTH are identical to those due to secondary
pulmonary hypertension, for example, cases due to cardiac disease with a left-to-right shunt.

12. Correct choice: C

PRIMARY LYMPHANGIOMYOMATOSIS—The defining histopathologic change of this condition is widespread smooth muscle proliferation arising from bronchi, bronchioles, veins, and lymphatics. Grossly, lymphangiomymomatosis may resemble emphysema (early stages) or honeycombing (advanced stages). Cystic spaces are separated by thick septa occupied by fascicles of smooth muscle cells. This condition is usually bilateral and diffuse and affects women of reproductive age. Some of these patients have tuberous sclerosis, which also leads to the development of angiomyolipomas. A feature common to both angiomyolipomas and lymphangiomymomatosis is immunoreactivity for HMB-45. The main differential diagnosis is with benign metastasizing leiomyomas from the uterus.

13. Correct choice: A

ALVEOLAR PROTEINOSIS—This clinicopathologic syndrome may be associated with a variety of conditions (e.g., disseminated severe bacterial, fungal or viral infections, leukemia and lymphoma, radiation or chemotherapy) that usually develop in immunocompromised patients. However, most cases are idiopathic. Radiographically, the disease may mimic pulmonary edema. Histologically, the alveolar cavities are filled with granular, PAS-positive homogeneous material, which is derived from abnormal precipitation of surfactant apoprotein.

14. Correct choice: C

COP—COP or cryptogenic organizing pneumonia bronchiolitis obliterans with organizing pneumonia or BOOP. COP is characterized by fibromyxoid plugs filling the air spaces. Within any given biopsy, these appear to be at approximately the same stage. Subsequently, scarring may result, or they may completely clear. UIP is also characterized by acute and chronic scarring, but the scarring process is in different phases and is also characterized by the presence of fibroblastic foci. DIP is characterized by the presence of clumps of alveolar macrophages, which are generally without scarring of the interstitium, although in later changes, they may be associated with some scarring. Diffuse alveolar damage is characterized by an exudative process in the presence of inflammatory cells, which include lymphocytes as well as some neutrophils. The pathognomonic feature is the presence of hyaline fibrinous membranes surrounding the alveolar spaces and applied to the alveolar surfaces. Respiratory bronchiolitis is characterized by filling of the alveolar spaces with alveolar macrophages containing brown and black pigment and peribronchiolar chronic inflammation. The interstitium may become scarred, at which time the lesion is considered respiratory bronchiolitis with interstitial lung disease.

15. Correct choice: B

DIFFUSE ALVEOLAR DAMAGE—The exudative process with inflammatory cells and the characteristic hyaline membrane identifies the lesion as alveolar damage. The diffuseness of the process has to be confirmed from the biopsy or from its radiographic appearance. As noted above, DAD may clear entirely or it may result in interstitial fibrosis. Choice A: Bronchopneumonia is characterized by an exudative process, but the air spaces are filled with inflammatory cells, and a hyaline membrane is not present. The hyaline membrane of DAD can be thought of as the result of capillary damage with leakage of fibrin, which elicits the intense inflammatory response. Choice C: Desquamative interstitial pneumonitis is characterized by the filling of the alveolar spaces with clumps of alveolar macrophages usually with little to no damage to the interstitium and without the presence of lymphocytes or neutrophils. The macrophages may assume an epithelioid appearance. Choice D: Extrinsic allergic alveolitis is a bronchialocentric lesion, which is characterized by the presence of poorly formed granulomas and alveolar
inflammation, which is sometimes granulomatous but more often chronic inflammatory. Choice E: Bronchiocentric granulomatosis is also bronchiocentric or bronchiolocentric, but it is characterized by extensive well-formed granulomas, which efface the bronchi, but generally spare the adjacent arteries.

16. Correct choice: D

RESPIRATORY BRONCHIOITIS—The figure shows the presence of peribronchiolar inflammation and alveolar macrophages containing brown pigment scattered throughout the air spaces. Interstitial fibrosis may or may not accompany this lesion. Choice A: Desquamative interstitial pneumonitis is characterized by the filling of air spaces with macrophages. Choice B: Bronchocentric granulomatosis is characterized by the presence of extensive granulomas, which are well formed and affect the peribronchiolar or peribronchial area and efface the bronchus, but spare the arteries. Choice C: IgG4-related disease is characterized by compression of the airspaces, which are filled with foamy macrophages. It is associated with scarring and a lymphoplasmacytic infiltrate in the interstitium. Choice E: Lymphangioleiomyomatosis is characterized by bronchiolar cysts surrounded by smooth muscle and containing hemosiderin-laden macrophages. The smooth muscles surrounding these dilated bronchial-based spaces stain positively for HMB-45.

17. Correct choice: E

PULMONARY LANGERHANS HISTIOCYTOSIS—The stellate interstitial lesion is the result of fibrosis associated with the presence of Langerhans cells, which are characterized by large and grooved nuclei. Eosinophils are often present but are not required for the diagnosis of pulmonary Langerhans histiocytosis or eosinophil granuloma. In later stages, there may be a paucity or even absence of the atypical histiocytes, which are replaced by fibrosis. However, a stellate bronchiolocentric interstitial scar is suggestive of the presence of “burned out” pulmonary Langerhans histiocytosis. Choice A: Giant cell pneumonia is characterized by the presence of an intense interstitial pulmonary fibrosis with inflammation and the presence of multinucleated giant cells. Choice B: Usual interstitial pneumonia is characterized by the presence of interstitial fibrosis of varying ages and areas of interstitial inflammation with fibroblastic foci. Choice C: IgG4-related disease is characterized by compressed airspaces filled with foamy macrophages, interstitial fibrosis, and a lymphoplasmacytic infiltration staining for IgG4.

Choice D: Nonspecific interstitial pulmonary fibrosis is similar in many ways to usual interstitial pneumonia, but it lacks the variation in stage of the inflammatory process and fibrosis, and it also lacks the fibroblastic foci.

18. Correct choice: B

EXTRINSIC ALLERGIC ALVEOLITIS—The lesion is bronchiolocentric and is characterized by interstitial inflammation, organizing pneumonia, and the presence of poorly defined granulomas, which usually contain one or a few multinucleated histiocytes. Choice A: Cryptogenic organizing pneumonia is characterized by fibromyxoid plugs in the alveolar spaces, but does not contain granulomas. Choice C: Nonspecific interstitial pneumonitis is characterized by interstitial fibrosis, but without variation in the stage of the fibrosis across the lesion. Interstitial inflammation may be a small or fairly prominent part of the process. IgG4-related disease is characterized by compressed air spaces with foamy macrophages scarring and a lymphoplasmacytic infiltrate. Choice E: Bronchocentric granulomatosis is characterized by well-formed granulomas compressing and effacing the bronchi.

19. Correct choice: C

LYMPHANGIOLEIOMYOMATOSIS—The bronchiocentric cysts have a relatively thick smooth muscle wall, which stains for HMB-45; the intervening lung is relatively normal. Alveolar macrophages containing large amounts of hemosiderin are characteristic of the lesion. Choice A: Centrilobular emphysema is also located in the center of the lobule and bronchiolocentric, but there is fibrosis around the cystic space, and not smooth muscle. Hemosiderin-laden macrophages are not a component of this lesion. Choice B: Pulmonary Langerhans cell histiocytosis is bronchiocentric, but is predominantly interstitial and is characterized by the Langerhans cells, which are S100 positive. Choice D: Metastatic leiomyosarcoma may have a heavy component of smooth muscle, which may be surprisingly well differentiated. The center may be necrotic, but it is not cystic to the extent seen in the lymphangioleiomyomatosis lesion. Choice E: Honeycomb lung is characterized by the presence of diffuse scarring sometimes with a large centrilobular cysts or panacinar emphysema to blebs with destruction of the pulmonary architecture.
Again, fibrosis may be a prominent part, but smooth muscle is rarely as prominent as seen in this lesion and, if present, does not stain with HMB-45.

20. Correct choice: D

IgG4-RELATED LUNG DISEASE—IgG4-related disease is characterized by the presence of an intense lymphoplasmacytic infiltrate, which is associated with fibrosis and compression of the air spaces. Identification of the lesion requires documentation of the presence of IgG4-producing cells by immunohistochemistry. However, the histology is relatively nonspecific in appearance, and the constellation requires clinical pathologic correlation. Choice A: Usual interstitial pneumonia is characterized by temporal difference in the fibrosis as well as the presence of fibroplastic foci. Choice B: Cryptogenic organizing pneumonia is characterized by the fibromyxoid lesions, which are usually confined to the air spaces and rarely involve the interstitium. A lymphoplasmacytic infiltrate is not a characteristic. Choice C: Lymphocytic interstitial pneumonia is characterized by the presence of large amounts of lymphoid cells, which are usually without plasma cell differentiation. They tend to track along the septa of the lung and are not associated with extensive fibrosis. Choice E: Pulmonary Langerhans histiocytosis is associated with interstitial fibrosis and the presence of large mononuclear cells, which are histiocytic in type, S100 positive by immunohistochemistry, and characterized by the presence of nuclear grooves.

21. Correct choice: C

CHURG-STRAUSS ARTERITIS—The lesion is characterized by the presence of an eosinophilic infiltrate with preservation of the architecture of the arteries. Choice A: Granulomatous pulmonary arteritis (WG) is characterized by a lymphoplasmacytic infiltrate of the arterial wall, frequently with compression and obstruction of the wall and association with geographic necrosis in the surrounding lung. Choice B: Lymphomatoid granulomatosis is characterized by the presence of atypical lymphocytes, which are angiocentric and angiodestructive. The characterization of the atypical cells by immunohistochemistry reveals that they are a T-cell–rich B-cell lymphoma. Choice D: Periarteritis nodosa is characterized by a lymphoplasmacytic infiltrate frequently with fibrinoid necrosis of the arteries. Choice E: Tuberculosis may mimic all of the lesions noted above. The arteritis may be intense, and the destruction of lung may be similar to that seen in granulomatous pulmonary arteritis. The diagnostic feature is the presence of the tubercle bacilli.

22. Correct choice: D

VENOOCCLUSIVE DISEASE—The location indicates that the vessels involved are veins. There is fibrosis and scarring of the veins without necrosis. Choice A: Pulmonary hypertension is usually restricted to the arterial component, which is normally found in conjunction with the bronchial or bronchiolar structures. Choice B: Usual interstitial pneumonia is characterized by variation in the temporal stage of interstitial fibrosis and fibroplastic foci. Arterial involvement is usually not intense. Choice C: In granulomatous pulmonary arteritis (WG), the lesions are angiocentric and usually involve arteries. Geographic necrosis is a common accompaniment of this lesion. Choice E: Plexogenic arteriopathy is characterized by the presence of sieve-like vascular channels in muscular arteries and does not usually involve veins.

23. Correct choice: C

DESTRUCTION OF PULMONARY ARCHITECTURE—In cryptogenic organizing pneumonia, the pulmonary architecture is preserved. Healing is thought to frequently resolve and return to a normal pulmonary parenchyma or a pulmonary parenchyma with interstitial fibrosis, but with architectural preservation. Choice A: The lesion is clinically associated with an elevated erythrocyte sedimentation rate suggestive of the inflammatory process, which occurs in the lung. Choice B: Restrictive pulmonary function tests are found despite the presence of obstruction in the alveoli and characteristically in the bronchioles as well. Type II pneumocyte hyperplasia may be present, but is usually not a prominent feature, and there is not atypia of these cells. Choice E: In any given biopsy, there tends to be a uniformity of stage of organization.

24. Correct choice: E

DIFFUSE ALVEOLAR DAMAGE (DAD)—Type II pneumocytes may be so markedly reactive that they may stimulate either a neoplastic process or viral disease with intranuclear viral inclusions. Choice A: Both may result in interstitial fibrosis, but UIP is an ongoing fibrosis, whereas fibrosis following DAD is temporally similar. Choice B: The disease is
characteristically diffuse. Choice C: Neutrophils are usually rare in the alveolar spaces; if numerous, they suggest a secondary bacterial pneumonia. Choice D: The lesion progresses rapidly, although it may be preceded by an acute respiratory infection some weeks prior to its onset. The symptoms rapidly progress to shortness of breath and respiratory insufficiency.

25. Correct choice: C

DIFFUSE ALVEOLAR DAMAGE—Although smoking may contribute to DAD, it is found in both smokers and nonsmokers, and another, often serious, cause, such as shock, drowning, and drugs, is usually evident. Choice A (Respiratory bronchiolitis) and choice B (Respiratory bronchiolitis with interstitial lung disease) are smoker’s lesions with damage characteristically to the respiratory bronchiole, which is often associated with centrilobular emphysema. Choice D: Desquamative interstitial pneumonitis is rarely found in nonsmokers and is considered to be a response to cigarette smoke. Choice E: Pulmonary Langerhans cell histiocytosis is primarily found in smokers.

26. Correct choice: D

PULMONARY LANGERHANS CELL HISTIOCYTOSIS—Although pulmonary Langerhans cell histiocytosis lesions may enlarge, they do not cavitate. However, the spiculated configuration of the interstitial scar on x-ray or CT scan may lead to suspicion of malignancy for which the lesion may be removed. Choice A: The lesions are bronchiolocentric, but choice B extends into the surrounding interstitium. Choice C: Characteristically, the histiocytes have grooved nuclei, are S100 positive, and by electron microscopy contain Birbeck granules. Choice E: Surprisingly, the lesion often referred to as eosinophilic granuloma may contain very few eosinophils. The characteristic cell is the large histiocyte.