Top 3 Differentials in Radiology

A Case Review

Second Edition

William T. O'Brien Sr.

Thieme


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Dedicated in memory of

William E. Shiels II, DO, FAOCR

1954–2015

For decades, Dr. Shiels dedicated his life to caring for children and training residents and fellows in the art of pediatric imaging. His pioneering and innovative contributions will certainly outlive his tenure, which was cut far too short. He will be sorely missed but will never be forgotten.

Contents

Preface

It is a distinct pleasure to present the second edition of *Top 3 Differentials in Radiology: A Case Review*. This book is primarily intended for radiology residents and staff physicians who are preparing for board examinations or looking for a refresher in differentials for common and important imaging gamuts. This second edition has new illustrative cases and updated content throughout, while retaining its high-yield format.

The book is organized into 12 core sections based on specific radiology subspecialties. Each section contains a series of common and important imaging gamuts. On the first page of each case, readers are presented with images from a patient whose diagnosis is as yet unknown, along with a clinical history and detailed image legend. The illustrative cases are meant to highlight a key finding or gamut, which is the basis for the case discussion. The second page identifies the key finding, from which a list of differentials is broken down into the "Top 3" and additional differential diagnoses. The discussion section of each case provides a brief review of important imaging and clinical manifestations for all entities on the list of differentials, and imaging pearls are provided at the end of each case to allow for a quick review of key points. The final diagnosis is provided for each case; however, it is by no means the focus of this review book. In fact, many illustrative cases have a final diagnosis which would not be considered in the "Top 3" for the particular gamut. Instead, the primary aim of the book is to generate and have an understanding of a reasonable list of gamut-based differentials, rather than obtaining the "correct" answer. The final section, titled "Roentgen Classics," contains cases from each of the previous core subspecialty sections that have imaging findings characteristic of a single diagnosis; thus, no differential is presented. A detailed discussion of the final diagnosis follows.

As with the first edition and subspecialty "Top 3" books, it is important to realize that the differentials and discussions are based on the key finding or gamut and not necessarily the illustrative cases that are shown. This is by design, since I felt it would be more high-yield to base the differentials and discussions on the overall gamut/key finding rather than the illustrative case presented. Having an understanding of gamut-based differentials will allow one to subsequently tailor the list of differentials for any case that is shown within the gamut, whereas basing the differentials on the selected images would be limited in terms of future utility.

I sincerely hope that you find the "Top 3" case-based approach enjoyable and useful, and I wish you all the best in your future endeavors.

William T. O'Brien Sr.

Acknowledgments

This book would not have been possible without the contributions of numerous colleagues. First and foremost, I am indebted to the faculty of David Grant USAF Medical Center, the University of California at Davis, Oakland Children's Hospital (where I completed my radiology residency training), and the University of Cincinnati and Cincinnati Children's Hospital Medical Center (where I completed my neuroradiology fellowships). The dedicated staff at these institutions have had a profound impact on my career. Their influence is what inspires me to remain in academics in the hopes of having a similar impact on the next generation of radiologists.

Each of the section editors for this second edition devoted many hours to ensure high quality content in the form of new illustrative cases and updated content throughout, while maintaining the central "Top 3" theme. In many instances, they utilized their own teaching file cases to enhance the contributions of case contributors within their chapter. I am grateful to the chapter editors and contributors for the time and expertise they offered to this project.

Lastly, I would like to thank my family for their continuous love and support, as well as the sacrifices they made during completion of this project. I have been blessed with a wonderful wife, Annie; two sons, Patrick and Liam; and a daughter, Shannon. Annie and I have been together for over two decades, and we could not be more proud of our three incredible children. I am grateful beyond words for the joy that they bring into my life each and every day.

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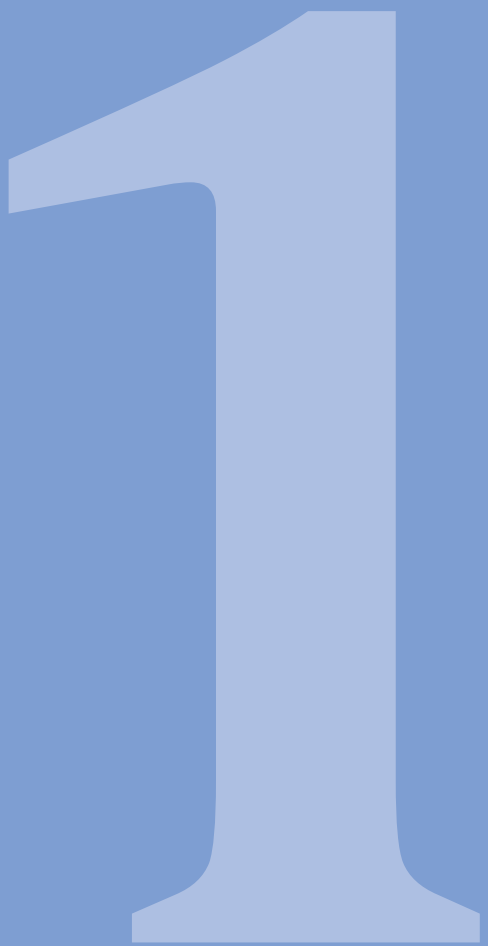
Part 1

Chest and Cardiac Imaging



Case 1

Arash J. Momeni

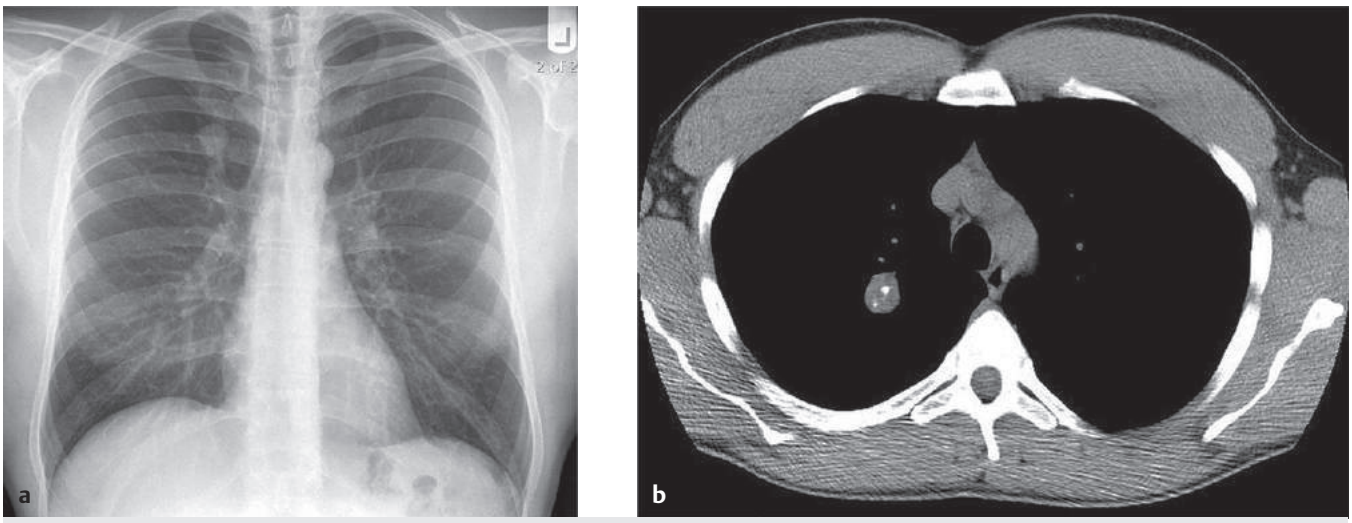


Fig. 1.1 **(a)** Posteroanterior radiograph of the chest demonstrates a solitary pulmonary nodule within the right upper lobe. **(b)** Unenhanced computed tomography (CT) image through the upper chest shows a circumscribed right upper lobe pulmonary nodule with regions of coarse central calcification and small subtle foci of macroscopic fat.

Clinical Presentation

Preoperative evaluation, asymptomatic (▶ Fig. 1.1).

Key Imaging Finding

Solitary pulmonary nodule

Top 3 Differential Diagnoses:

- **Granuloma**. Granulomas are produced secondary to an infectious or inflammatory process, such as tuberculosis, fungal disease, and vasculitides. They present radiographically as solitary or multiple pulmonary nodules. If benign patterns of calcification are identified (central, diffuse, popcorn, or laminated), no further work-up is necessary. Eccentric, speckled, or amorphous calcifications, however, are suspicious for a neoplastic process until proven otherwise. Calcified hilar and mediastinal lymph nodes are commonly seen with granulomatous disease.
- **Neoplasm**. Both primary lung cancer and metastatic disease may present as a solitary pulmonary nodule or mass (>3 cm). Irregular borders or suspicious calcifications (speckled, eccentric) suggest neoplasm over granulomatous disease. Adenocarcinoma characteristically presents as a solid, part-solid, or ground-glass nodule in a peripheral location and is considered the most common primary malignant lung neoplasm. Squamous cell and small cell carcinomas are associated with smoking and tend to occur centrally. Squamous cell carcinoma has a proclivity to cavitate. Small cell carcinoma typically presents as a perihilar mass with associated lymphadenopathy.
- **Hamartoma**. Hamartomas are composed of normal tissue assembled in a disorganized fashion. They are the most common benign tumor of the lung, accounting for 5 to 10% of solitary pulmonary nodules. Classically, they are well-defined, solitary masses less than 4 cm in diameter. Focal macroscopic fat, in addition to a benign pattern of calcification (such as popcorn calcification), is most helpful in making the diagnosis. Evaluation for fat should rely on visible fat, and not on Hounsfield unit (HU), which may falsely be low from averaging with air.

Additional Differential Diagnoses:

- **Round pneumonia**. Typically seen in pediatric patients younger than 8 years, pneumonia may have a rounded mass-like appearance. It is due to centrifugal spread of the rapidly replicating bacteria through the pores of Kohn and canals of Lambert from a single primary focus in the lung.
- **Arteriovenous malformation (AVM)**. AVMs are abnormal communications between a pulmonary or systemic artery and a pulmonary vein. When multiple, nearly 90% are associated with Osler–Weber–Rendu syndrome (hereditary hemorrhagic telangiectasia), characterized by epistaxis, telangiectasia of skin and mucous membranes, and gastrointestinal bleeding. Although usually congenital, AVMs may be acquired in the setting of cirrhosis, trauma, or certain infections. Radiographically, they are well defined. Contrast-enhanced imaging reveals avidly enhancing nodules or masses with an enlarged feeding artery and draining vein. It is critical to prospectively identify AVMs, since inadvertent biopsy can have catastrophic consequences. AVMs are typically treated with embolization (coils or detachable balloons).

Diagnosis

Hamartoma

✓ Pearls

- Benign calcification patterns for a pulmonary nodule include central, diffuse, popcorn, or laminated.
- Malignant features include irregular borders, suspicious calcifications, or eccentric soft-tissue mass.
- Evaluation for fat should rely on visible fat, and not on HU, which may falsely be low from averaging with air.

Suggested Readings

Suut S, Al-Ani Z, Allen C, et al. Pictorial essay of radiological features of benign intrathoracic masses. Ann Thorac Med. 2015; 10(4):231–242

Truong MT, Ko JP, Rossi SE, et al. Update in the evaluation of the solitary pulmonary nodule. Radiographics. 2014; 34(6):1658–1679

Case 2

Philip Yen

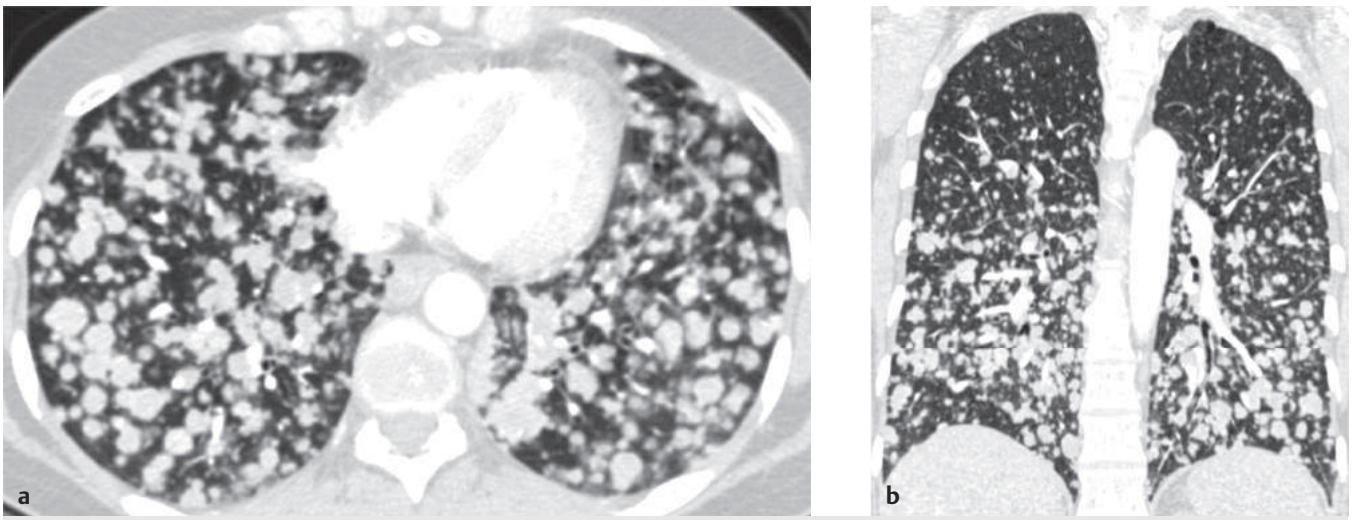


Fig. 2.1 **(a)** Axial contrast-enhanced computed tomography (CT) shows randomly distributed, well-defined pulmonary nodules. **(b)** Coronal CT shows lower lobe predominance.

Clinical Presentation

A 39-year-old woman undergoing treatment for breast cancer (▶Fig. 2.1).

Key Imaging Finding

Multiple pulmonary nodules

Top 3 Differential Diagnoses

- **Metastatic disease**. The majority of patients with multiple metastatic pulmonary nodules have a known primary malignancy. Although nodules can be found anywhere in the lung as they are primarily spread hematogenously, they tend to be randomly distributed with a lower lobe predominance because of the increased blood flow as compared to the upper lobes. Nodules may vary in size, reflecting separate episodes of metastases or varying growth rates. Nodules can be small and numerous or large "cannon-ball" lesions, which are generally seen with sarcomas and gastrointestinal primary malignancies.
- **Tuberculosis or fungal granulomatous disease**. *Mycobacterium tuberculosis* is an aerobic bacterium that disseminates by inhalation of airborne respiratory droplets. Primary and secondary (reactivation) patterns of pulmonary involvement may be seen. Hematogenous spread of the disease results in multiple 1- to 2-mm nodules dispersed in a random distribution. Common fungal causes of multiple granulomatous nodules include histoplasmosis and coccidioidomycosis, both of which may manifest with a miliary pattern accompanied by hilar and/or mediastinal adenopathy.
- **Septic emboli**. Patients with septic emboli usually have a concomitant history of intravenous (IV) drug abuse, bacterial endocarditis, or other source of systemic infection. Patients present with multiple bilateral nodules that tend to be peripheral in location and well defined. A vessel may be identified coursing directly into the center of a nodule, termed the "feeding vessel sign," thought to represent the hematogenous source of the nodule. However, this sign is not specific for septic emboli, as it may also be seen in metastatic disease.

Additional Differential Diagnoses

- **Granulomatosis with polyangiitis (GPA)**. GPA (formerly Wegener granulomatosis) is a vasculitis that affects multiple organs, including the kidneys, upper and lower airways, and lungs. Detection of serological markers such as antineutrophil cytoplasmic antibodies (c-ANCA) frequently aids in diagnosis. Pulmonary manifestations include multiple lung nodules that may range from 2 to 10 cm. These nodules may cavitate and appear thick-walled with air–fluid levels. A history of multiple sinus infections is common.
- **Rheumatoid arthritis (RA)**. Pulmonary involvement of RA occurs after musculoskeletal manifestations. The most common pulmonary manifestation is a pleural effusion. Rheumatoid nodules are not a common presentation of RA, but when they do occur, they can be as small as 2 mm or as large as 5 cm. Nodules may be solitary or multiple and usually are peripheral and well defined. Of particular note is the tendency to disappear with successful therapy as subcutaneous rheumatoid nodules heal.

Diagnosis

Metastatic disease

✓ Pearls

- Because of blood flow, hematogenous spread (whether of infection or neoplasm) tends to favor the lower lobes.
- Tuberculosis and fungal infections can have similar imaging features; TB may be primary or secondary.
- Septic emboli occur in patients with IV drug abuse, endocarditis, or systemic infections and may cavitate.
- The majority of RA patients have musculoskeletal manifestations prior to pulmonary involvement.

Suggested Readings

Collins J, Stern EJ. Chest Radiology: The Essentials. Philadelphia, PA: Lippincott Williams & Wilkins; 2008

Walker CM, Abbott GF, Greene RE, Shepard JA, Vummidi D, Digumarthy SR. Imaging pulmonary infection: classic signs and patterns. AJR Am J Roentgenol. 2014; 202(3):479–492

Webb R, Higgins C. Thoracic Imaging: Pulmonary and Cardiovascular Radiology. Philadelphia, PA: Lippincott Williams & Wilkins; 2010

Case 3

Philip Yen and Matthew L. Lutynski

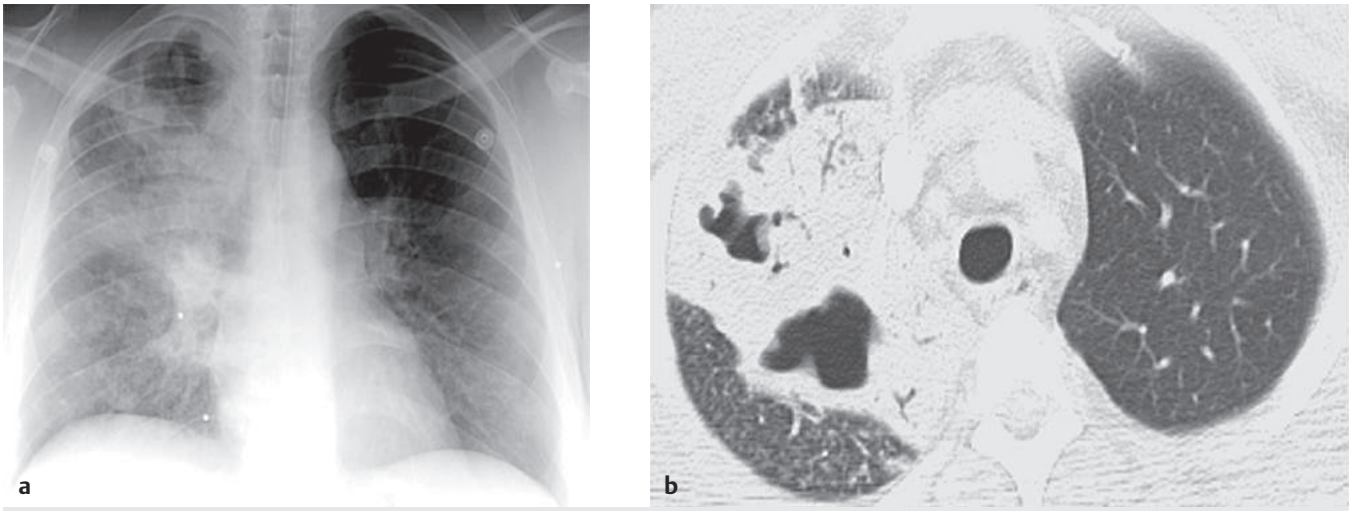


Fig. 3.1 **(a)** Posteroanterior chest radiograph shows a right upper lobe consolidation with a large right apical cavitation. **(b)** Contrast-enhanced axial computed tomography (CT) image shows a thick-walled right upper lobe consolidation with two cavitations.

Clinical Presentation

A 40-year-old man with fevers, night sweats, hemoptysis, and weight loss (▶Fig. 3.1).

Key Imaging Finding

Cavitary pulmonary nodule/mass

Top 3 Differential Diagnoses

- **Tuberculosis**. Primary tuberculosis (TB) may be asymptomatic or present with lobar air-space consolidations. Cavitations are unusual in this phase but can occasionally be found. Reactivation disease presents clinically with night sweats, fever, and weight loss. Radiographically, this phase manifests as multiple cavitations reflecting the increased inflammation and necrosis. These are predominantly in the upper lobes where the higher oxygen tension enables the aerobic bacterium to thrive and where lymphatic clearance is less than that of the lower lobes.
- **Fungal disease**. The most common fungal causes of cavitary lesions are the endemic fungi. Histoplasmosis is endemic to the Ohio and Mississippi River valleys. It particularly favors the nitrogen-rich soil found in bat- or avian guano–laden areas such as caves and chicken houses. Coccidioidomycosis is a soil-borne fungus that is endemic to the Southwestern United States and is spread by inhalation. As with reactivation TB, chronic cavitary fungal colonization preferentially involves the upper lobes.
- **Squamous cell carcinoma (primary or metastatic)**. Approximately 30% of all primary lung cancers arise from primary squamous cell carcinoma (SCC). It is typically located centrally with involvement of hilar or mediastinal lymph nodes. Patients frequently have a history of smoking. Metastatic SCC originates from either a head and neck primary or cervical cancer in females. SCC commonly cavitates.

Additional Differential Diagnoses

- **Pyogenic infection (pulmonary abscess, septic emboli)**. *Staphylococcus aureus* is the most common bacterial infection to result in cavitation. It typically causes a widespread consolidation and may lead to cavitation and abscess formation. In the setting of multiple widespread cavities, a source of showering septic emboli should be considered.
- **Granulomatosis with polyangiitis (GPA)**. GPA (formerly Wegener granulomatosis) is a multiorgan vasculitis that affects the airways, lungs, and kidneys. Patients present with sinus disease, along with cough and hemoptysis. Serum antineutrophil cytoplasmic antibodies (c-ANCA) are frequently detected. The most common pulmonary manifestations are multiple lung nodules, followed by air-space consolidations, ground-glass opacities, and thick-walled cavitations.
- **Rheumatoid arthritis (RA)**. Although not a common manifestation, RA occasionally can present with well-defined pulmonary nodules, which tend to cavitate. The lesions commonly regress with treatment of the underlying disease process. More frequently, thoracic involvement of RA consists of a pleural effusion.

Diagnosis

Reactivation tuberculosis

✓ Pearls

- Cavitation within the upper lobes and superior segment involvement are associated with postprimary TB.
- Histoplasmosis and coccidioidomycosis are the most common fungal pulmonary infections in the United States.
- SCC (primary or secondary) commonly cavitates; primary SCC is associated with smoking.
- GPA is a vasculitis that affects the airways, lungs, and kidneys; pulmonary nodules cavitate.

Suggested Readings

Collins J, Stern EJ. Chest Radiology: The Essentials. Philadelphia, PA: Lippincott Williams & Wilkins; 2008

Walker CM, Abbott GF, Greene RE, Shepard JA, Vummidi D, Digumarthy SR. Imaging pulmonary infection: classic signs and patterns. AJR Am J Roentgenol. 2014; 202(3):479–492

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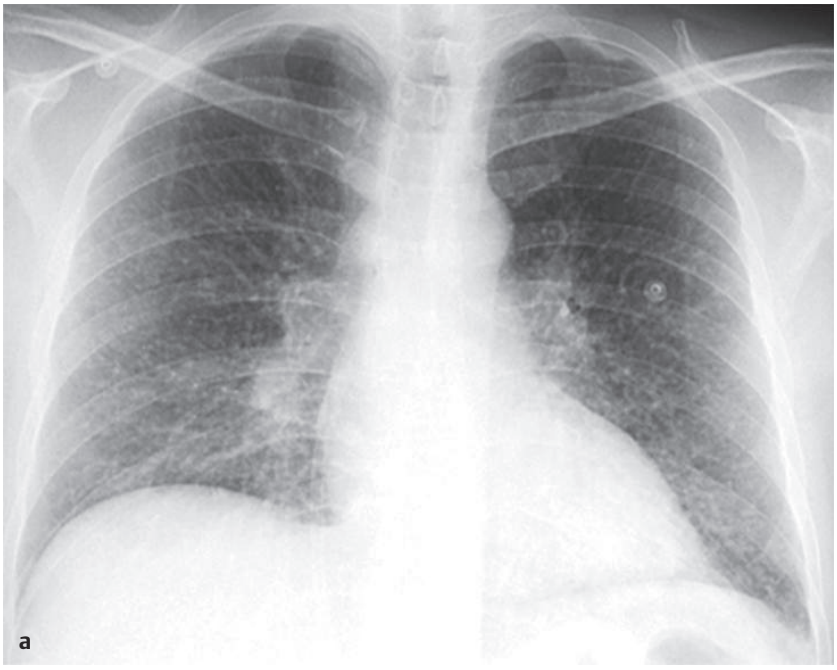
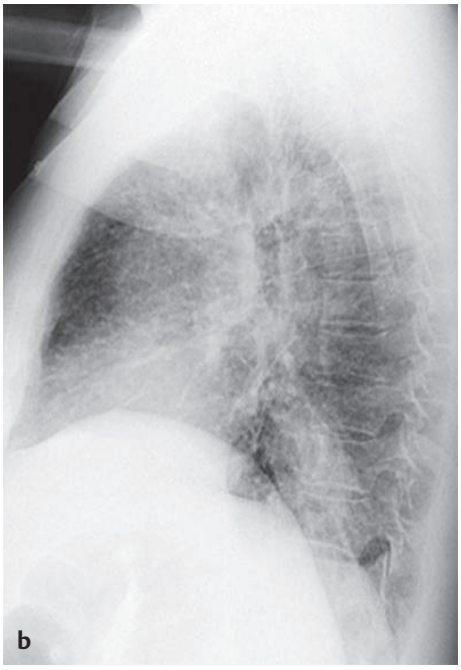
Case 4

John P. Lichtenberger III





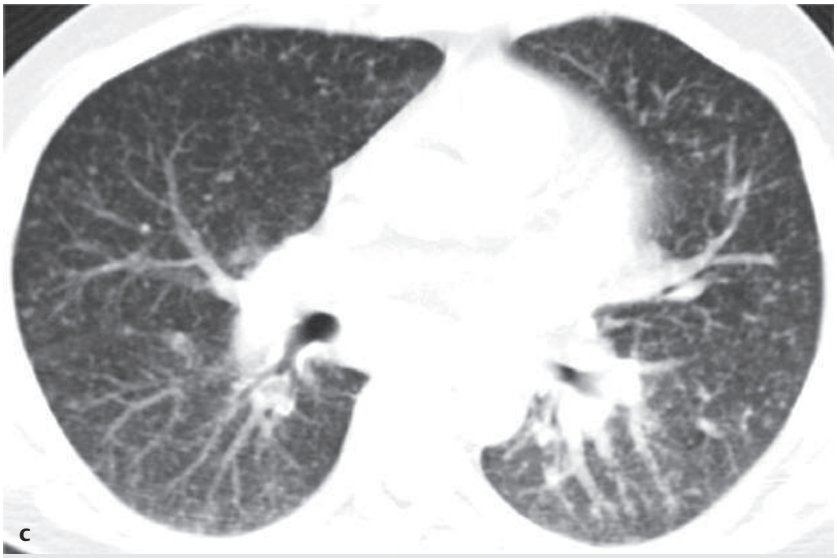


Fig. 4.1 **(a)** Frontal and **(b)** lateral chest radiographs reveal diffuse bilateral tiny nodular densities, along with bilateral hilar fullness. **(c)** Axial computed tomography (CT) image in lung window demonstrates diffuse bilateral 1- to 3-mm miliary nodules in a random distribution.

Clinical Presentation

A 28-year-old immunocompromised patient with chronic cough (▶Fig. 4.1).

Key Imaging Finding

Miliary pulmonary nodules

Top 3 Differential Diagnoses

- **Tuberculosis (TB)**. Pulmonary infection with *Mycobacterium tuberculosis* is classified as primary or reactivation based on clinical and radiographic features. Predominantly, a lower lobe air-space disease, mediastinal lymphadenopathy, and pleural effusions are typical radiographic manifestations of primary infection. Reactivation TB, however, characteristically presents as upper lobe consolidation with regions of cavitation and fibrosis, endobronchial spread with ill-defined centrilobular opacities in a "tree-in-bud" configuration, or hematogenous spread with randomly distributed 1- to 3-mm nodules, referred to as a miliary pattern. The miliary pattern is more common in children, elderly, and immunocompromised patients (HIV, transplant, etc.).
- **Fungal disease**. Fungal infection in the lung encompasses both primary infection in immunocompetent patients (e.g. *Histoplasma*, *Coccidioides*,blastomycosis) and opportunistic infection in immunosuppressed patients (e.g., *Aspergillus*, *Candida*, *Cryptococcus*). Although radiographic features are somewhat organism dependent, disseminated fungal infection can result in a miliary pattern identical to miliary TB. Chronic fungal infections may cavitate.
- **Metastases**. Although hematogenous metastases to the lung may occur with numerous primary malignancies, thyroid carcinoma, renal cell carcinoma (RCC), and melanoma are the most common primary malignancies to produce a miliary pattern of dissemination in the chest.

Additional Differential Diagnoses

- **Pneumoconioses**. The pneumoconioses result from inhalation of particulate matter as a result of occupational exposure. Silicosis and coal worker's pneumoconiosis are among the most common entities. Radiographic findings consist of multiple upper lobe fibrotic nodules ranging from 1 to 10 mm in size. When small, the appearance mimics that of miliary disease processes. As the disease progresses, fibrosis ensues and the nodular densities coalesce. Eggshell calcifications may be seen within hilar and mediastinal lymph nodes. A late complication referred to as progressive massive fibrosis presents radiographically as upper lobe masslike opacities in the setting of underlying fibrosis. Patients are at an increased risk of superinfection, especially TB.
- **Healed varicella**. Acute varicella pneumonia is a severe form of primary infection which occurs primarily in children and pregnant patients. The infection presents as multifocal regions of patchy air-space disease, and affected patients are very ill. Healed varicella presents radiographically as calcified miliary pulmonary nodules in a random distribution.

Diagnosis

Fungal disease (coccidioidomycosis)

✓ Pearls

- Miliary spread of TB is most common in children, elderly, and immunocompromised patients.
- Disseminated fungal disease may present with a miliary pattern; chronic fungal disease can cavitate.
- Thyroid carcinoma, RCC, and melanoma are the most common primary neoplasms with a miliary pattern.
- Pneumoconioses result from occupational exposure; eggshell lymph node calcifications may be seen.

Suggested Readings

Collins J, Stern EJ. Chest Radiology: The Essentials. Philadelphia, PA: Lippincott Williams & Wilkins; 2008

Mcloud TC, Boiselle PM. Thoracic Radiology: The Requisites. Philadelphia, PA: Mosby; 2010

Walker CM, Abbott GF, Greene RE, Shepard JA, Vummidi D, Digumarthy SR. Imaging pulmonary infection: classic signs and patterns. AJR Am J Roentgenol. 2014; 202(3):479–492

Case 5

Paul B. DiDomenico

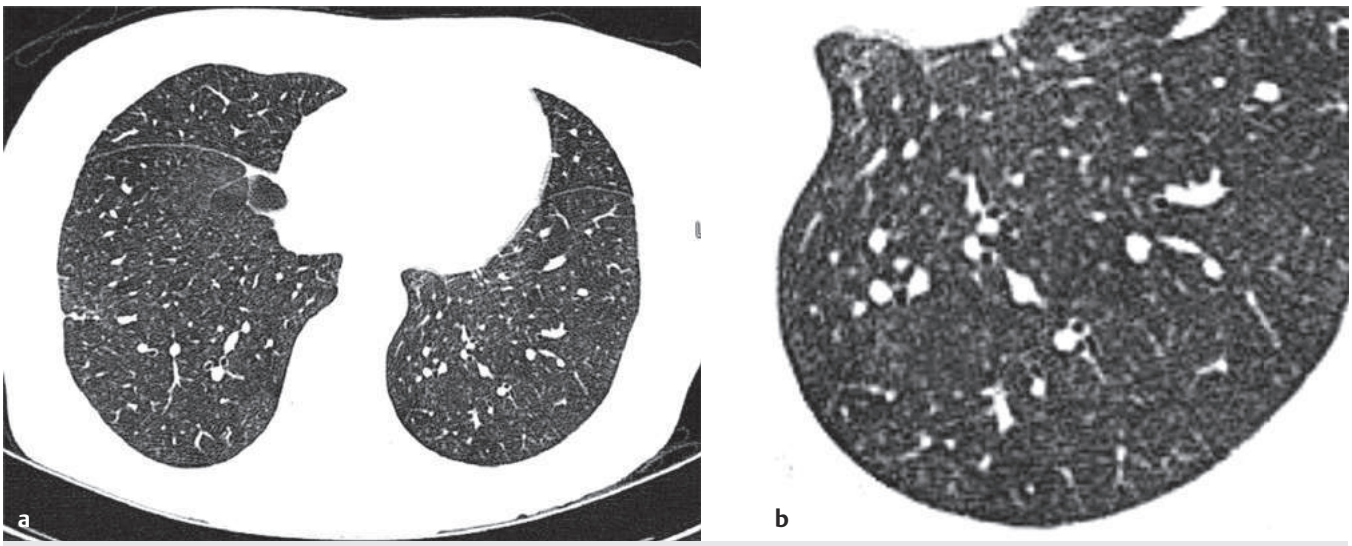


Fig. 5.1 **(a)** Axial CT image through the chest and **(b)** coned-down view of the left lower lobe in lung window reveals diffuse bilateral ill-defined centrilobular ground-glass nodules.

Clinical Presentation

A 50-year-old woman with cough and shortness of breath (▶ Fig. 5.1).

Key Imaging Finding

Centrilobular pulmonary nodules

Top 3 Differential Diagnoses

- **Infectious bronchiolitis (*Mycobacterium avium-intracellulare* infection [MAI], tuberculosis [TB])**. Infection from both tuberculous and nontuberculous mycobacterial (NTMB) organisms may manifest as numerous small nodular opacities centered on the bronchiole of the secondary pulmonary lobule with sparing of the subpleural space (centrilobular nodules). Numerous species of mycobacteria are ubiquitous in the environment, but *M. avium intracellulare* and *Mycobacterium Kansasii* account for most nontuberculous infections. A pattern of clustered nodules with branching opacities (so-called tree-in-bud pattern) in the lingula and right middle lobe is typical of MAI and is often seen in elderly women; this pattern is termed "Lady Windermere" syndrome. NTMB infections may be indistinguishable from postprimary (reactivation) TB.
- **Hypersensitivity pneumonitis (HP)**. Also known as extrinsic allergic alveolitis, HP results from exposure to environmental antigens inhaled as dust particles. Varying sources of organic dusts may result in farmer's lung (moldy hay), bird fancier's lung (avian proteins), or humidifier lung (thermophilic bacteria), which are indistinguishable radiographically. Following exposure, the acute phase may exhibit fine nodular or ground-glass opacities. Centrilobular nodules with ground-glass opacities are typical of the subacute phase of the disease, most often in the mid- to lower lung zones. If exposure continues, there may be progression to end-stage lung disease with fibrosis.
- **Endobronchial spread of tumor**. Metastatic spread of tumors within the chest may take a variety of routes including direct, hematogenous, lymphatic, and endobronchial spread. While hematogenous spread is the most common form of widespread dissemination, endobronchial spread is also possible in late stages of disease and may manifest as centrilobular nodules in any part of the lung.

Additional Differential Diagnoses

- **Respiratory bronchiolitis-associated interstitial lung disease (RB-ILD)**. RB-ILD is usually seen in smokers 30 to 50 years of age. Findings at computed tomography (CT) include either upper lobe predominance or diffuse centrilobular nodules, representing accumulated pigmented macrophages, with patchy ground-glass opacities.
- **Pneumoconioses**. Silicosis and coal worker's pneumoconiosis result from occupational exposure and present as upper lobe–predominant interstitial lung diseases. Either perilymphatic or centrilobular nodules may be present, which may eventually coalesce to form masslike opacities and potentially calcify. Involved lymph nodes may also calcify peripherally, resulting in a characteristic "eggshell" appearance. Late findings may include masslike fibrosis with peripheral emphysema (progressive massive fibrosis).

Diagnosis

Hypersensitivity pneumonitis

✓ Pearls

- Mycobacteria are common causes of infectious bronchiolitis; a "tree-in-bud" appearance is characteristic.
- HP results from exposure to inhaled antigens; centrilobular nodules and ground-glass opacities are typical.
- Endobronchial spread of tumor occurs in late stages of malignancy and may involve any part of the lung.
- RB-ILD is a smoking-related process with diffuse or upper lobe–predominant centrilobular nodules.

Suggested Readings

Collins J. CT signs and patterns of lung disease. Radiol Clin North Am. 2001; 39(6):1115–1135

Walker CM, Abbott GF, Greene RE, Shepard JA, Vummidi D, Digumarthy SR. Imaging pulmonary infection: classic signs and patterns. AJR Am J Roentgenol. 2014; 202(3):479–492

Webb WR, Müller NL, Naidich DP. High-Resolution CT of the Lung. Philadelphia, PA: Lippincott Williams & Wilkins; 2009

Case 6

John P. Lichtenberger III

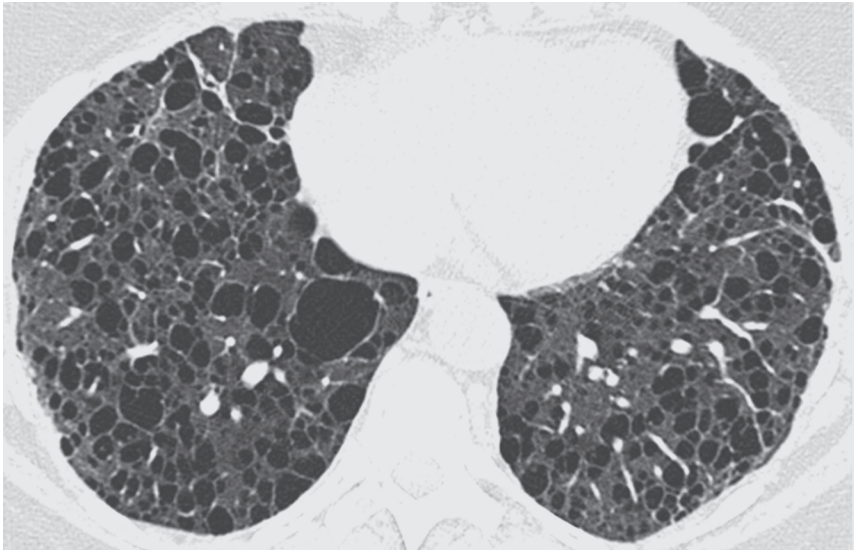


Fig. 6.1 Unenhanced axial CT image of the chest shows numerous bilateral thin-walled cysts, most of which are similar in size. The intervening lung is normal.

Clinical Presentation

A 31-year-old woman with a smoking history, chronic cough, and shortness of breath (▶Fig. 6.1).

Key Imaging Finding

Cystic lung disease

Top 3 Differential Diagnoses

- **Emphysema**. Emphysema is characterized by nonfibrotic enlargement of the airways distal to the terminal bronchioles with destruction of alveolar walls. Computed tomography (CT) further characterizes this process based on the location of these cystic spaces at the lobular level. Centrilobular emphysema involves the central portion of the secondary pulmonary lobule with upper lobe predominance and is highly associated with cigarette smoking. Panlobular emphysema involves the entire lobule and is classically associated with alpha-1 antitrypsin deficiency. Paraseptal emphysema is predominantly subpleural, involving the alveolar ducts and sacs. Bronchiectasis refers to enlargement and thickening of portions of the airways.
- **Lymphangioleiomyomatosis (LAM)**. A relatively rare disease affecting women of reproductive age, LAM is characterized by proliferation of smooth muscle cells around bronchioles. This results in air trapping and characteristic thin-walled lung cysts. The cysts are typically uniform in size. Air trapping predisposes patients to pneumothoraces. A similar process involving the lymphatics results in chylous pleural effusions. LAM may occur as an isolated abnormality or in association with tuberous sclerosis.
- **Pulmonary Langerhans cell histiocytosis (LCH)**. Seen predominantly in young and middle-aged adults and almost exclusively in smokers, pulmonary LCH is an idiopathic disease of mature histiocyte proliferation. On imaging, numerous small (<1.0 cm) upper lobe predominant nodules, many of which demonstrate cavitation, are eventually replaced by irregular thin walled cysts of varying sizes. Costophrenic sulci are typically spared. Associated pneumothoraces are common. Occasionally, LCH may progress to interstitial fibrosis and honeycombing.

Additional Differential Diagnoses

- **Pneumocystis pneumonia**. The most common cause of diffuse pneumonia in immunocompromised patients, *Pneumocystis* pneumonia classically results in central ground glass opacities with or without reticulonodular interstitial thickening on plain radiographs. CT most commonly demonstrates ground glass opacification with abnormal air spaces, to include thin walled cysts, pneumatoceles, and pneumothoraces, as well as interlobular septal thickening.
- **Lymphocytic interstitial pneumonitis (LIP)**. Lymphocytic infiltration of the alveolar septa characterizes lymphocytic interstitial pneumonitis, a lower lobe–predominant hyperplasia of bronchus-associated lymphoid tissue. A reticulonodular interstitial pattern on plain radiographs is better characterized on CT as centrilobular nodules with thin-walled cystic air spaces and regions of ground-glass opacification. LIP is commonly seen in the setting of Sjögren's syndrome and AIDS, particularly in children.

Diagnosis

Lymphangioleiomyomatosis

✓ Pearls

- Cysts and emphysema can coexist but are distinct processes; centrilobular emphysema is most common.
- Cysts must be distinguished from bronchiectasis by establishing lack of continuity with the airways.
- LAM occurs almost exclusively in women of child-bearing age; chylous pleural effusions may be seen.
- LCH is seen almost exclusively in smokers and tends to spare the costophrenic sulci.

Suggested Readings

Collins J, Stern EJ. Chest Radiology: The Essentials. Philadelphia, PA: Lippincott Williams & Wilkins; 2008

Koyama M, Johkoh T, Honda O, et al. Chronic cystic lung disease: diagnostic accuracy of high-resolution CT in 92 patients. AJR Am J Roentgenol. 2003; 180(3):827–835

Case 7

William T. O'Brien, Sr.

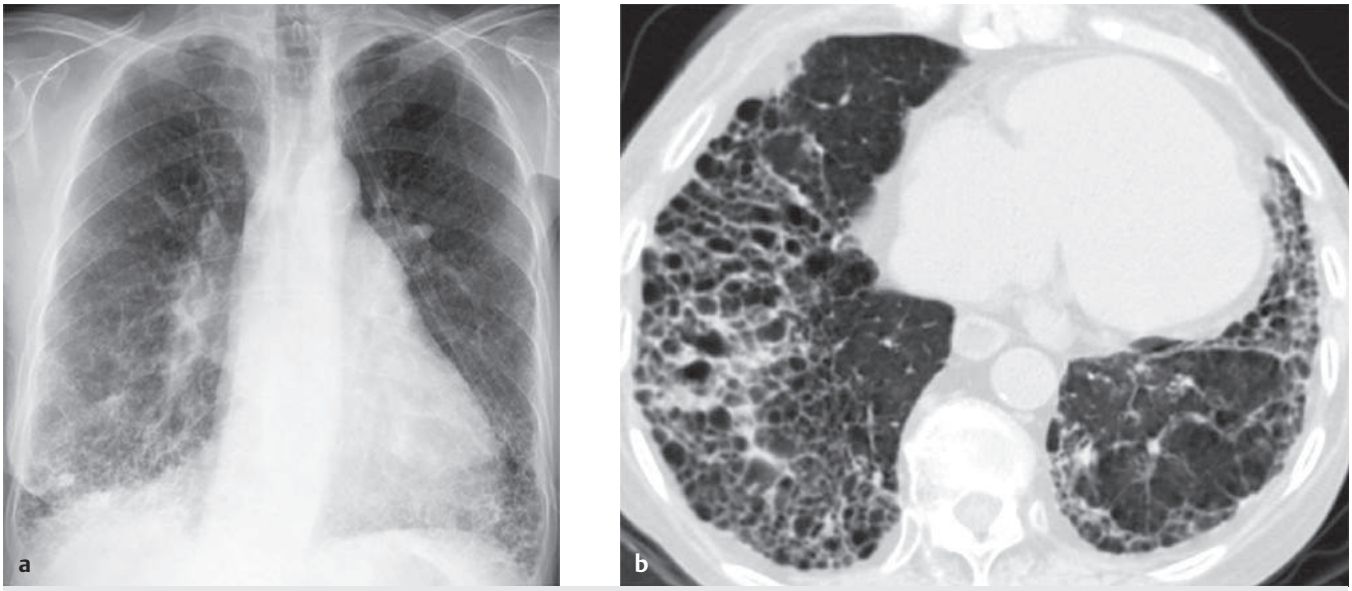


Fig. 7.1 **(a)** Frontal chest radiograph demonstrates coarsened interstitial lung markings predominantly at the lung bases. **(b)** Axial CT image shows traction bronchiectasis and fibrotic changes predominantly in the lower lobes and a dilated, patulous, fluid-filled esophagus.

Clinical Presentation

A 50-year-old woman with chronic shortness of breath and difficulty swallowing (▶Fig. 7.1).

Key Imaging Finding

Lower lobe interstitial lung disease (ILD)

Top 3 Differential Diagnoses

- **Usual interstitial pneumonia (UIP)**. UIP is a relatively common cause of chronic fibrosing lung disease. Patients often present with worsening progressive cough and shortness of breath. Plain radiographs reveal increased interstitial markings at the lung bases with normal-to-low lung volumes. On high-resolution computed tomography (HRCT), UIP presents as irregular septal thickening along the peripheral aspect of the lower lobes with subpleural honeycombing and traction bronchiectasis in advanced cases. Occasionally, ground-glass opacities may been seen, which correlate with regions of active alveolitis. Once diagnosed, median survival is approximately 3 years. About 70% of UIP have no identifiable cause are termed idiopathic pulmonary fibrosis (IPF).
- **Collagen vascular diseases**. Collagen vascular diseases include **scleroderma**, **rheumatoid arthritis** (RA), and **systemic lupus erythematosus** (SLE) and involve the lungs to varying degrees. Scleroderma is a systemic process that affects young females and involves many organ systems—the lungs, esophagus, and musculoskeletal system. Pulmonary findings include pneumonitis and ILD with striking lower lobe predominance. HRCT reveals irregular septal thickening and subpleural honeycombing. Secondary findings include a dilated, patulous esophagus. RA is a systemic disease that most severely affects articular surfaces. Pulmonary involvement may be seen in up to 50% of cases. Unilateral pleural effusion is the most common presentation of RA within the chest. Additional manifestations include lower lobe ILD, necrobiotic pulmonary nodules, and pericarditis. Caplan syndrome refers to coal worker's pneumoconiosis superimposed upon RA. SLE is a systemic process that occurs in young females. Pleural and pericardial effusions are the most common manifestation of SLE in the chest.
- **Asbestos-related disease**. Asbestos-related lung disease results from occupational exposure, as can be seen in former shipyard workers and mechanics. The clinical and radiographic manifestations usually present 20 years after the initial exposure. Pleural thickening and pleural plaques are the most common radiographic manifestation and are typically bilateral, involving predominantly the lower thorax. The plaques often calcify. A benign exudative pleural effusion may also occur. Rounded atelectasis is a common finding and presents as a rounded mass, which abuts the pleura in a region of underlying pleural thickening. Vessels can be seen "swirling" into the lesion. HRCT findings include subpleural lines and parenchymal bands extending to the pleura. Lung involvement can include lower lobe septal thickening and subpleural honeycombing. When there is pulmonary fibrosis, it is called asbestosis.

Additional Differential Diagnoses

- **Drug toxicity**. Drug toxicity from chemotherapeutic agents or illicit drug abuse may rarely result in predominantly lower lobe ILD characterized by irregular septal thickening and subpleural honeycombing. Other patterns of injury include pulmonary edema, pulmonary hemorrhage, and hypersensitivity pneumonitis. Bleomycin and busulfan are the most common chemotherapeutic agents that cause lower lobe ILD. Amiodarone may also result in lower lobe ILD, along with characteristic dense opacities.

Diagnosis

Connective tissue disorder (scleroderma)

✓ Pearls

- UIP is the histologic and imaging descriptor of many diseases; when the cause is unknown, it is called IPF.
- Scleroderma is the most common collagen vascular disease that produces lower lobe fibrosis.
- Asbestos exposure and asbestosis are related but different; asbestosis is defined by interstitial fibrosis.
- Chemotherapeutic agents may predominantly cause lower lobe ILD; amiodarone produces dense opacities.

Suggested Readings

Mueller-Mang C, Grosse C, Schmid K, Stiebellehner L, Bankier AA. What every radiologist should know about idiopathic interstitial pneumonias. Radiographics. 2007; 27(3):595–615

Case 8

Bang Huynh

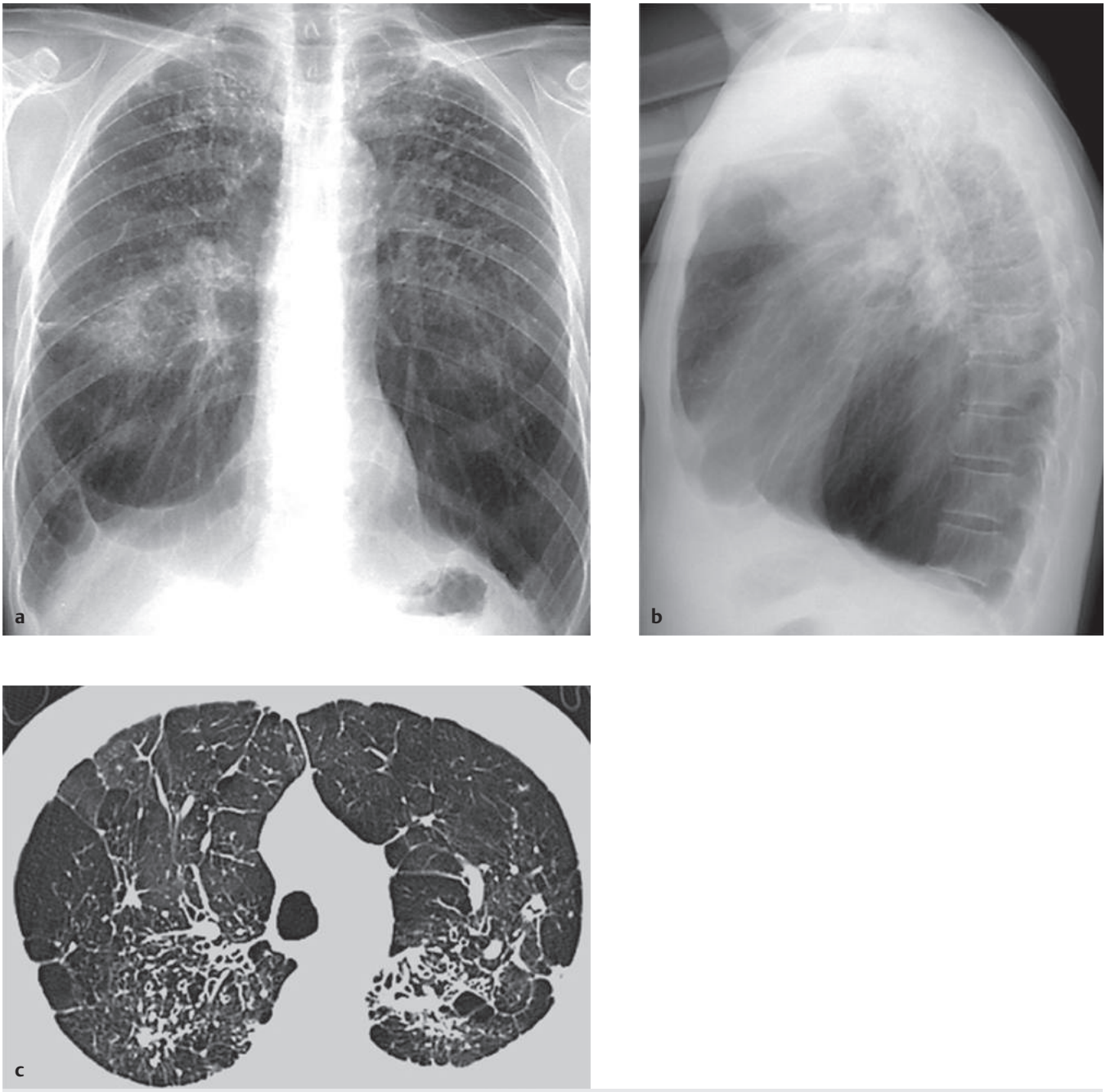


Fig. 8.1 **(a)** Frontal and **(b)** lateral chest radiographs show hyperinflated lungs with narrowing of the mediastinum and flattening of the diaphragm, better seen on the lateral view, as well as superior retraction of the hila. There are coarsened, irregular interstitial markings and countless small nodules predominantly within the upper lobes. A masslike opacity is seen in the right upper lobe extending from the hilum. **(c)** Axial CT image in lung window reveals multiple 2- to 3-mm nodules, interlobular septal thickening with traction bronchiectasis, and scarring with distortion of the normal architecture within the upper lobes.

Clinical Presentation

Chronic progressive shortness of breath (▶ Fig. 8.1).

Key Imaging Finding

Upper lobe interstitial lung disease

Top 3 Differential Diagnoses

- **Postprimary tuberculosis (TB)**. Primary TB can have a broad range of presentations but is typically characterized by mediastinal and unilateral hilar lymphadenopathy (sometimes of low attenuation). Parenchymal involvement can range from poorly defined opacities to lobar consolidations. Pleural effusions are not uncommon. Nodal calcification is present in about a third of cases. There is no distribution preference, and cavitation, though possible, is not common. In contrast, postprimary TB favors the posterior segments of the upper lobes or superior segments of the lower lobes primarily because of higher oxygen tension and less efficient lymphatic clearance. Cavitation and fibrosis are distinct features of postprimary TB. Other manifestations of TB include miliary and endobronchial patterns.
- **Sarcoidosis**. Sarcoidosis is a systemic disease of unknown etiology characterized by noncaseating granulomas. Thoracic disease may include mediastinal/hilar and parenchymal involvement. Hilar adenopathy is often bilateral and symmetric. Parenchymal involvement usually consists of countless 2- to 4-mm centrilobular and subpleural (perilymphatic) nodules. There is an upper lobe and peribronchovascular distribution. Sarcoid granulomas can resolve completely; however, they occasionally cause severe and extensive fibrosis that resembles progressive massive fibrosis that is seen in complicated silicosis.
- **Cystic fibrosis (CF)**. CF is a genetic disorder affecting Caucasian children and young adults. The lungs and gastrointestinal tract are most affected with thick secretions from exocrine glands. Pulmonary involvement occurs in more than 90% of patients. Viscous secretions result in upper lobe–predominant mucus plugging, air trapping, bronchial wall thickening, and bronchiectasis.

Additional Differential Diagnoses

- **Silicosis/coal worker's (CW) pneumoconiosis**. Pneumoconiosis is a broad term that describes reactions of the lungs to inhaled dust particles. CW pneumoconiosis and silicosis are pathologically different but have very similar radiographic appearances. Silicosis is caused by the inhalation of free silica, a compound associated with mining of heavy metal, sandblasting, and stonework. Patients may present acutely following heavy exposure whereby silicoproteinosis develops or, more commonly, with chronic silicosis from decades of exposure. The chronic form results from cyclic inflammation, resulting in fibrosis with upper lobe predominance. Complicated silicosis is an extreme progression where the fibrosis coalesces into masslike opacities, a process called progressive massive fibrosis.
- **Langerhans' cell histiocytosis (LCH)**. Pulmonary LCH is characterized by abnormal histiocyte proliferation and is seen predominantly in young and middle-aged smokers. Imaging findings include numerous small upper lobe–predominant nodules, many of which cavitate, and irregular cysts of varying sizes. Costophrenic sulci are typically spared. Associated pneumothoraces are common.

Diagnosis

Complicated silicosis

✓ Pearls

- Postprimary TB may result in upper lobe cavitary pulmonary nodules and fibrosis.
- Sarcoidosis presents with symmetric hilar adenopathy; perilymphatic pulmonary nodules may also be seen.
- Pulmonary manifestations of CF include upper lobe mucous plugging, air trapping, and bronchiectasis.
- Silicoproteinosis is a unique form of silicosis that is acute and indistinguishable from alveolar proteinosis.

Suggested Readings

Mueller-Mang C, Grosse C, Schmid K, Stiebellehner L, Bankier AA. What every radiologist should know about idiopathic interstitial pneumonias. Radiographics. 2007; 27(3):595–615

Pipavath S, Godwin JD. Imaging of interstitial lung disease. Clin Chest Med. 2004; 25(3):455–465

Pipavath S, Godwin JD. Imaging of interstitial lung disease. Radiol Clin North Am. 2005; 43:589–599

Case 9

Arash J. Momeni

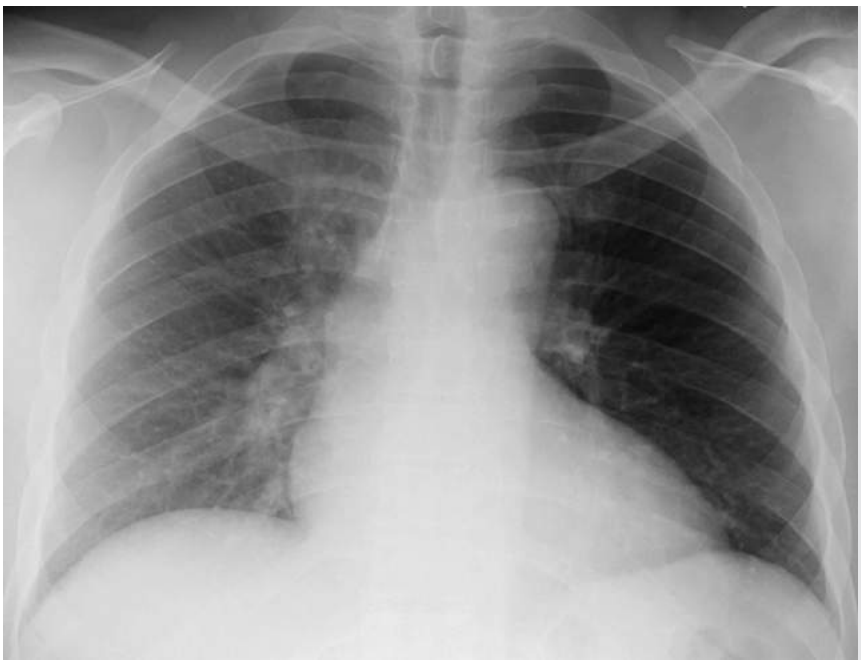


Fig. 9.1 Frontal chest radiograph demonstrates hyperlucency of the left hemithorax. Lung volumes are normal and symmetric.

Clinical Presentation

A 40-year-old man with occasional dyspnea on exertion (▶ Fig. 9.1).

Key Imaging Finding

Unilateral hyperlucent lung

Top 3 Differential Diagnoses That You Cannot Afford to Miss

- **Airway obstruction**. The hallmark of airway obstruction in children is air trapping, which is manifested as failure of the lung to decrease in volume, and subsequently increase in opacification, on expiratory chest radiographs. The mediastinum may shift to the contralateral side. Air trapping occurs when an endobronchial lesion causes a check-valve type of obstruction. In children, this is usually secondary to foreign body aspiration (which may be radiolucent). Expiratory views, decubitus views, and fluoroscopy may be helpful.
- **Pulmonary embolism**. Oligemia of the lung beyond the occluded pulmonary vessel, or the Westermark sign, is a helpful but not commonly encountered sign of pulmonary embolism. A large unilateral thrombus, whether bland, septic, or neoplastic, can result in a unilateral hyperlucent lung.
- **Pneumothorax**. A pneumothorax results in hyperlucency of the ipsilateral hemithorax and typically presents with central displacement of the visceral pleural line, absence of lung markings distal to the displaced pleural line, and possible contralateral shift of the mediastinum (tension pneumothorax). In a supine patient, a pneumothorax may collect anteromedially, in the nondependent portion of the thorax. This will result in the pneumothorax presenting as lucency in the anteromedial chest rather than as a pleural line. A deep hyperlucent sulcus may also be seen. A tension pneumothorax is a true emergency.

Additional Differential Diagnoses

- **Chest wall abnormality**. Several chest wall abnormalities may result in a hyperlucent lung. For example, mastectomy results in relative radiolucency on the side of breast tissue removal. Poland syndrome is characterized by a spectrum of abnormalities ranging from isolated absence of the pectoralis major muscle to additional abnormalities of the ipsilateral extremity, such as syndactyly, brachydactyly, and rib anomalies.
- **Swyer–James syndrome**. Swyer–James syndrome, or MacLeod syndrome, is a form of obliterative bronchiolitis that occurs following an insult, classically viral, to the developing lung. This process affects small bronchi and bronchioles. The portions of the lung ventilated by abnormal airways remain inflated by collateral air drift. Chest radiographs demonstrate unilateral hyperlucency because of reduced lung perfusion. Air trapping may be seen on expiratory radiographs or high-resolution computed tomography (HRCT). Lung volumes on the affected side may be normal or decreased. On CT, additional findings may include bronchiectasis and attenuated vessels in areas of decreased lung attenuation.
- **Acute asthmatic attack**. Hyperlucency in asthma is secondary to bronchoconstriction and compensatory vasoconstriction of hypoventilated portions of the lung. As this is usually a more central process, findings are commonly bilateral but may be asymmetric.

Diagnosis

Chest wall abnormality (Poland syndrome)

✓ Pearls

- Expiratory views, decubitus views, and fluoroscopy may be helpful in evaluating for foreign bodies.
- Always consider pulmonary embolism and pneumothorax (in a supine patient) with a hyperlucent lung.
- Swyer–James syndrome is usually a unilateral process where the affected lung is reduced in size.

Suggested Readings

Collins J, Stern EJ. Chest Radiology: The Essentials. Philadelphia, PA: Lippincott Williams & Wilkins; 2008

Reid L, Simon G. Unilateral lung transradiancy. Thorax. 1962; 17:230–239