CHEST

Lung Hyperlucency

A Clinical-Radiologic Algorithmic Approach to Diagnosis

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Areas of diminished lung density are frequently identified both on routine chest radiographs and chest CT examinations. Colloquially referred to as hyperlucent foci of lung, a broad range of underlying pathophysiologic mechanisms and differential diagnoses account for these changes. Despite this, the spectrum of etiologies can be categorized into underlying parenchymal, airway, and vascular-related entities. The purpose of this review is to provide a practical diagnostic algorithmic approach to pulmonary hyperlucencies incorporating clinical history and characteristic imaging patterns to narrow the differential. CHEST 2020; 157(1):119-141

KEY WORDS: airway; lung hyperlucency; parenchymal; pulmonary hyperlucency; vascular

Areas of abnormal decreased lung density, both focal and diffuse, are frequently identified on chest radiographs (CXRs) and corresponding thoracic CT examinations. Referred to colloquially as hyperlucent lung, the finding of markedly diminished areas of lung density poses a diagnostic challenge because there are multiple and diverse potential explanations for the finding. To date, no systematic approach to the differential diagnosis has been proposed.[1-7] An approach that incorporates clinical history and physical examination findings with specific chest imaging features, with supportive evidence provided by ancillary studies (pulmonary function testing, ventilation perfusion scans [V/Qs]), and echocardiography), can provide a diagnostic framework. The purpose of this report is to provide a systematic approach (Fig 1) to the evaluation of a finding of abnormally decreased lung density. Emphasis is placed on categorizing the most common

ABBREVIATIONS: AATD = alpha-1-antitrypsin deficiency; BPS = bronchopulmonary sequestration; CB = constrictive bronchiolitis; CLO = congenital lobar overinflation; CPAM = congenital pulmonary airway malformation; CTEPH = chronic thromboembolic pulmonary hypertension; CXR = chest radiograph; DIPNECH = diffuse idiopathic pulmonary neuroendocrine cell hyperplasia; EDAC = excessive dynamic collapse of airways; PA = pulmonary artery; PAH = pulmonary arterial hypertension; PE = pulmonary embolism; TBM = tracheobronchomalacia; V/Q = ventilation perfusion scan

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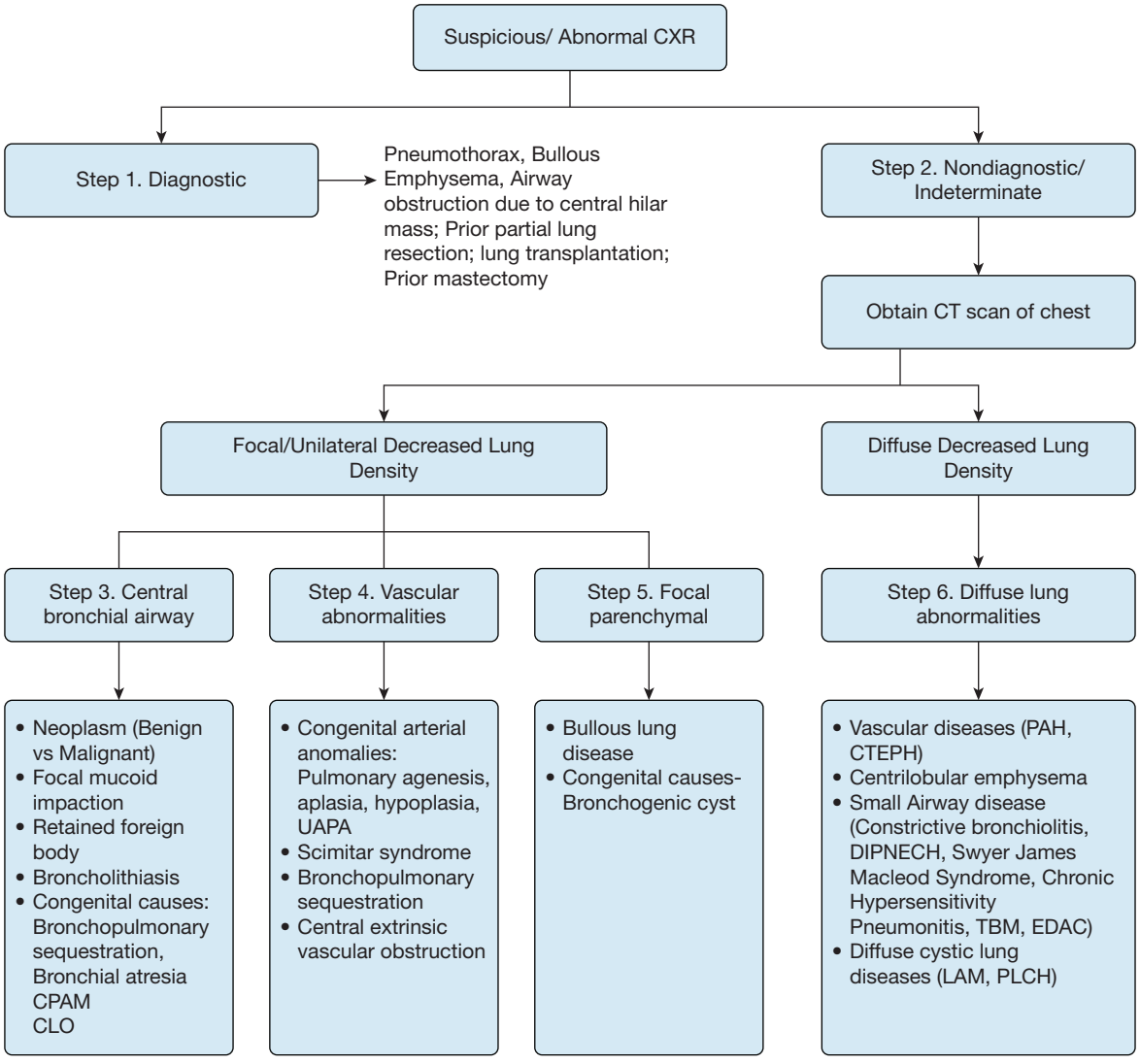

Figure 1 – *Flowchart showing algorithmic approach to pulmonary hyperlucency. CLO = congenital lobar overinflation; CPAM = congenital pulmonary airway malformation; CTEPH = chronic thromboembolic pulmonary hypertension; CXR = chest radiograph; DIPNECH = diffuse idiopathic pulmonary neuroendocrine cell hyperplasia; EDAC = excessive dynamic collapse of airways; LAM = lymphangioleiomyomatosis; PAH = pulmonary arterial hypertension; PLCH = pulmonary Langerhans cell histiocytosis; TBM = tracheobronchomalacia; UAPA = unilateral absence of the pulmonary artery.*

disorders while acknowledging that several entities may appear in more than one category.

Step 1: Routine Diagnostic Radiographic Interpretation

Routine CXRs are typically the first diagnostic test that suggest regions of diminished lung density. Initial interpretation should include excluding pseudo causes of hyperlucency because of technical factors. A rotated posterior-anterior view (Fig 2) may result in artifactual decreased attenuation on the side rotated anteriorly because of a shorter distance between the patient and the incident x-ray beam. Asymmetrical distance between the medial clavicles and spinous processes should alert to this possibility.[1,8]

A skinfold artifact caused by air trapped between skinfolds can also artifactually produce a relative hyperlucency of one lung that may simulate a pneumothorax. The absence of a thin opaque visceral pleural edge, an anatomic configuration atypical for pleural air, and extension of the artifact beyond the thoracic cavity are helpful findings to aid recognition (Fig 3).

A number of well-described etiologies for the appearance of asymmetrical hyperlucent lung are typically diagnosed on presentation (Table 1). These most often include the following: pleural causes

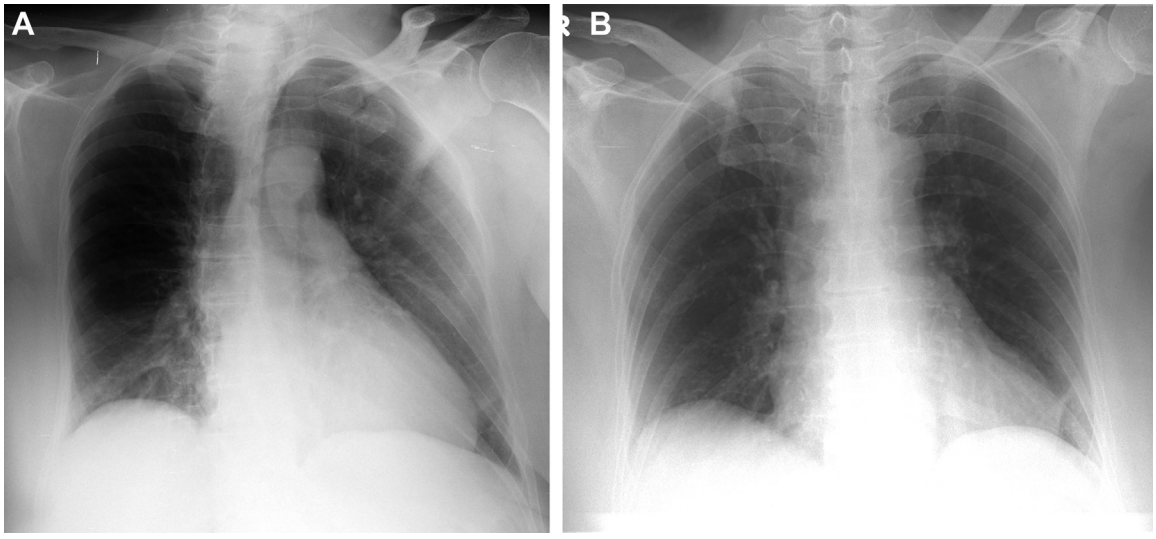

Figure 2 – *A, Portable anterior-posterior chest radiograph in a 75-y-old man with right anterior oblique positioning. The right hemithorax appears more hyperlucent because of increased x-ray transmission from closer proximity to the x-ray beam. B, Repeat nonrotated chest radiograph in the same patient showing symmetrical appearances.*

including unilateral pneumothorax (spontaneous [Fig 4], posttraumatic, or iatrogenic) and parenchymal causes, especially bullous emphysema (Fig 5A, 5B); central airway tumors resulting in partial airway obstruction with resultant peripheral hyperinflation; and postsurgical etiologies including prior partial lung resection, lung transplantation (Fig 6), and a prior mastectomy (Fig 7). Less common etiologies include various chest wall anomalies, such as congenital absence or hypoplasia of the pectoral muscles (Poland syndrome [Fig 8] or surgical removal of muscles from the chest wall to be used as muscle flaps [Fig 9]).[1,9] In most cases, when correlated with the history and physical examination, the underlying etiology is suggested on routine radiographs. Of course, follow-up CT examinations may still be warranted in select cases for further evaluation.

Step 2: Indeterminate and/or Nondiagnostic Radiographic Findings

As outlined in Figure 1, in the absence of a definitive CXR diagnosis, a follow-up CT evaluation should be considered

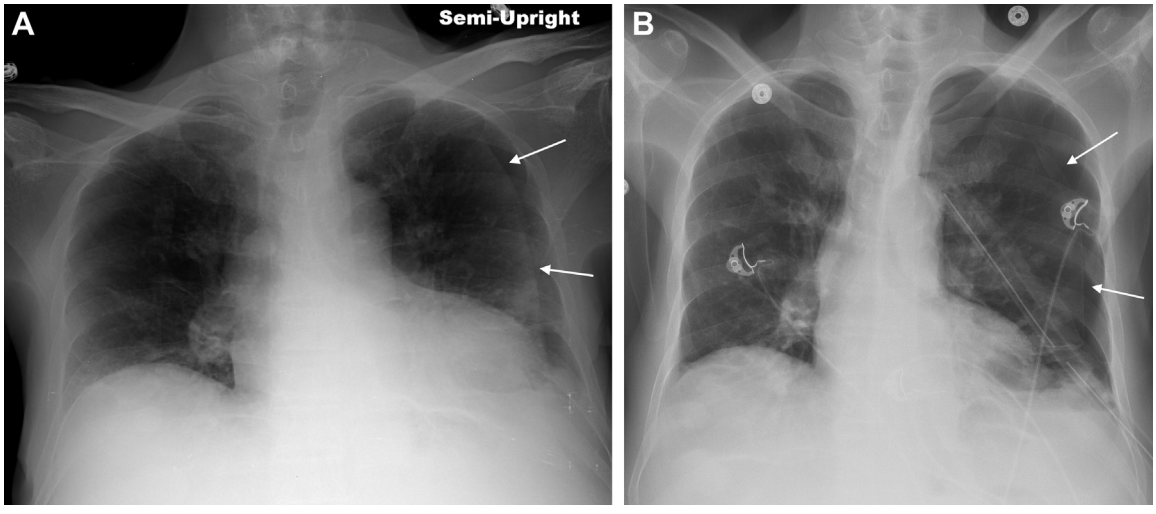

Figure 3 – *A, Portable anterior-posterior chest radiograph in a 69-y-old man with chest pain demonstrating a skinfold with no visceral pleural edge projecting over the lateral left hemithorax (arrows). The finding was misinterpreted as a left pneumothorax and prompted chest tube placement. B, Follow-up portable AP chest radiograph in the same patient after chest tube placement resulting in a new iatrogenic left pneumothorax with thin opaque visceral pleural edge now visible (arrows).*

TABLE 1] Hyperlucent Lung: Routine Chest Radiographic Diagnoses

A. Pleural disease
1. Spontaneous vs iatrogenic PTXs
2. PTX associated with underlying cystic lung diseases
B. Central airways and/or bronchial diseases
1. Acquired bronchial obstruction because of benign vs malignant endobronchial neoplasia, extrinsic compression of central airways, mucoid impaction, aspiration radiopaque foreign bodies, broncholithiasis
C. Vascular abnormalities
1. Congenital anomalies—pulmonary arterial aplasia and/or hypoplasia, sequestration, scimitar syndrome with associated hypogenetic lung
2. Extrinsic vascular compression (lung cancer, granulomatous disease)
D. Focal parenchymal disease
1. Unilateral and/or asymmetrical bullous emphysema (vanishing lung syndrome)
2. Congenital lobar emphysema
E. Diffuse lung disease
1. Centrilobular predominantly upper lobe emphysema
2. Panlobular predominantly lower lobe emphysema (alpha-1-antitrypsin deficiency)
3. Diffuse underlying cystic lung disease (LAM and/or tuberous sclerosis)
F. Chest wall anomalies
1. S/P partial lung resection, unilateral lung transplantation
2. Chest wall surgery and/or mastectomy
3. Congenital abnormality—Poland syndrome
G. Technical factors

LAM = lymphangioleiomyomatosis; PTX = iatrogenic pneumothorax; S/P = status post/after.

CT Technique

A routine, non-contrast-enhanced CT scan is typically performed. A routine noncontrast CT scan is performed with contiguous axial 2- or 5-mm and contiguous 1-mm high-resolution CT lung images reconstructed using an edge-enhancing and/or high-definition reconstruction algorithm, and axial 2- or 5-mm mediastinal soft tissue images. Coronal and sagittal multiplanar reconstructions should also be obtained. Additional select end-exhalation high-resolution CT images or dynamic volumetric expiratory images can be obtained, when clinically indicated, to assess for regional air trapping or tracheobronchomalacia (TBM), respectively. Although IV contrast administration may be useful when underlying vascular pathology is suspected, many vascular anomalies are identifiable even without contrast administration. When contrast-enhanced studies are clinically indicated, a CT pulmonary angiographic technique should typically be used; however, some entities (eg, bronchopulmonary sequestration [BPS]) are better assessed during the systemic arterial phase of enhancement.[10]

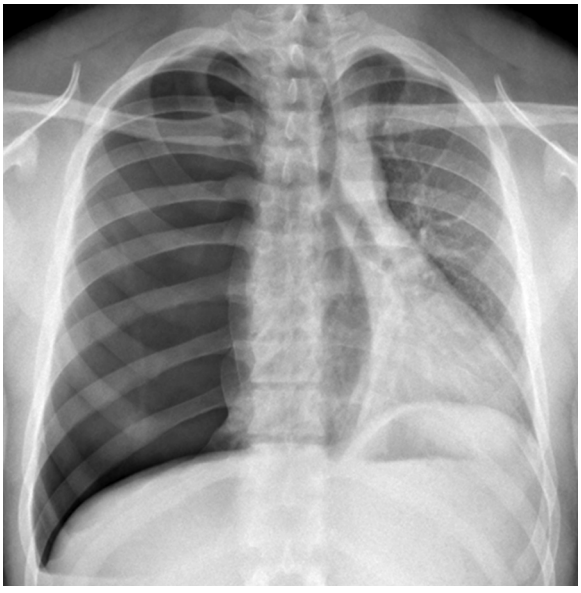

Figure 4 – *Posterior-anterior chest radiograph in a 25-y-old man presenting with acute right chest pain and dyspnea, showing spontaneous right pneumothorax, with hyperexpansion of the right hemithorax, depression of the right diaphragm, medial displacement of the partially collapsed right lung, compression of the left lung, and shift of the mediastinum and heart to the left with concave deformity of the right heart border.*

Anatomic causes of hyperlucency include possible airway, focal vascular, and parenchymal etiologies. For the purpose of organization, we focus on focal abnormalities (defined as involving one lobe of the lung), followed by diffuse abnormalities (defined as involving all five lobes of the lung).[11]

Step 3: Evaluate for Possible Central Airway and/or Bronchial Obstruction

The presence of focal and/or unilateral diminished radiographic lung density should prompt obtaining a CT scan (Table 2). The various etiologies to be considered include the following: focal airway and/or bronchial obstruction as a result of neoplastic and nonneoplastic causes including focal mucoid impaction, retained aspirated foreign bodies, and rarely, broncholiths.[1] Less commonly, congenital causes of focal airway obstruction should be considered including bronchial atresia, BPS, congenital lobar overinflation (CLO), and congenital pulmonary airway malformation (CPAM).

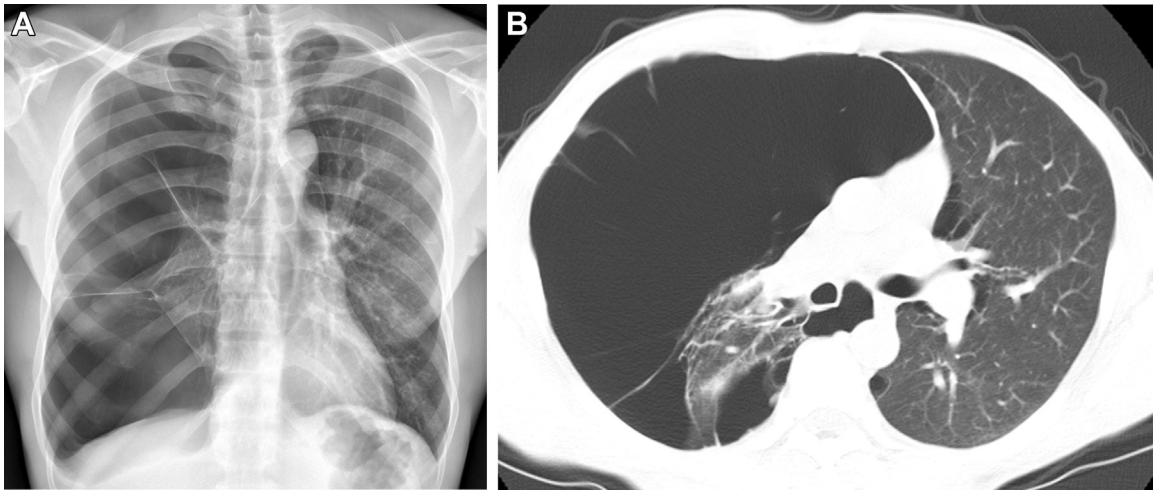

Figure 5 – *A, Posterior-anterior chest radiograph in a 44-y-old man with 10 pack-years smoking history and idiopathic giant bullous emphysema (vanishing lung syndrome) showing a hyperexpanded right hemithorax with severe bullous changes, and associated medial displacement and compression of the residual lung parenchyma in the right lower lobe. B, Axial CT image on lung windows in the same patient showing hyperexpansion and bullous replacement of the right lung with relative sparing of the compressed right lower lobe.*

Endobronchial Tumor

Most endobronchial tumors are associated with lobar or complete atelectasis because of complete occlusion of the airway. However, slow growing tumors such as typical carcinoid[2,12] (Fig 10) or less commonly adenoid cystic or mucoepidermoid cancer of the bronchus[13] may cause partial airway obstruction as a result of a secondary ball valve effect, resulting in hyperinflated lung.[12] Varying by location and size, patients may present with cough, shortness of breath, and/or hemoptysis.[12] Other benign, slow growing tumors such as bronchial leiomyoma may result in similar physiology.[14]

Mucus Plugging

Mucus plugging may cause focal hyperlucency (Fig 11) because of airway obstruction associated with collateral ventilation from adjoining lung parenchyma along with associated reflex vasoconstriction.[15] Mucoid impaction is characteristically identified radiologically as a branched (Y- or V-shaped) tubular parenchymal opacity. These typically form in preexisting foci of

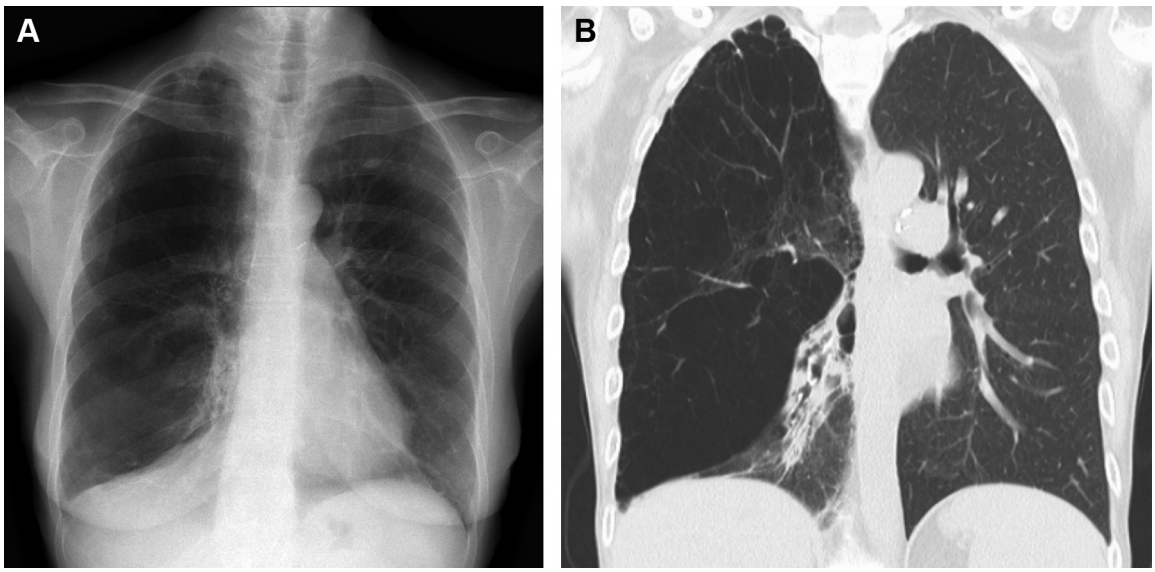

Figure 6 – *A, Posterior-anterior chest radiograph in a 66-y-old woman with severe emphysema and status post unilateral left lung transplant showing unilateral hyperlucency in native right lung with underlying emphysema. B, Coronal CT image on lung windows showing the hyperexpanded right lung with underlying emphysema and transplanted normal left lung.*

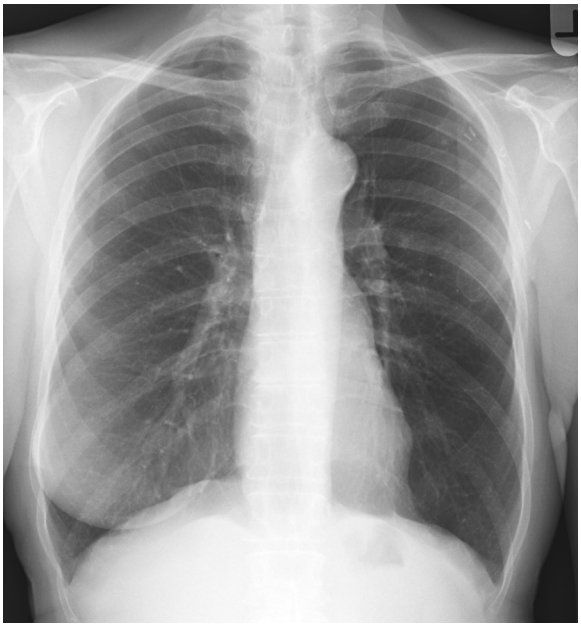

Figure 7 – *Posterior-anterior chest radiograph in a 71-y-old woman with history of left mastectomy for breast cancer demonstrating relative left-sided hyperlucency secondary to absent breast shadow.*

underlying bronchiectasis; however, the presence of underlying bronchiectasis may be obscured when involved airways are entirely filled with retained secretions. When extensive, mucus plugging may rarely be associated with contralateral mediastinal shift. Occasionally, a check valve mechanism may develop leading to focal air trapping and progressive distension of the affected lung segments behind the inspissated mucus, the so-called tension physiology. This is especially true when the patient is on mechanical ventilation.[16] The presence of a newly identified mucus plug should alert the physician to the possibility of an endobronchial tumor.[17]

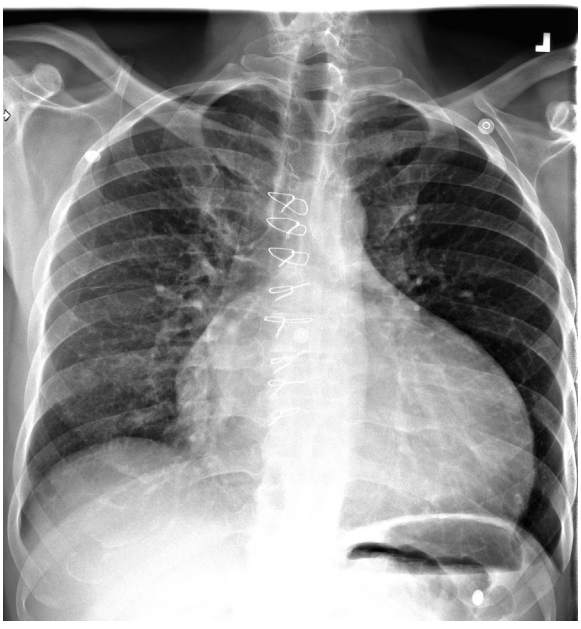

Figure 8 – *Posterior-anterior chest radiograph in a 45-y-old man with a history of nonischemic cardiomyopathy and Poland syndrome showing relative hyperlucency in the left hemithorax.*

Foreign Body Aspiration

Foreign body aspiration, although most commonly seen in children < 3 years, may result in pulmonary hyperlucency in adults, especially those with impaired consciousness, swallowing disorders, and/or postsurgical abnormalities of the pharynx.[1,2]

Aspirated foreign bodies are seen more commonly on the right side given the larger diameter and more vertical course of these airways compared with the left side. Common examples include food, in particular, which is typically nonradiopaque. As a result, the only abnormality seen on CXRs (Fig 12) may be isolated or localized air trapping leading to delayed diagnosis. In these cases, images obtained during forced exhalation can be especially helpful.[2,18] The use of chest CT scan can identify the site of airway occlusion serving as a potential roadmap for possible bronchoscopic intervention.[1,19]

Bronchial Atresia

Bronchial atresia is a congenital airway abnormality that results from focal interruption of lobar, segmental, or subsegmental bronchi and is often associated with peripheral mucus impaction and a resulting mucocele. Hyperlucency in this setting is caused by abnormally formed underlying cystic lung parenchyma and air trapping. This most commonly affects the apicoposterior segment of the left upper lobe; however, any segment may be affected.[20]

Bronchial atresia is frequently asymptomatic, more common in men, and usually identified incidentally on CXRs as a select subcategory of causes of focal typically lobar air trapping, associated with proximal mucoid impaction (Fig 13).[1,2,20] Air trapping is thought to be secondary to collateral ventilation from adjoining lung parenchyma accentuated by associated intrapulmonary vasoconstriction.[20]

CPAM

CPAM, previously known as congenital cystic adenomatoid malformation, primarily results from disorganized bronchiolar proliferation. Involved small

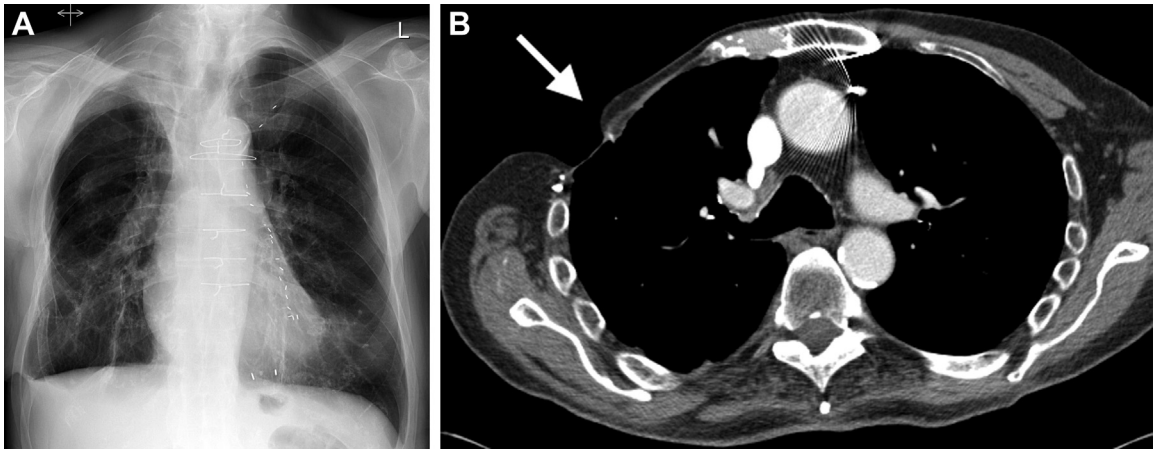

Figure 9 – *A, Posterior-anterior chest radiograph in 91-y-old man showing relative hyperlucency in the right mid- to upper zones (arrow). B, Axial CT image on soft tissue windows showing prior surgical resection of the right pectoral muscles and chest wall fat (arrow) for use as myocutaneous flap graft.*

airways communicate with the proximal bronchial tree and are thought to represent localized arrested development of the fetal bronchial tree.[21] CPAM is classified into five subtypes based on the number of cysts and the histologic resemblance to segments of the developing bronchial tree. Type 1 and type 2 CPAM

TABLE 2] Airway Causes of Pulmonary Hyperlucencies

Focal and/or Unilateral Causes	Clinical Features and Background	Radiographic Findings
Bronchial asthma	- Frequent episodes of wheezing, shortness of breath	-Mosaic pattern prominent on expiration, with associated bronchial wall thickening
Endobronchial tumor (slow growing tumors: carcinoid, mucoepidermoid cancer; benign tumors: leiomyoma)	-Cough, shortness of breath, hemoptysis -Carcinoid—asymptomatic in 25% of cases	-Usually lobar or complete atelectasis -Rarely PL restricted to lobe more prominent with expiration, with associated mucocele -Carcinoid tumors usually enhanced with contrast, and calcification seen in 30% of cases
Mucus plug	-Could be the sign of an endobronchial tumor -May cause tension physiology if patient is being mechanically ventilated	-Usually atelectasis—segmental, subsegmental, or lobar -Occasionally lobar hyperlucency more prominent on expiration
Foreign body aspiration	-More commonly in children < 3 y -May be seen in adults, especially in patients with impaired consciousness, swallowing disorders	-Lobar hyperlucency commonly on right side, more prominent on expiration
Bronchial atresia	-Frequently asymptomatic -More common in men	-Branched V- or Y- shaped parahilar opacity, representing a mucocele, with no visible communication with the central tracheobronchial tree and distal air trapping -Most commonly affects apicoposterior segment of LUL
CPAM	-May present with recurrent pneumonia, pneumothoraces, or hemoptysis -Rarely asymptomatic -Increased risk of malignancy	-Single lobar hyperlucency with multiple cysts (type 1 CPAM with cyst size > 2 cm, type 2 CPAM with cyst size < 2 cm)
Congenital lobar emphysema	-Secondary to an intrinsic cartilaginous abnormality or extrinsic compression of airway -Usually seen in early infancy, rarely seen in adult men -Presents with respiratory distress secondary to pressure effects	-PL restricted to a lobe usually in the left upper lobe, followed by right middle lobe, with mediastinal shift to the opposite side

CPAM = congenital pulmonary airway malformation; LUL = left upper lobe; PL = pulmonary hyperlucency.

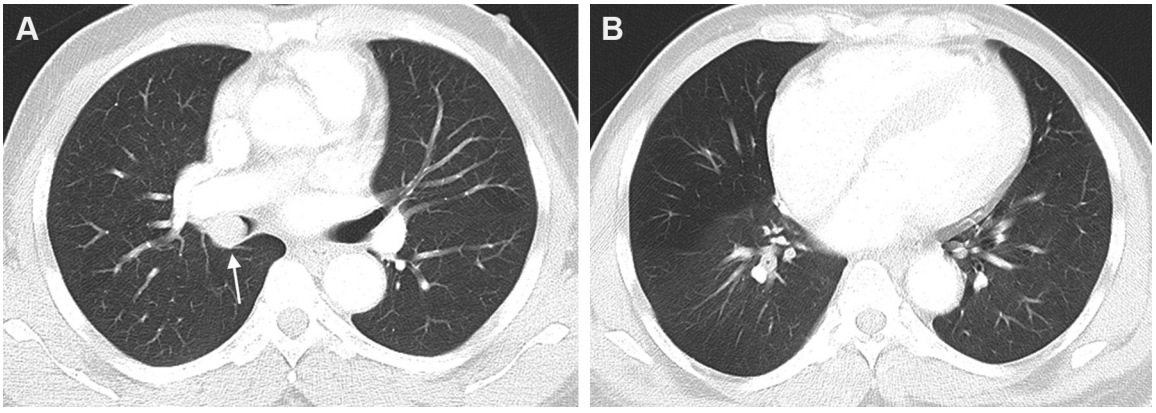

Figure 10 – *A-B, Axial CT images on lung windows in a 61-y-old man with cough secondary to a 2-cm carcinoid tumor in the bronchus intermedius (A, arrow) with associated air trapping in the right middle lobe (B).*

may present as isolated hyperinflataion, usually in a single lobe on CXR, that demonstrate multiple cysts on CT scan (Fig 14).[22]

Type 1 CPAM has either a bronchial or bronchiolar origin with large cysts > 2 cm, whereas type 2 CPAM has bronchiolar origin with smaller cysts < 2 cm.[22,23] CPAMs are usually identified early in infancy when patients present with recurrent pneumonia, pneumothorax, or hemoptysis.[21] Rarely, they remain asymptomatic and are found incidentally in adulthood.[21] CPAMs are associated with an increased risk of malignancy including pleuropulmonary blastoma and adenocarcinoma in situ.[23,24]

CLO

CLO is usually diagnosed in the neonatal period or early infancy, but may be a rare cause of hyperlucency in adults. It is characterized by lobar hyperinflation, typically of the left upper lobe, less commonly the middle lobe, and is more frequent in men.[2,22] The underlying cause, although unknown, likely results either from an intrinsic cartilaginous abnormality or extrinsic compression of the airway either secondary to an anomalous blood vessel or mass resulting in air trapping.[22]

CXRs usually demonstrate a hyperlucent, hyperexpanded lobe (Fig 15). This results in atelectasis of surrounding lung parenchyma, sometimes associated with contralateral mediastinal shift.[2] CLO usually presents with respiratory distress during early childhood, but rarely it may present in adulthood.[2,25] CLO may be misdiagnosed as a pneumothorax; however, the presence of organized lung markings help establish the diagnosis.[22]

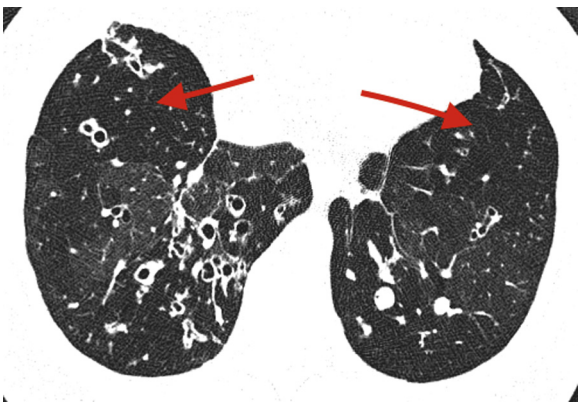

Figure 11 – *High-resolution CT image in a 61-y-old woman with history of severe chronic atypical mycobacterial infection showing bilateral lower lobe bronchiectasis, bronchial wall thickening, mucoid impaction, and associated air trapping (arrows).*

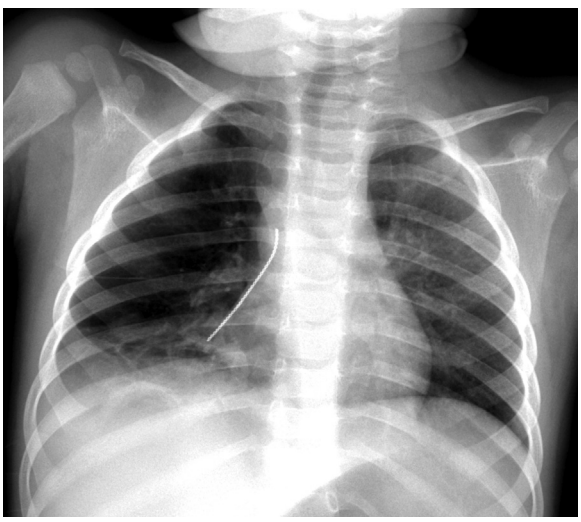

Figure 12 – *Portable chest radiograph in a 9-mo-old girl who presented with cough and fever showing hyperlucency in the right lung and a linear radiopaque foreign body. A pipe cleaner was retrieved from the right mainstem bronchus on bronchoscopy.*

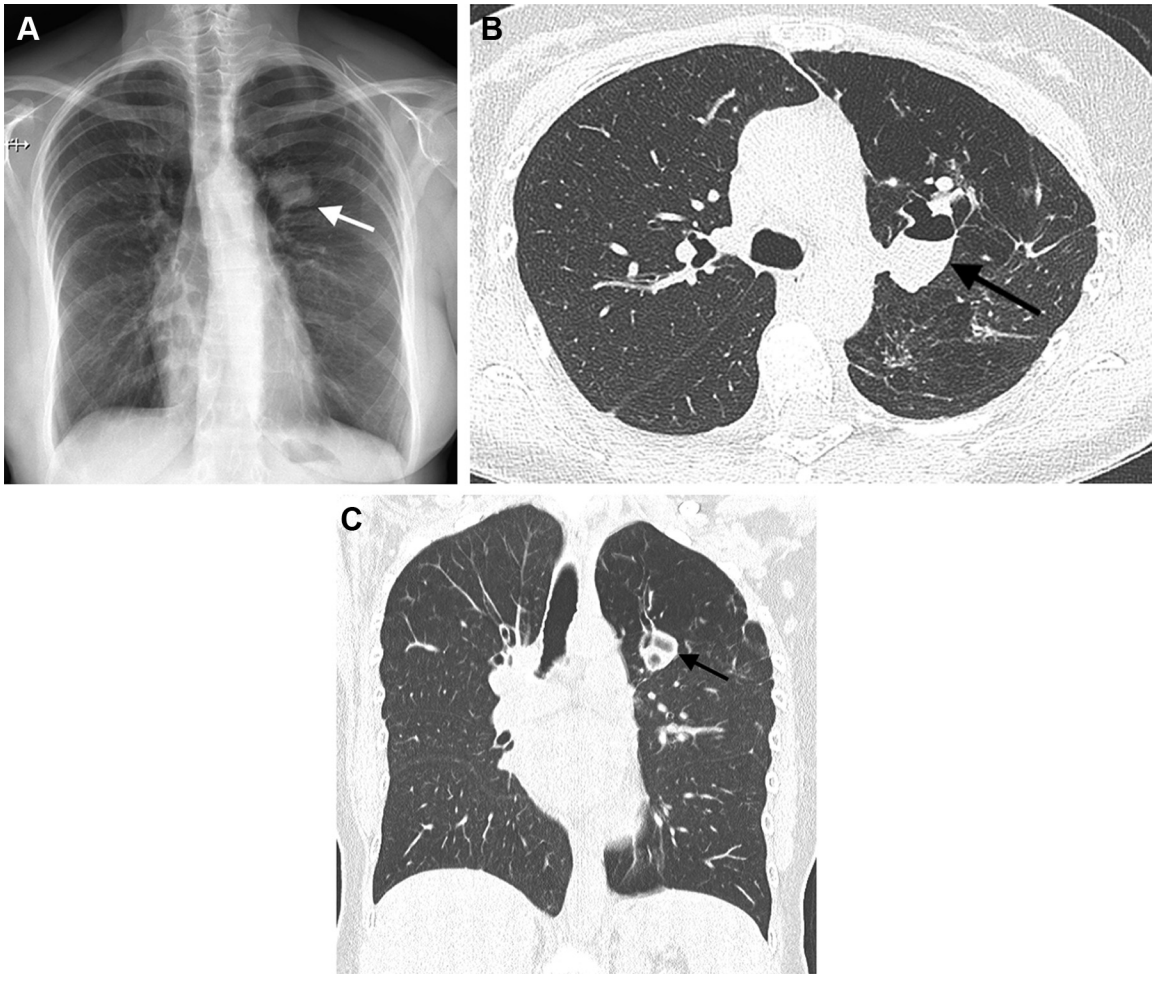

Figure 13 – *A, Posterior-anterior chest radiograph in 55-y-old woman with a history of bronchial atresia showing hyperlucent left upper lobe and a well-defined rounded mass-like opacity in the left suprahilar region (arrow) correlating with a bronchocoele. B, Axial and (C) coronal CT images on lung windows in the same patient showing left upper lobe air trapping and a blind ending dilated fluid-filled left upper lobe bronchus (arrows).*

Step 4: Evaluate for Possible Vascular Etiologies

Vascular etiologies with resultant oligemia are important causes of diminished lung density (Table 3). These include a number of congenital abnormalities, typically characterized by structural vascular anomalies associated with a reduction in ipsilateral lung volume, a small hilum with ipsilateral shift of the mediastinum, and compensatory hyperinflation of the contralateral lung. Also common are extrinsic compression or scarring causing vascular narrowing or encasement, as occurs in cases of advanced sarcoidosis or fibrosing mediastinitis. In contrast, most other acquired causes of decreased lung density typically result in bilateral, diffuse oligemia (subsequently discussed after Step 6). It should be emphasized that identification of anomalous vasculature typically is best made on CT scan by viewing mediastinal rather than lung windows.

Pulmonary Arterial Anomalies

Pulmonary agenesis, aplasia, and hypoplasia refer to congenital lung underdevelopment with a variable amount of lung and vascular tissue.[23,26] The cause for these conditions is not well understood.

Pulmonary agenesis refers to the total absence of the lung, bronchus, and pulmonary artery (PA), whereas pulmonary aplasia is characterized by congenital absence of lung and the PA, with a remaining rudimentary bronchus. CT scan is needed to differentiate between the two.[26,27]

Pulmonary hypoplasia is characterized by the presence of both a PA and bronchus, but both are hypoplastic, with a variable degree of development of associated lung parenchyma.[23,26,27]

Proximal interruption of the PA or agenesis of the PA (Fig 16) appears similar to pulmonary hypoplasia with

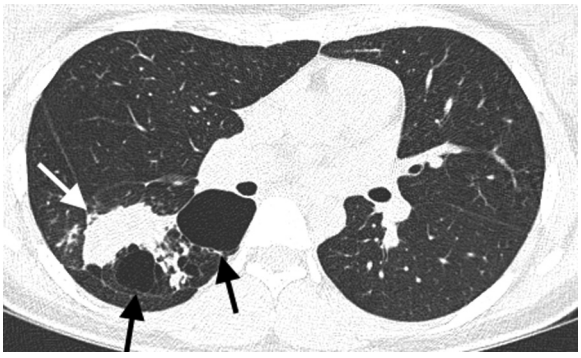

Figure 14 – *Axial CT image on lung windows in a 40-y-old woman with congenital pulmonary airway malformation showing a large mixed soft tissue and cystic lesion involving the posterior segment of the right upper lobe and superior segment of the right lower lobe (arrows).*

hypoplastic lung parenchyma and PA; however, the bronchial anatomy is normal.[26-28]

Unilateral agenesis, aplasia, and hypoplasia of the lung can affect either side, but is more common on the left, whereas proximal interruption of the PA however is almost always on the right.[26] All of these anomalies are compatible with life and have been diagnosed incidentally in asymptomatic adults or in association with recurrent infections and/or pneumothoraces.[26,29-33]

BPS

BPS is a rare congenital tracheobronchial anomaly with nonfunctional lung tissue lacking a normal communication to the bronchial tree and PAs. Instead, the arterial supply derives from the systemic circulation, most often the aorta. There are two types of BPS: intralobar BPS (75% of cases) and extralobar BPS. Differentiation is based on the presence of separate visceral pleura encasing involved lung associated with systemic venous drainage, features diagnostic of extralobar sequestration.[28,34]

Radiographically, intralobar BPS almost always involves the lower lobes (98% of cases) appearing either as a mass or mass-like consolidation associated with recurrent infections resulting frequently in air-fluid levels.[28] BPS, especially when complicated by advanced chronic inflammation, may appear as a cystic lesion,[35] which is identified as an area of focal hyperlucency with air trapping (Fig 17) caused by the collateral air drift from neighboring lung.[10,28] The key to the diagnosis on CT scan is identification of anomalous arterial supply which typically can be identified even with non-contrast-enhanced mediastinal windows. Clinical manifestations include recurrent infections and pneumonia; however, up to 15% of cases may remain asymptomatic and are discovered incidentally on chest imaging.[34]

Scimitar Syndrome

Scimitar syndrome or hypogenetic lung syndrome is a rare congenital anomaly consisting of hypoplasia of the right lung and PA, with ipsilateral partial anomalous pulmonary venous connection and anomalous systemic arterial supply characteristically involving the right lung.[26,28,36]

This condition often can be definitively diagnosed on CXR by the presence of a characteristic vertically oriented curvilinear opacity paralleling the right heart border, correlating with an anomalous pulmonary vein draining to the inferior vena cava, or less commonly the right atrium or portal vein. The anomalous pulmonary

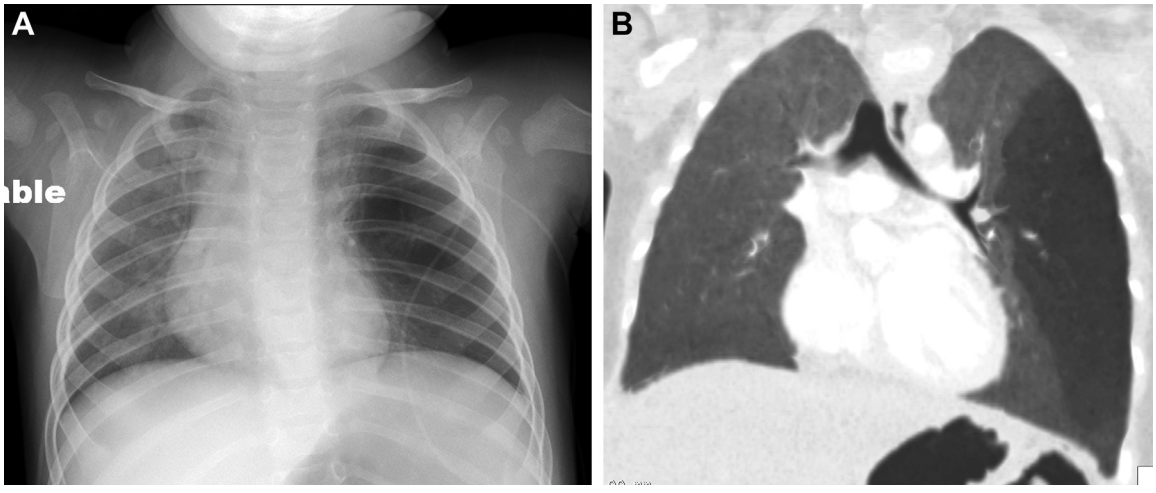

Figure 15 – *A, Anterior-posterior chest radiograph in a 7-mo-old boy with a history of congenital lobar overinflation showing hyperexpansion and hyperlucency of the left hemithorax. B, Coronal minimum intensity projection image on lung windows in the same patient showing relatively diffuse air trapping in the left upper lobe and lingula with sparing of the apex and left lower lobe and normal central airway anatomy.*

TABLE 3] Vascular Causes of Pulmonary Hyperlucency

Vascular Causes	Clinical Features and Background	Radiographic Findings
Pulmonary agenesis, aplasia, and/or hypoplasia complex	-Recurrent respiratory infections -Asymptomatic in minority	-Unilateral volume loss commonly on left side with mediastinal shift and compensatory hyperinflation of contralateral lung with herniation to the affected side
Unilateral absence of PA	-Recurrent respiratory infections, shortness of breath -PAH in 20%-44% -Rarely asymptomatic	-Ipsilateral volume loss on right side with mediastinal shift to right side -Absent PA
Bronchopulmonary sequestration	-Recurrent respiratory infections and pneumonia -Asymptomatic and incidental findings in 15% of cases	-Mass or opacity with rarely air-fluid level in the lower lobes -May be seen as focal hyperlucency with air trapping
Scimitar syndrome	-Recurrent respiratory infections, hemoptysis, PAH -Asymptomatic in 30%-40% of cases	-Scimitar vein along right heart border -Mediastinal shift to right side -Right lung appears hyperlucent likely secondary to decreased vascularity
Acute pulmonary embolism	-Sudden-onset shortness of breath with or without pleuritic chest pain	-PL known as Westermark sign has a sensitivity of 14% and specificity of 92% for acute PE Acute PE: CT scan: expansile filling defect within central portion of the vessel lumen with or without signs of right heart strain, with or without pulmonary infarcts with normal vascular caliber
CTEPH and/or PAH	-Chronic shortness of breath -H/o PE and/or DVT in case of CTEPH -Untreated PAH and/or CTEPH will progress to right heart failure	-V/Q is the most sensitive test for diagnosis -Mosaic appearance on CT scan in 50% of cases with CTEPH and 15% of cases with PAH -PAH: mosaic appearance is characterized by perivascular and patchy areas of GGO; abruptly tapering or corkscrew vessels -CTEPH: well-demarcated segmental or subsegmental distribution of mosaicism; blood vessels show nonexpansile mural-based adherent thrombus, eccentric wall thickening, and webs

CTEPH = chronic thromboembolic pulmonary hypertension; H/o = history of; GGO = ground-glass opacity; PA = pulmonary artery; PAH = pulmonary arterial hypertension; PE = pulmonary embolism; V/Q = ventilation/perfusion scan. See Table 2 legend for expansion of other abbreviation.

vein has been likened to a type of Turkish sword (based on its vertical curvilinear orientation), hence the term scimitar, from which it derives its name.[26,28]

Like the other congenital pulmonary arterial anomalies, there is associated compensatory hypertrophy of the contralateral lung with ipsilateral mediastinal shift.

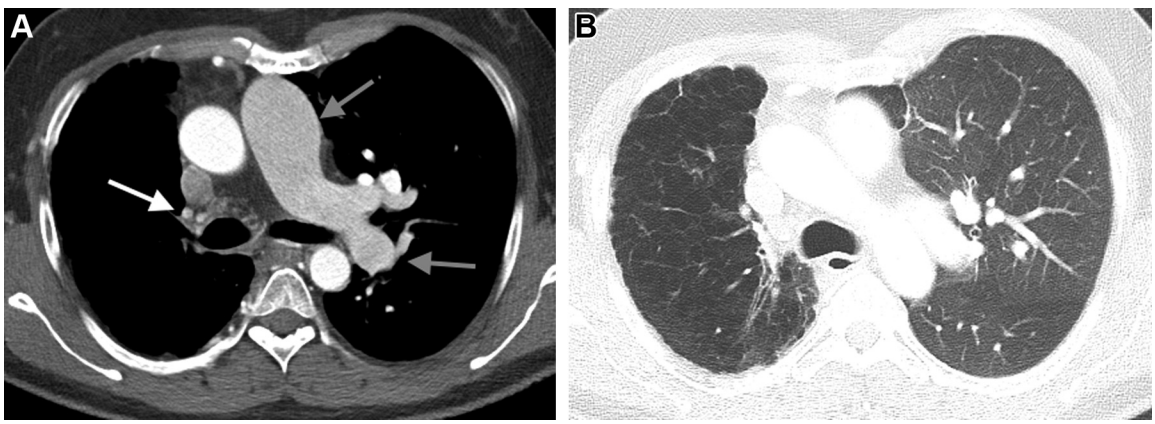

Figure 16 – *A, Axial contrast-enhanced CT image on mediastinal windows in a 35-y-old man with congenital proximal interruption of the right pulmonary artery showing complete absence of the right pulmonary artery (PA), dilatation of the main PA and left PA (arrows), and bronchial artery collaterals at the right hilum (arrow). B, Axial CT image on lung windows in the same patient showing decreased volume and hyperlucency in the right lung.*

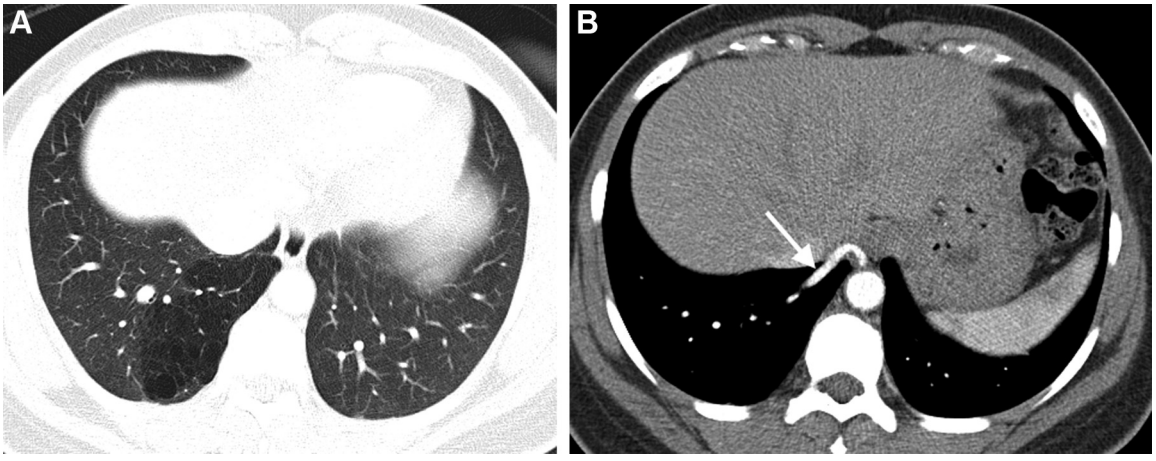

Figure 17 – *A, Axial CT image on lung windows in a 45-y-old asymptomatic woman showing focal air trapping and mild cystic changes in the medial and posterior basilar segments of the right lower lobe. B, Axial contrast-enhanced CT image on mediastinal windows in the same patient showing aberrant arterial supply from the distal descending thoracic aorta consistent with bronchopulmonary sequestration (arrow).*

However, the hyperlucency is more pronounced in the affected right lung, likely secondary to decreased vascularity—findings best appreciated on corresponding CT scan.[26,28] The clinical spectrum ranges from repeated infections and right heart failure when found in infants, to an incidental finding on CXRs in young adults, or with symptoms of dyspnea depending on the degree of left-right shunting with consequent PAH.[36]

Pulmonary Embolism

In both acute and chronic pulmonary embolism (PE), there is associated regional decrease in pulmonary blood volume secondary to obstruction of a lobar or segmental PA. This presents as hyperlucency on plain CXRs, referred to as Westermark sign (Fig 18),[1,37] with a reported sensitivity of 14% and specificity of 92% for acute PE.[38]

Acute PE is differentiated from chronic PE based on imaging characteristics and the clinical context. Both appear as filling defects on contrast-enhanced CT scans, but the vessel caliber is maintained after acute PE (Fig 19), whereas abrupt reduction in vascular caliber is seen along with other intimal irregularities, bands, webs, and rarely calcification in patients with chronic PE.[39] Another feature differentiating acute from chronic PE is the finding that the clot forms an acute angle with the vessel wall in acute PE vs an obtuse angle in chronic PE (Fig 20).[39]

Step 5: Evaluate for Focal Parenchymal Etiologies

In this category are a large number of diverse focal parenchymal causes of increased parenchymal destruction (Table 4). This varies from underlying cavitary or cystic lung diseases[11] to extensive underlying emphysema associated with smoking or recreational drug use or even underlying congenital syndromes as occurs in patients with a family history of recurrent spontaneous pneumothoraces, or the finding on physical examination of fibrofolliculomas and angiofibromas.[11] Alternatively, a history of prior chest trauma or infection increases the possibility of a pneumatocele, whereas compensatory hyperinflation may be identified after lobectomy.

Clearly, given these widely disparate etiologies, diagnosis requires correlation with meticulous history and physical examination.

Bullous Lung Disease

Bullae by definition are areas of focal paraseptal emphysema $>$ 1 cm in diameter, bounded by a very thin wall.[11] Giant bullous lung disease, or vanishing lung syndrome, refers to the presence of large bullae, which occupy at least one-third of the hemithorax with compression of the surrounding lung parenchyma.[40,41] This rare form of bullous disease is more commonly seen unilaterally, in the upper lobes and in young male smokers.[41] CXRs demonstrate asymmetrical unilateral upper lobe hyperlucency (Fig 5). Although seen mostly in smokers, especially in patients with concomitant marijuana abuse, this entity may be seen in nonsmokers,[41] and may be a presenting manifestation of Ehlers-Danlos syndrome, Marfan syndrome, and alpha-1-antitrypsin deficiency.[11,42,43] Although often asymptomatic, dyspnea and chest pain may be seen secondary to overdistension of bullae causing atelectasis of adjacent lung parenchyma. Other sequelae include

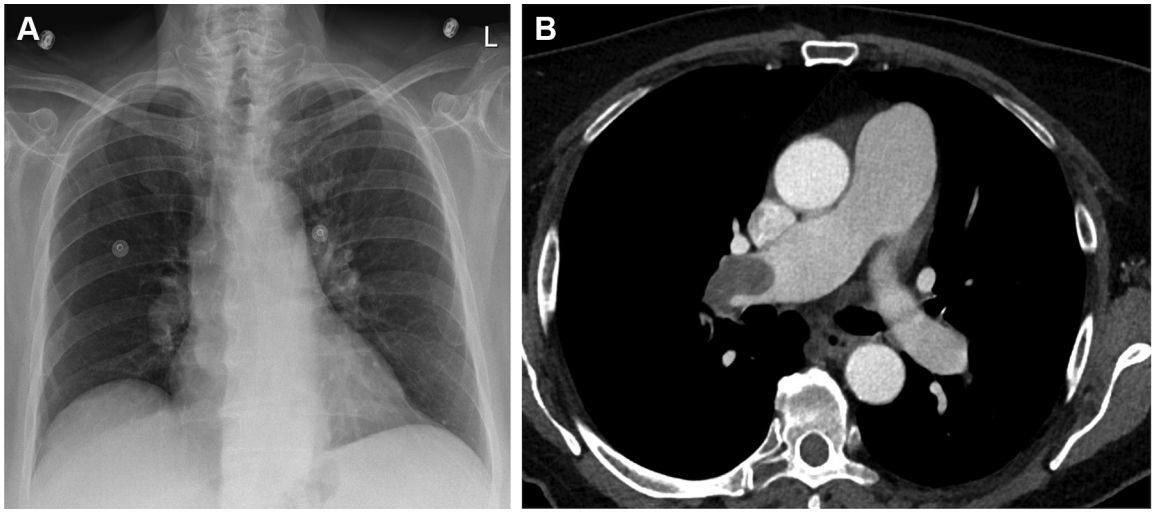

Figure 18 – *A, Posterior-anterior chest radiograph in a 76-y-old woman with shortness of breath and chest pain, demonstrating asymmetrical lucency in the right lung consistent with Westermark sign and dilated right inferior pulmonary artery (Palla sign). B, CT scans (axial view) showing large right pulmonary embolism.*

pneumothorax (Figs 21A, 21B), hemoptysis, and recurrent infections.[42]

Bullous lung disease can be easily misdiagnosed as a pneumothorax; however, pneumothorax is associated with lung collapse toward the hilum, whereas in bullous lung disease the compressed lung falls away from the hilum toward the cardiophrenic angle. In cases for which radiographic differentiation between bullous emphysema and

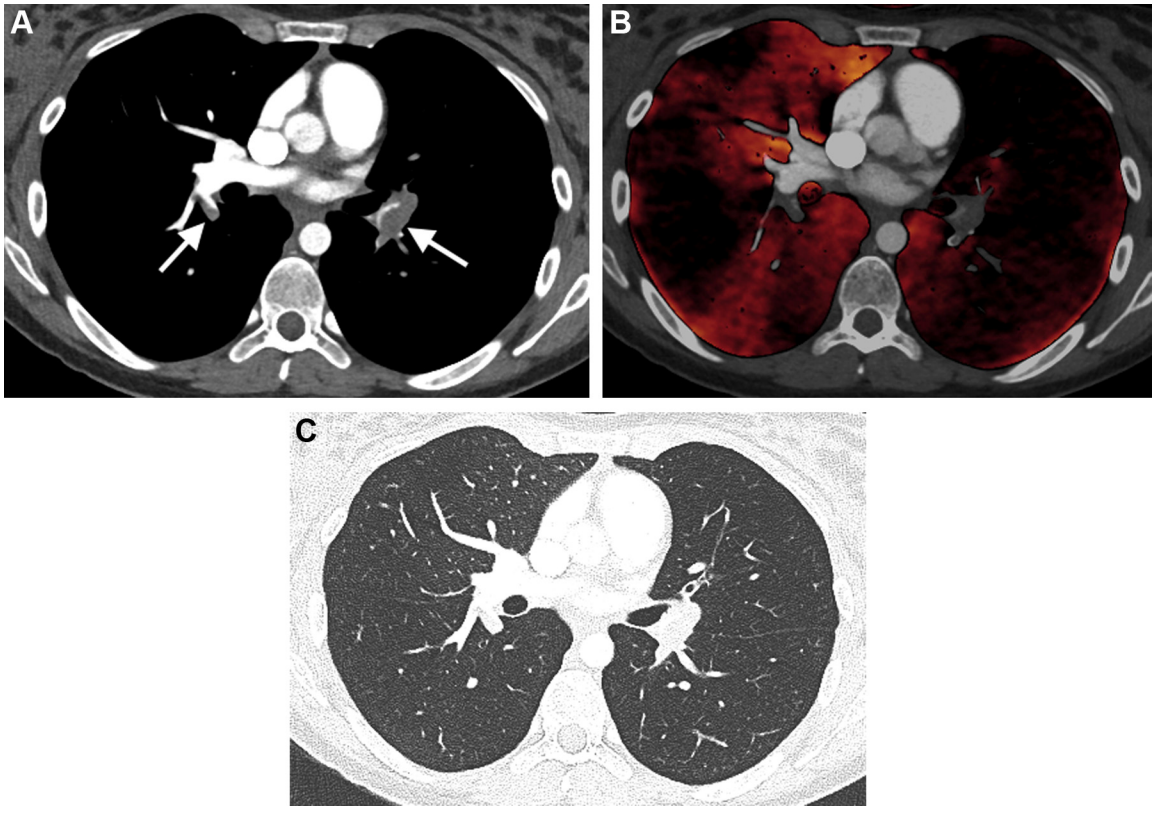

Figure 19 – *A, Axial CT pulmonary angiographic image on mediastinal windows in a 24-y-old woman with acute pulmonary emboli showing bilateral central expansile pulmonary arterial filling defects (arrows). B, Axial postprocessed iodine perfusion map in the same patient showing extensive perfusional defects in both lungs. C, Axial CT image of chest in the same patient showing no evidence of mosaicism.*

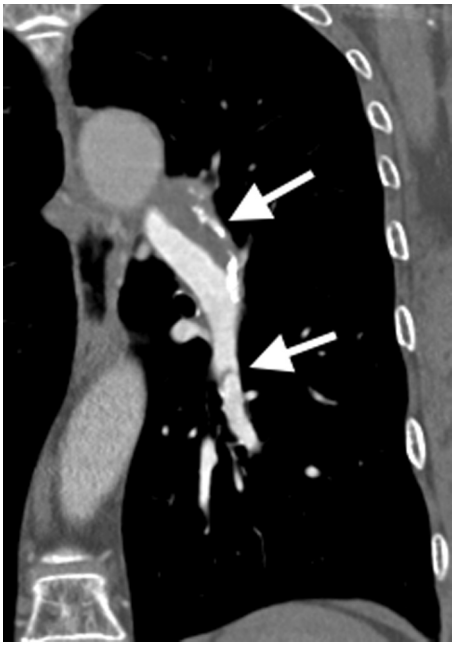

Figure 20 – *Coronal CT pulmonary angiographic image on mediastinal windows in a 72-y-old woman with history of chronic pulmonary embolism and pulmonary hypertension showing a mural-based partially calcified filling defect in the distal left main pulmonary artery (arrow) and a small web in the left lower lobe pulmonary artery (arrow).*

pneumothorax is difficult, CT scan is characteristically diagnostic.[44]

Step 6: Evaluate for Diffuse Parenchymal Etiologies

In this category are a number of disparate airway, parenchymal, and vascular entities, all of which can result in the appearance of diffuse, bilateral diminished lung density. This includes primary parenchymal etiologies such as diffuse cystic lung diseases as occurs in patients with lymphangioleiomyomatosis and/or tuberous sclerosis, or in association with severe diffuse airway pathology as occur in patients with cystic fibrosis. Also included are cases of emphysema presenting as diffuse, bilateral lung disease, including bilateral upper lobe centrilobular emphysema, and bilateral lower lobe emphysema as associated with alpha-1-antitrypsin deficiency.

Diffusely Diminished Lung Density Primarily Because of Underlying Parenchymal Disease

Cystic Lung Diseases: Diffuse cystic lung diseases, which carry a broad differential, may present as extensive bilateral decreased lung density. An algorithmic approach to this entity has been described in a previous issue of *CHEST*.[11]

Emphysema and/or Alpha-1-Antitrypsin Deficiency: Emphysema may be the most well-recognized cause for bilateral pulmonary hyperlucency. Centrilobular and paraseptal emphysema are usually in the upper lobes and are associated with cigarette smoking (Fig 22), whereas panlobular emphysema, which is seen in alpha-1-antitrypsin deficiency (AATD) (Fig 23), is associated with acinar destruction and may have a lower lobe predominance.[1] Given the enlargement of air spaces, along with reduction in caliber of pulmonary vessels (because of hypoxic

TABLE 4] Lung Parenchymal Causes of Pulmonary Hyperlucency

Parenchymal Causes	Clinical Features and Background	Radiographic Findings
Bullous lung disease	-Usually seen in young male cigarette smokers -Presenting manifestation of Ehler-Danlos syndrome, Marfan syndrome, and placental transmogrification of the lung -Often asymptomatic, but may be complicated with dyspnea and chest pain secondary to overdistension of the bulla, along with pneumothorax, hemoptysis, and recurrent infections	-Upper lobe predominant subpleural PL along with downward displacement of hilum with or without mediastinal shift
Compensatory hyperinflation	-Develops secondary to lung expansion when a part of the lung collapses or is removed -May result in respiratory limitations, if the remainder of the lung parenchyma is diseased	-PL seen with ipsilateral mediastinal shift, secondary to net volume loss
Emphysema and/or alpha-1-antitrypsin deficiency	-Most well-recognized cause of bilateral PL -Develops secondary to destruction of lung parenchyma along with hypoxic vasoconstriction -Shortness of breath and cough can lead to cor pulmonale in advanced cases	-B/l generalized PL often with lower lobe predominance (in alpha-1-antitrypsin deficiency) along with normal or enlarged hilar PA

B/l = bilateral. See Table 2 and 3 legends for expansion of other abbreviations.

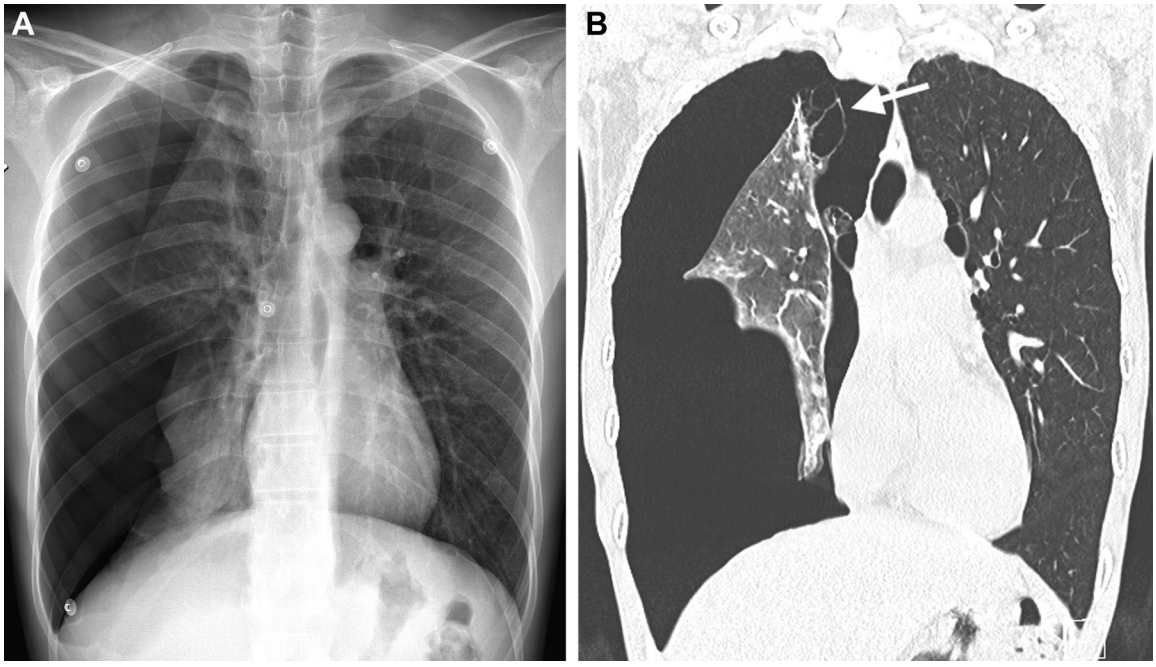

Figure 21 – *A, Posterior-anterior chest radiograph in 34-y-old man with acute right chest pain and history of daily marijuana use, demonstrating a large right pneumothorax. B, Coronal CT image in the same patient demonstrating a large right pneumothorax and relatively advanced premature paraseptal emphysema and bullous changes (arrow).*

vasoconstriction), the end result is increased, characteristically bilateral, parenchymal hyperlucency on CXRs.

AATD is an uncommon genetic condition, which predisposes to early-onset lung disease including emphysema and bronchiectasis, and chronic liver diseases including cirrhosis and hepatocellular cancer. Another rare manifestation is skin disease with panniculitis, and AATD is associated with cytoplasmic antineutrophil cytoplasmic antibody positive vasculitis.[45] Emphysema in AATD is characteristically early onset in nonsmokers (fourth to fifth decades), and as early as the third decade in smokers,[46] and may show a basilar predominance. Pulmonary function test shows airflow obstruction, often with partial reversibility. A serum level of AAT $<$ 57 mg/dL (normal range, 100-220 mg/dL) in combination with genetic testing demonstrating alleles associated with severe

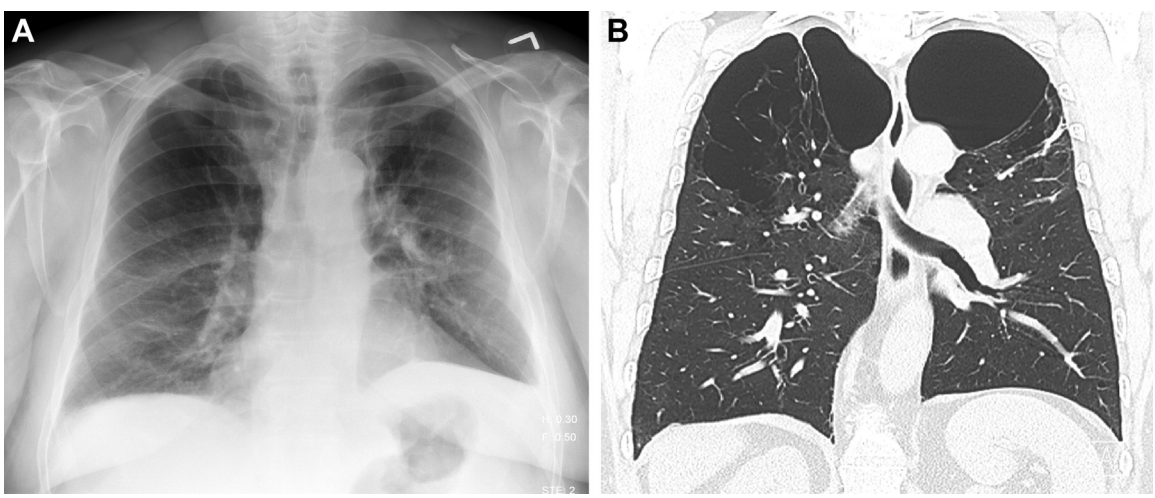

Figure 22 – *A, Posterior-anterior chest radiograph in a 63-y-old man with heavy smoking history showing relative hyperlucency in the upper zones bilaterally. B, Coronal CT image on lung windows in the same patient showing severe centrilobar and paraseptal emphysema with bullous changes, with characteristic upper lobe predominance typical for smoking-related emphysema.*

deficiency of AAT (eg, Pi*ZZ) confirms the diagnosis of AATD.[45]

Diffusely Diminished Lung Density Primarily Because of Underlying Small Airways and/or Bronchiolar Disease

Although large airway obstruction may result in air trapping involving an entire lobe or lung, small airways disease produces a so-called mosaic pattern, characterized by geographic, alternating lobular or segmental areas of dark (air trapped) and more opaque (normal) lung (Table 5). However, pulmonary vascular disease may produce an identical radiologic appearance, secondary to regional variations in perfusion (so-called mosaic perfusion). Irrespective of etiology, vessel caliber is diminished within the darker portions of the lung, and helps distinguish this appearance from geographic parenchymal ground-glass opacities that may superficially mimic this pattern (eg, in patients with *Pneumocystis jirovecii* infection). Paired inspiratory and expiratory high-resolution CT scans have been used to distinguish the mosaic attenuation of primary airways disease from primary vascular etiologies, with increased expiratory mosaicism favoring airways disease (Fig 24), and a static appearance favoring vascular disease (Fig 25).[47] However, as small airway dysfunction is commonly associated with pulmonary hypertension,[48] the assumption that expiratory

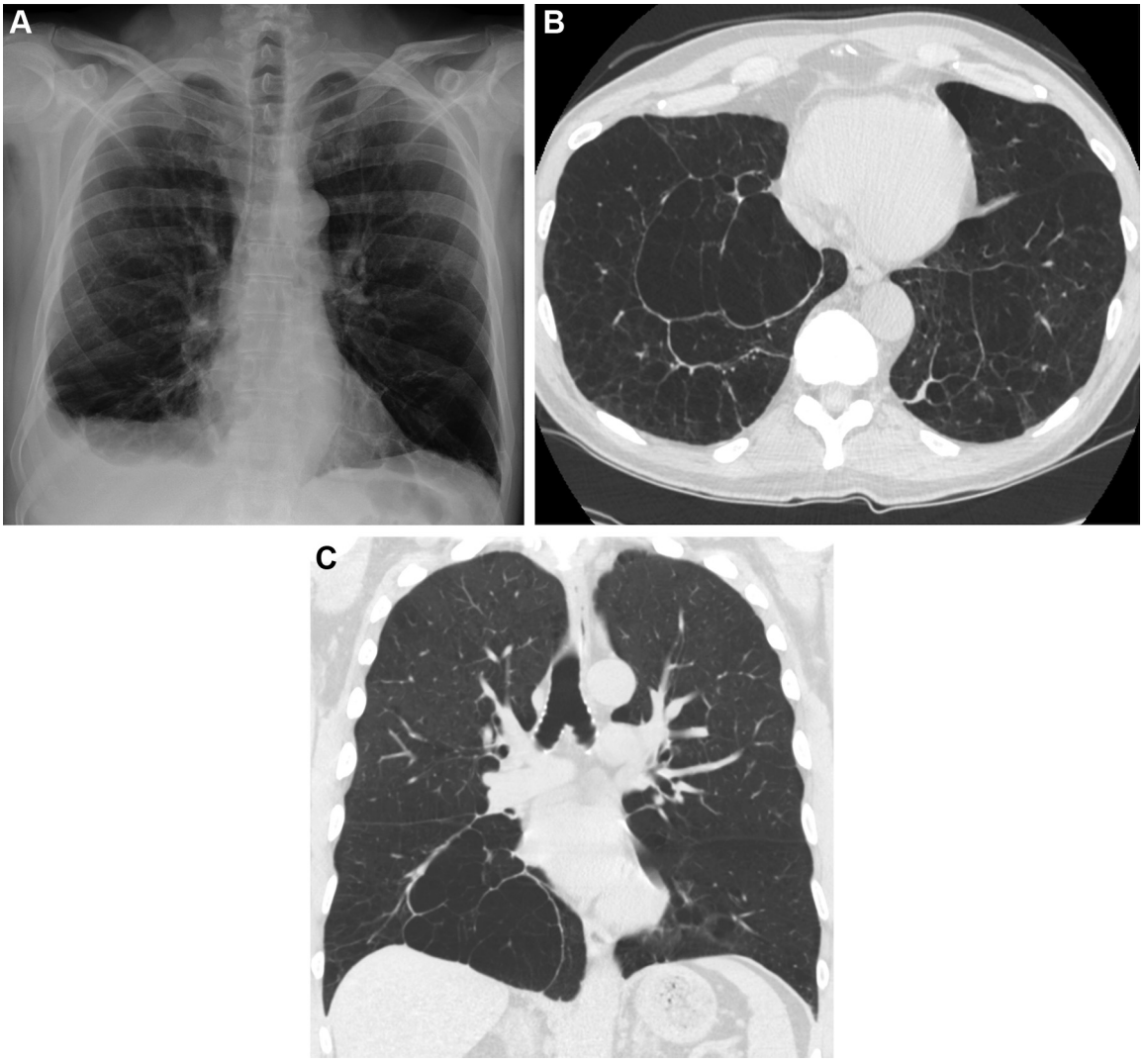

Figure 23 – *A, Posterior-anterior chest radiograph in a 56-y-old man with ZZ genotype alpha-1-antitrypsin deficiency on augmentation therapy with alpha-1 proteinase inhibitor showing lower lobe predominant hyperexpanded and hyperlucent lungs. B, Axial and coronal (C) CT images on lung windows in the same patient showing severe panacinar emphysema in the lower lobes and centrilobular emphysema in the upper lobes.*

high-resolution CT scan can always reliably distinguish these entities is likely overly simplistic, given the complex interplay of coexisting perfusional and ventilatory defects in many patients.

Constrictive Bronchiolitis

Constrictive bronchiolitis (CB) is characterized by a distinctive pattern of peribronchiolar fibrosis with cicatrization of the bronchiolar lumen with resultant extrinsic compression and obliteration of the airway. Swyer-James-Macleod syndrome (subsequently detailed) is an example of CB acquired in childhood with resultant maldevelopment of the lung. Other conditions commonly associated with CB (Table 6) include connective tissue diseases, previous infections, toxic fume inhalation, post-lung and -bone marrow transplantation, and a wide variety of drugs. Importantly, CB is only rarely idiopathic.[49,50]

CXRs may show bilateral hyperinflation and hyperlucency. CT scans show a characteristic mosaic pattern of lung attenuation, frequently identifiable even on inspiratory images; however, findings are more often diagnostic on corresponding expiratory images. The clinical course in most patients with CB tends to be progressive with poor response to corticosteroids.[49]

Swyer-James-Macleod Syndrome

Swyer-James-Macleod syndrome is a rare syndrome of CB after a lower respiratory tract infection in childhood.[51] Infectious agents commonly implicated include adenovirus, respiratory syncytial virus, and mycoplasma.[2] This causes arrested development of distal lung parenchyma, beyond the bronchioles, along with hypoplasia of the ipsilateral PA.[51] Moreover, review of the reported cases seems to show a more common occurrence in the left side, particularly the left lower lobe.[51] In fact, although frequently appearing unilateral on CXRs, Swyer-James-Macleod syndrome almost always involves both lungs, albeit asymmetrically, usually associated with

TABLE 5] Airway Causes of Diffusely Decreased Lung Density (Bilateral Pulmonary Hyperlucencies)

Airway Causes	Clinical Features and Background	Radiographic Findings
Constrictive bronchiolitis	-Peribronchiolar fibrosis and cicatrization of bronchiolar lumen, secondary to a variety of causes (Table 6) -Progressive shortness of breath with poor response to treatment	-Bilateral pulmonary hyperlucencies -Mosaic appearance prominent on expiration
Swyer-James-Macleod syndrome	-Usually the result of an infectious insult in childhood—adenovirus, RSV, mycoplasma -Recurrent childhood infections, bronchiectasis, hemoptysis -Rarely incident finding	-Unilateral hyperlucent lung, prominent on expiration, with associated ipsilateral mediastinal displacement (asymmetrical involvement of both lungs may be seen on CT scan) -Associated bronchiectatic airways seen
DIPNECH	-Mainly seen in nonsmoking women in fifth or sixth decades -Secondary to proliferation of neuroendocrine cells around bronchioles -Most with prolonged cough and dyspnea	-Mosaic appearance more prominent on expiration -Associated centrilobular and peribronchial nodules
TBM and/or EDAC	-May be congenital or acquired as in previous tracheostomy, endotracheal intubations, chest trauma, relapsing polychondritis, recurrent infections -Frequently misdiagnosed as bronchial asthma or COPD -Most with cough, wheezing, frequent infections	-Mosaic appearance with air trapping prominent on expiration -Associated tracheal thickening or calcification may be seen -> 70% collapse of trachea on expiration on dynamic CT scan
Hypersensitivity pneumonitis	-Secondary to repeated inhalation of organic antigens that evokes bronchiolocentric granulomatous and lymphocytic alveolitis -Clinically presents with chronic cough, shortness of breath, fatigue	-Upper and middle lobe predominant mosaic attenuation, air trapping along with centrilobular GGO in subacute HP; chronic HP shows mosaic attenuation, fibrosis (reticulation and traction bronchiectasis) in a peribronchovascular distribution, predominantly in upper lobes

DIPNECH = diffuse idiopathic pulmonary neuroendocrine cell hyperplasia; EDAC = excessive dynamic airway collapse; HP = hypersensitivity pneumonitis; RSV = respiratory syncytial virus; TBM = tracheobronchomalacia. See Table 3 legend for expansion of other abbreviation.

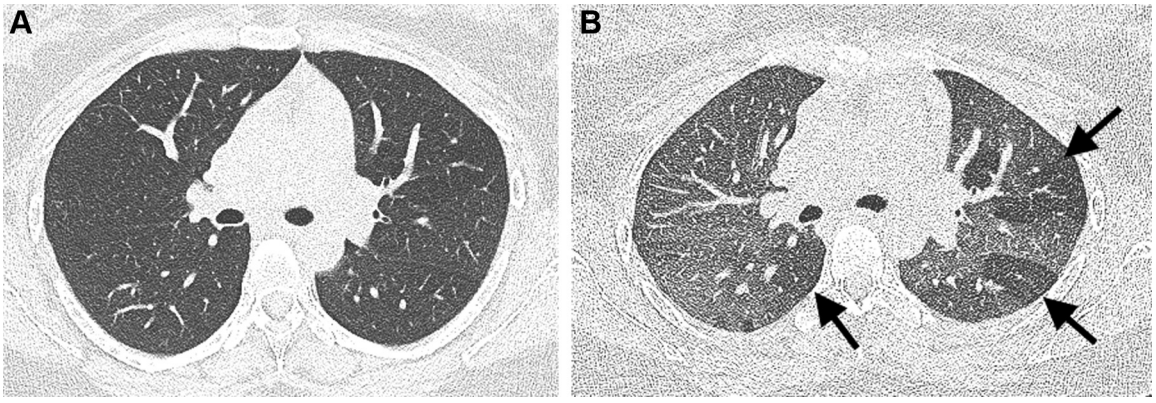

Figure 24 – *A-B, Inspiratory and expiratory high-resolution (HR) CT images in a 31-y-old woman with asthma. A, Inspiratory HR CT scan demonstrates relatively uniform lung attenuation. B, Expiratory HR CT scan demonstrates mosaic attenuation with geographic regions or darker (air trapped) lung in the left upper and both lower lobes (arrows).*

bronchiectatic foci consistent with prior underlying infection on corresponding CT scan (Fig 26).[52]

Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia

Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) is a rare, likely underrecognized, condition seen predominantly in nonsmoking women in their fifth or sixth decade.[53,54] It is characterized pathologically by the proliferation of neuroendocrine cells that involve both the terminal and respiratory bronchioles with resultant CB in many cases.[47]

CXRs may be normal, or have findings suggestive of chronic airways disease. CT scan typically shows diffuse mosaic attenuation and numerous lung nodules (Fig 27) secondary to neuroendocrine cell hyperplasia, carcinoid tumorlets (< 5 mm), and carcinoid tumors (> 5 mm).[54,55] Although sometimes an incidental finding, many patients have symptomatic airways disease and are often mislabeled as having bronchial asthma or COPD.[53,54]

Bronchial Asthma

Bronchial asthma, which is characterized by airway inflammation and reversible airway obstruction with hyperreactivity, may be associated with bilateral hyperinflation.[1,56] Although bronchial wall thickening is the most common abnormality seen on CXRs, generalized hyperinflation is usually only seen when bronchial asthma has associated small airways disease with mucus plugging or bronchiolar spasm. It is occasionally seen in patients during an acute exacerbation.[56,57] In these settings, CT scans demonstrate a mosaic pattern, which is accentuated on expiratory imaging (Fig 24).[56]

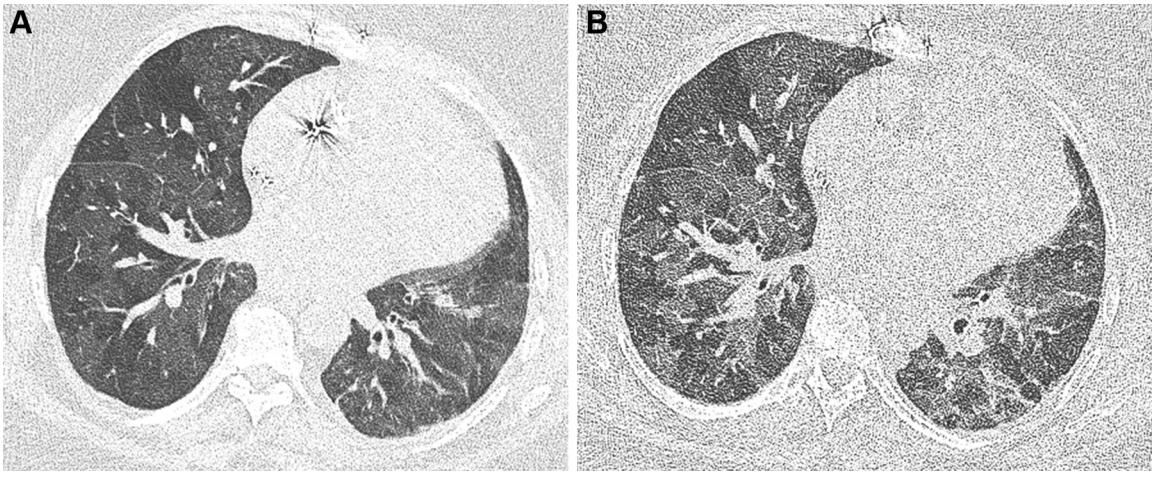

Figure 25 – *A, Inspiratory and (B) expiratory HR CT images in a 69-y-old woman with known pulmonary hypertension, showing relatively static mosaic attenuation on expiration. See Figure 24 legend for expansion of abbreviation.*

TABLE 6] Causes and Underlying Disorders Associated With Constrictive Bronchiolitis

Cause	Examples
Postinfectious	Viral—adenovirus, RSV, influenza, parainfluenza Bacterial—mycoplasma
Autoimmune disorders	Collagen vascular diseases—RA, SLE, IBD Paraneoplastic pemphigus
Transplant related	Graft vs host disease Allogenic transplant (heart-lung, lung, bone marrow transplant)
Toxic fumes	Nitrogen dioxide, sulfur dioxide, chlorine, phosgene
Toxins	Sauropus androgynus
Drugs	Cocaine, gold, penicillamine, carmustine
Cryptogenic constrictive bronchiolitis	

IBD = inflammatory bowel disease; RA = rheumatoid arthritis; SLE = systemic lupus erythematosus. See Table 5 legend for expansion of other abbreviation.

TBM and Excessive Dynamic Collapse of Airways With Secondary Small Airways Disease

TBM refers specifically to the instability or weakness of the cartilaginous portion of the airway, whereas excessive dynamic collapse of airways (EDAC) refers to the airway narrowing caused by the encroachment of a slack posterior membrane into the airway lumen.[18,58] Both TBM and EDAC are frequently misdiagnosed as bronchial asthma or COPD and are associated with cough, dyspnea, wheezing, and recurrent infections. Diagnosis is based on the presence of > 70% collapse of the airways on dynamic CT scan of the chest.[58]

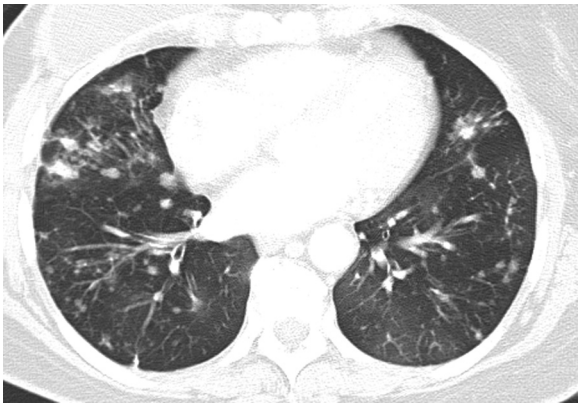

Figure 27 – *Axial CT image on lung windows in a 69-y-old woman with a history of diffuse idiopathic pulmonary neuroendocrine cell hyperplasia and right lower lobe wedge resection for carcinoid tumor showing numerous bilateral lung nodules (carcinoid tumors and tumorlets), bronchial wall thickening, and mosaic attenuation.*

Although rarely reported as a cause of pulmonary hyperlucency, it is likely underrecognized because chest CT scan frequently shows a mosaic pattern with associated air trapping (Fig 28). Although the exact mechanism is unclear, associated intrinsic defect in the cartilage of small airways or chronic inflammation of the small airways because of recurrent infections secondary to impaired clearing of secretions may explain the high proportion of small airways disease in

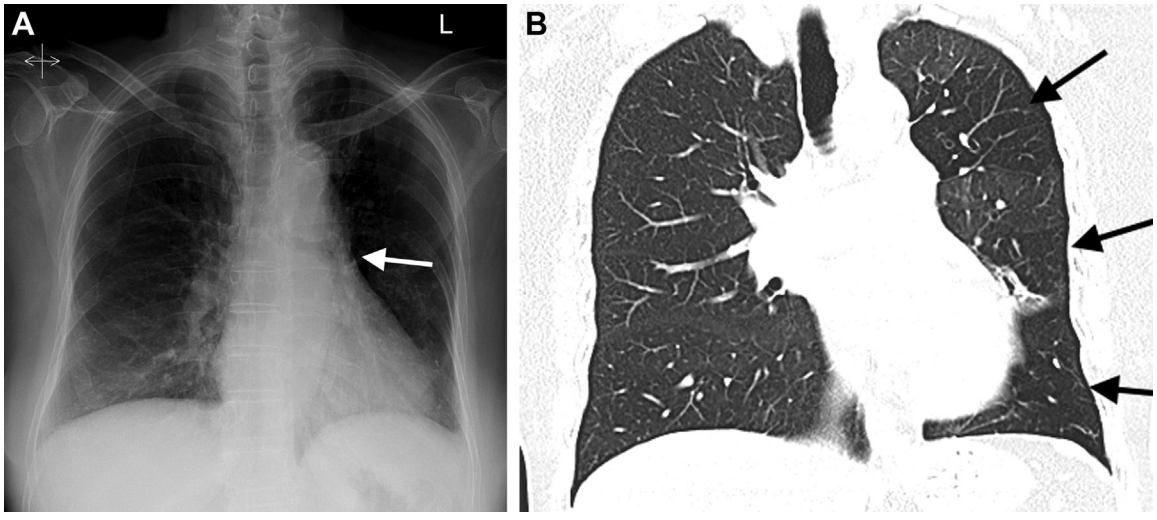

Figure 26 – *A, Posterior-anterior chest radiograph in a 73-y-old man with a history of Swyer-James-Macleod syndrome showing a smaller volume, hyperlucent left lung with smaller left hilar vascular shadow (arrow). B, Coronal CT image on lung windows in the same patient showing a smaller volume left lung with air trapping (arrows).*

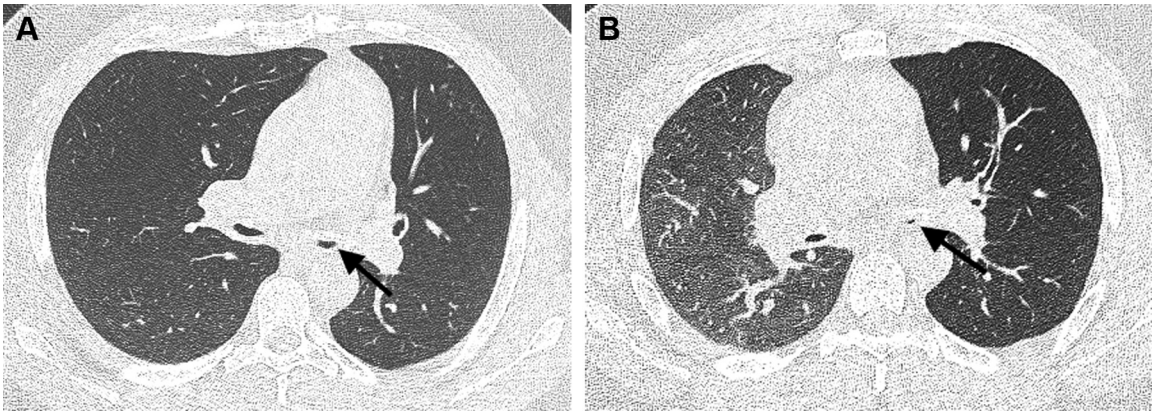

Figure 28 – *A-B, Paired inspiratory and expiratory HR CT images in a 34-y-old woman with history of relapsing polychondritis. A, Inspiratory HR CT scan shows thickened (partially calcified) cartilaginous bronchial rings and small caliber but patent left main bronchus (arrow) with uniform lung attenuation. B, Expiratory HR CT scan shows normal increased density in the right lung but complete collapse of the left main bronchus (arrow) with associated air trapping and paradoxical increased volume in the left lung. See Figure 24 legend for expansion of abbreviation.*

this group.[59,60] Two small series of TBM found air trapping and mosaic appearance in 100% of the patients.[59,60]

Hypersensitivity Pneumonitis

Hypersensitivity pneumonitis is a complex syndrome caused by repeated exposure to a variety of organic antigens in susceptible individuals. It is pathologically characterized by a bronchiolocentric granulomatous and lymphocytic alveolitis that may evolve to fibrosis in chronic advanced cases. Although numerous causative antigens have been identified, in nearly one-half of cases, no associated antigen is discovered. Clinical manifestations may reflect the severity and duration of exposure and range from insidious onset of dyspnea, fatigue, and cough, to rapid and marked respiratory insufficiency.

Acute hypersensitivity pneumonitis typically manifests as symmetric homogeneous airspace opacities, evolving to mid to upper lung predominant centrilobular ground-glass nodules in the subacute phase. In contrast, a pattern of focal airtrapping predominantly affecting discrete pulmonary lobules is frequently identified in patients with chronic fibrotic hypersensitivity pneumonitis (Fig 29). In these cases, fibrosis is additionally characterized by reticulation and traction bronchiectasis in a peribronchovascular distribution, predominantly involving the upper lobes. Diagnosis is based either on a history of exposure to known antigen and other clinical, radiologic, and histopathology findings.[61,62]

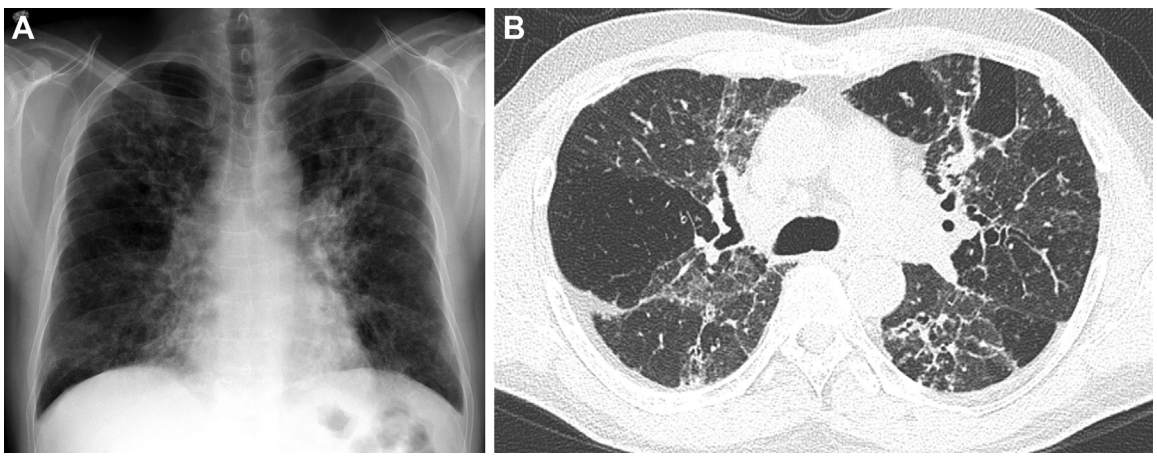

Figure 29 – *A, Posterior-anterior chest radiograph in a 56-y-old man with history of chronic hypersensitivity pneumonitis showing hyperinflation, patchy hyperlucency, coarsened markings, and central fibrotic opacities. B, HR CT image in the same patient showing mosaic attenuation and patchy ground-glass and reticular opacities typical for chronic hypersensitivity pneumonitis. See Figure 24 legend for expansion of abbreviations.*

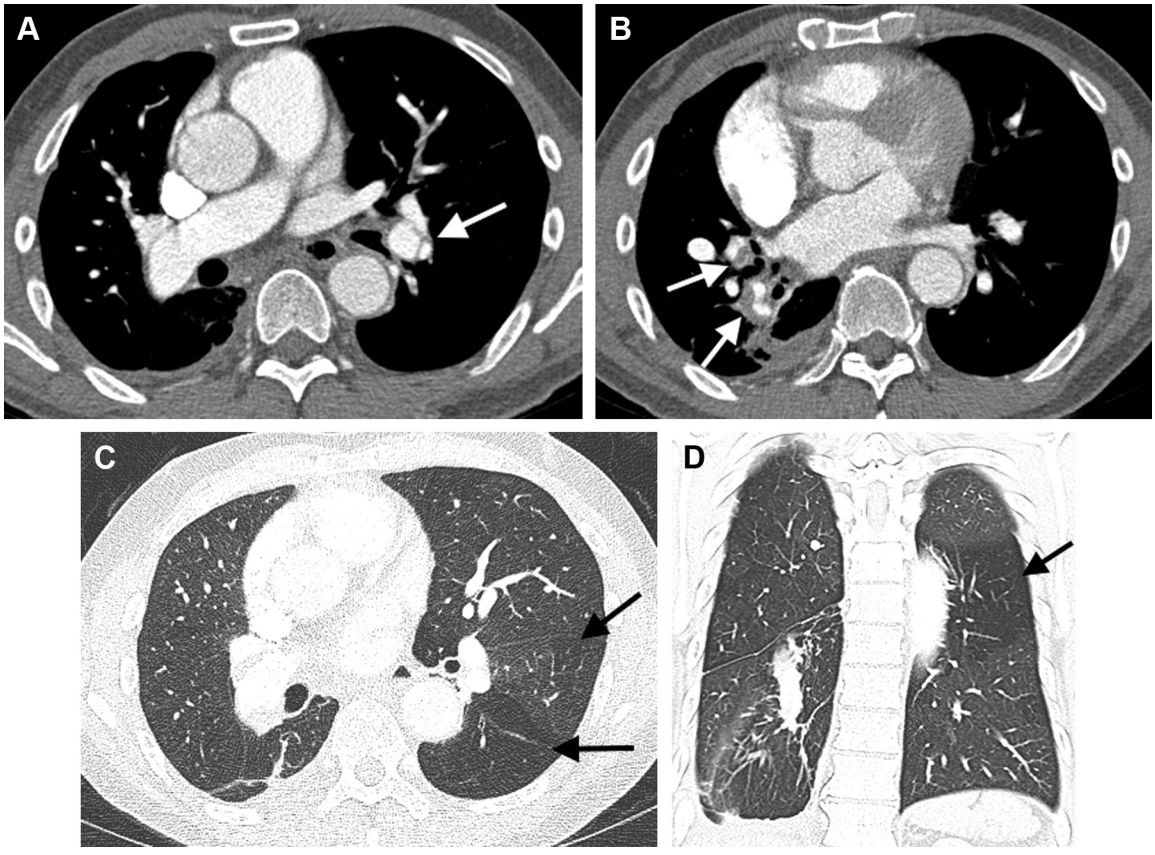

Figure 30 – *A-D, Axial CT pulmonary angiographic images on mediastinal windows in a 57-y-old man with a history of chronic pulmonary embolism and pulmonary hypertension. A, Proximal left lower lobe pulmonary artery web is shown (arrow). B, Right lower lobe mural-based segmental pulmonary arterial filling defects with associated luminal narrowing (arrows) is shown. C, Axial and coronal (D) CT images on lung windows in the same patient showing mosaic attenuation in the superior segment of the left lower lobe (arrows).*

Diffusely Diminished Lung Density Because of Underlying Vascular Etiologies

Pulmonary Arterial Hypertension and Chronic Thromboembolic Pulmonary Hypertension: Chronic thromboembolic pulmonary hypertension (CTEPH) and, less commonly, pulmonary arterial hypertension (PAH) may present on CT scan as diffuse, mosaic perfusion (Figs 30A-D). In a retrospective analysis of CT scans of 189 patients with pulmonary hypertension because of different causes, mosaic perfusion was seen in approximately 50% of patients with CTEPH, whereas only 15% of patients with idiopathic PAH demonstrated this finding.[63] Hence, although suggestive, a mosaic appearance is not especially sensitive for underlying pulmonary vascular disease, especially in the absence of enlarged pulmonary arteries. Although V/Q scans are more often diagnostic, it cannot be overemphasized that CT studies obtained for a variety of clinical indications—often including unexplained dyspnea—may be the first indication of underlying pulmonary vascular disease. V/Q scans and follow-up echocardiograms can be especially helpful in diagnosis.

Conclusions

There is a wide range of causes for focal and diffusely diminished lung density. We propose a practical combined clinical-radiographic algorithm emphasizing characteristic visual patterns of disease. Although the ultimate utility of this methodology will require further prospective validation, it is anticipated that this approach will prove of benefit in routine clinical practice.

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