

with predominant midlung field involvement include the dyskinetic/immotile cilia syndrome. Finally, predominant involvement of the central airways is reported in association with allergic bronchopulmonary aspergillosis (ABPA), in which an immune-mediated reaction to *Aspergillus* damages the bronchial wall. Congenital causes of central airway–predominant bronchiectasis resulting from cartilage deficiency include tracheobronchomegaly (Mounier-Kuhn syndrome) and Williams-Campbell syndrome.

In many cases, the etiology of bronchiectasis is not determined. In case series, as many as 25–50% of patients referred for bronchiectasis have idiopathic disease.

EPIDEMIOLOGY

The overall reported prevalence of bronchiectasis in the United States has recently increased, but the epidemiology of bronchiectasis varies greatly with the underlying etiology. For example, patients born with CF often develop significant clinical bronchiectasis in late adolescence or early adulthood, although atypical presentations of CF in adults in their thirties and forties are also possible. In contrast, bronchiectasis resulting from MAC infection classically affects nonsmoking women >50 years of age. In general, the incidence of bronchiectasis increases with age. Bronchiectasis is more common among women than among men.



In areas where tuberculosis is prevalent, bronchiectasis more frequently occurs as a sequela of granulomatous infection.

Focal bronchiectasis can arise from extrinsic compression of the airway by enlarged granulomatous lymph nodes and/or from development of intrinsic obstruction as a result of erosion of a calcified lymph node through the airway wall (e.g., broncholithiasis). Especially in reactivated tuberculosis, parenchymal destruction from infection can result in areas of more diffuse bronchiectasis. Apart from cases associated with tuberculosis, an increased incidence of non-CF bronchiectasis with an unclear underlying mechanism has been reported as a significant problem in developing nations. It has been suggested that the high incidence of malnutrition in certain areas may predispose to immune dysfunction and development of bronchiectasis.

PATHOGENESIS AND PATHOLOGY

The most widely cited mechanism of infectious bronchiectasis is the “vicious cycle hypothesis,” in which susceptibility to infection and poor mucociliary clearance result in microbial colonization of the bronchial tree. Some organisms, such as *Pseudomonas aeruginosa*, exhibit a particular propensity for colonizing damaged airways and evading host defense mechanisms. Impaired mucociliary clearance can result from inherited conditions such as CF or dyskinetic cilia syndrome, and it has been proposed that a single severe infection (e.g., pneumonia caused by *Bordetella pertussis* or *Mycoplasma pneumoniae*) can result in significant airway damage and poor secretion clearance. The presence of the microbes incites continued chronic inflammation, with consequent damage to the airway wall, continued impairment of secretion and microbial clearance, and ongoing propagation of the infectious/inflammatory cycle. Moreover, it has been proposed that mediators released directly from bacteria can interfere with mucociliary clearance.

Classic studies of the pathology of bronchiectasis from the 1950s demonstrated significant small-airway wall inflammation and larger-airway wall destruction as well as dilation, with loss of elastin, smooth muscle, and cartilage. It has been proposed that inflammatory cells in the small airways release proteases and other mediators, such as reactive oxygen species and proinflammatory cytokines, that damage the larger-airway walls. Furthermore, the ongoing inflammatory process in the smaller airways results in airflow obstruction. It is thought that antiproteases, such as α_1 antitrypsin, play an important role in neutralizing the damaging effects of neutrophil elastase and in enhancing bacterial killing. Bronchiectasis and emphysema have been observed in patients with α_1 antitrypsin deficiency.

Proposed mechanisms for noninfectious bronchiectasis include immune-mediated reactions that damage the bronchial wall (e.g., those associated with systemic autoimmune conditions such as Sjögren’s

syndrome and rheumatoid arthritis). *Traction bronchiectasis* refers to dilated airways arising from parenchymal distortion as a result of lung fibrosis (e.g., postradiation fibrosis or idiopathic pulmonary fibrosis).

CLINICAL MANIFESTATIONS

The most common clinical presentation is a persistent productive cough with ongoing production of thick, tenacious sputum. Physical findings often include crackles and wheezing on lung auscultation, and some patients with bronchiectasis exhibit clubbing of the digits. Mild to moderate airflow obstruction is often detected on pulmonary function tests, overlapping with that seen at presentation with other conditions, such as chronic obstructive pulmonary disease (COPD). Acute exacerbations of bronchiectasis are usually characterized by changes in the nature of sputum production, with increased volume and purulence. However, typical signs and symptoms of lung infection, such as fever and new infiltrates, may not be present.

DIAGNOSIS

The diagnosis is usually based on presentation with a persistent chronic cough and sputum production accompanied by consistent radiographic features. Although chest radiographs lack sensitivity, the presence of “tram tracks” indicating dilated airways is consistent with bronchiectasis. Chest computed tomography (CT) is more specific for bronchiectasis and is the imaging modality of choice for confirming the diagnosis. CT findings include airway dilation (detected as parallel “tram tracks” or as the “signet-ring sign”—a cross-sectional area of the airway with a diameter at least 1.5 times that of the adjacent vessel), lack of bronchial tapering (including the presence of tubular structures within 1 cm from the pleural surface), bronchial wall thickening in dilated airways, inspissated secretions (e.g., the “tree-in-bud” pattern), or cysts emanating from the bronchial wall (especially pronounced in cystic bronchiectasis; Fig. 312-1).

APPROACH TO THE PATIENT: Bronchiectasis

The evaluation of a patient with bronchiectasis entails elicitation of a clinical history, chest imaging, and a workup to determine the underlying etiology. Evaluation of focal bronchiectasis almost always requires bronchoscopy to exclude airway obstruction by an underlying mass or foreign body. A workup for diffuse bronchiectasis includes analysis for the major etiologies (Table 312-1), with an initial focus on excluding CF. Pulmonary function testing is an important component of a functional assessment of the patient.

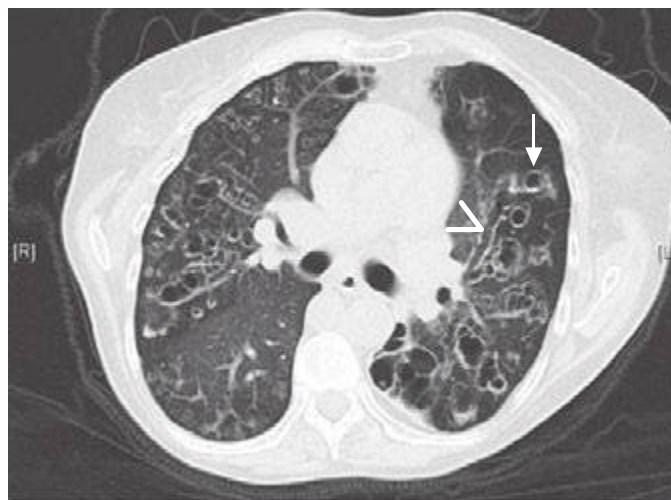


FIGURE 312-1 Representative chest computed tomography (CT) image of severe bronchiectasis. This patient’s CT demonstrates many severely dilated airways, seen both longitudinally (arrowhead) and in cross-section (arrow).