Diseases of the Trachea and Main-Stem Bronchi: Correlation of CT with Pathologic Findings¹

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This article presents the computed tomographic (CT) features of the most common abnormalities of the trachea and main-stem bronchi and correlates CT and pathologic findings. The abnormalities are classified into focal and diffuse. Focal disease tends to produce a decreased airway diameter, whereas diffuse diseases are divided into those that increase the airway diameter and those that decrease it. Conventional CT with 10-mm collimation was performed in 36 patients to assess their condition. Additional dynamic incremental thinsection (1.5-5.0-mm collimation) CT was performed in patients with focal abnormalities. Findings from conventional CT correlated closely with those from pathologic analysis of specimens from patients with diffuse disease, but dynamic thin-section scans are necessary for optimal assessment of focal abnormalities. CT demonstrates the location and extent of disease; helps characterize abnormal tissues; helps evaluate the thickness of the tracheal and bronchial walls; and helps determine the extent of extraluminal disease, including the presence of mediastinal extension and lymphadenopathy.

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■ INTRODUCTION

A number of diseases may affect the trachea and main-stem bronchi. Patients may not have any symptoms or may present with nonspecific symptoms such as cough, dyspnea, wheezing, and stridor. The clinical course is often long-term, and misdiagnosis of bronchial asthma is common.

Computed tomography (CT) can demonstrate the degree of widening or narrowing of the airway; the location and extent of tracheobronchial abnormalities; and the presence of associated mediastinal disease, postobstructive atelectasis, and pneumonitis.

This article reviews the CT features of the most common diseases of the trachea and main-stem bronchi and correlates the CT and pathologic findings. The article is based on the CT findings in 36 patients with a bronchoscopically or pathologically proved diagnosis. All patients underwent conventional contiguous 10-mm-collimation scanning. Additional dynamic incremental 1.5–5.0-mm-collimation scans were obtained in patients with focal abnormalities.

Diseases of the trachea and main-stem bronchi may be divided into those that cause focal disease and those that result in diffuse disease (1,2). Focal disease tends to produce decreased airway diameter. The diffuse dis-

Classification of Diseases of the Trachea and Main-Stem Bronchi

Focal diseases
Tracheal stricture
Benign neoplasms
Primary malignant neoplasms
Secondary malignant neoplasms
Diffuse diseases
Increased diameter
Tracheobronchomegaly (Mounier-Kuhn
disease)
Decreased diameter
Relapsing polychondritis
Amyloidosis
Sarcoidosis
Wegener granulomatosis
Tracheopathia osteoplastica
Tracheobronchitis associated with ulcer-
ative colitis
Saber-sheath trachea
Tracheomalacia and bronchomalacia
Infectious disorders

eases may be further divided into those that increase the diameter of the airway and those that decrease the diameter (Table).

FOCAL DISEASES

• Tracheal Stricture

Strictures of the trachea are usually caused by damage from a cuffed endotracheal or tracheostomy tube or trauma to the neck (1). It is hypothesized that the cuff pressure in these devices may exceed capillary pressure, lead-



Figure 1. Tracheal stricture. (a) CT scan (1.5-mm collimation) of a patient with stridor for 3 months after extubation demonstrates narrowing of the tracheal lumen. (b) CT scan obtained immediately above this level demonstrates normal tracheal diameter.



Figure 2. Squamous papilloma of the right mainstem bronchus. High-resolution (1.5-mm-collimation, high-spatial-frequency reconstruction algorithm) contrast material—enhanced CT scan obtained through the carina demonstrates an intraluminal polypoid mass in the right main-stem bronchus (arrow). The mass is well circumscribed, with no evidence of extension beyond the bronchial wall.

ing to ischemic necrosis and subsequent fibrosis (3). Stricture may also occur proximal to the cuff at the tracheostomy site.

CT scans can demonstrate the site of narrowing in most cases. However, a web or stenosis that involves a short segment may be missed because of volume averaging. CT may also result in the overestimation of the severity of a fixed stenotic segment and the underestimation of the length of the abnormal trachea (1,4). Better assessment of localized tracheal abnormalities can be achieved with contiguous 1.5-5.0-mm-collimation scans obtained through the area during a single breathhold (Fig 1).

• Benign Neoplasms

Benign neoplasms account for less than 10% of tumors involving the trachea and mainstem bronchi and include papillomas (Fig 2), submucosal salivary gland adenomas, and primary mesenchymal tumors such as hamartoFigure 3. Hamartoma of the left main-stem bronchus. (a) CT scan (10-mm collimation) demonstrates a 1.5-cm partially calcified lesion within the left mainstem bronchus (arrows). Areas of decreased attenuation within the mass had CT numbers compatible with those of fat. These findings are suggestive of hamartoma. (b) Bouin-fixed gross pathologic specimen obtained after a left lower lobectomy with sleeve resection of the left main-stem bronchus demonstrates endobronchial tumor filling the lumen with minimal extension into the wall. Scale is in centimeters. (c) Low-power photomicrograph (original magnification, ×25; hematoxylin-eosin stain) demonstrates fat and cartilage within the tumor.





mas (Fig 3). They all tend to be well circumscribed, rounded, smooth, and less than 2 cm in diameter. As in other cases of tracheal narrowing, stridor is the most common presenting symptom (1,5–8).

The CT scan will usually demonstrate the polypoid configuration and intraluminal location of the mass, which is limited by the tracheal cartilage. Hamartomas may be definitively diagnosed at CT if fat can be demonstrated (Fig 3).

• Primary Malignant Neoplasms

Primary malignant neoplasms of the trachea are uncommon, accounting for less than 1% of all thoracic malignancies (1,5,8). The presentation is variable, and the initial early symptoms are nonspecific, including dyspnea, wheezing, stridor, hemoptysis, or dysphagia. As with other causes of tracheal narrowing, the patient may initially be treated for bronchial asthma. The lesions typically are visible on chest radiographs but are rarely identified prospectively.

Bronchoscopy continues to be the mainstay of diagnosis and is highly successful in helping identify the mucosal and intraluminal portions of the tumor. It is difficult, however, to establish the extent of disease with this modality. Conversely, CT enables the precise evaluation of the extraluminal portion of the tumor, as well as the status of bronchi distal to tight stenoses. It is less useful in accurately distinguishing between mucosal and submucosal tumors. CT also tends to result in the underestimation of the longitudinal extent of the tumor. Bronchoscopy combined with CT is therefore complementary (5,9).

The most common primary tracheal malignancies are squamous cell carcinoma (Figs 4, 5) and adenoid cystic carcinoma (Fig 6).



Figure 4. Squamous cell carcinoma of the trachea. Contrast-enhanced CT scans (5-mm collimation) obtained at the level of the carina demonstrate circumferential thickening of the tracheal wall with irregular narrowing of the tracheal lumen.





Figure 5. Squamous cell carcinoma of the trachea. (a) Contrast-enhanced CT scan (10-mm collimation) obtained with the mediastinal settings through the trachea shows no abnormalities. (b) CT scan (10-mm collimation) with the lung settings shows a small, ill-defined region of increased attenuation along the right side of the trachea. (c) High-resolution (1.5-mm collimation, high-spatial-frequency reconstruction algorithm) contrast-enhanced CT scan obtained through the same region clearly demonstrates a small enhanced nodule arising from the right tracheal wall, with no evidence of extraluminal extension. Additional 1.5-mm-collimation CT scans were obtained because a small focal tracheal abnormality was seen at bronchoscopy.

c.



Figure 6. Adenoid cystic carcinoma of the trachea. (a) Contrast-enhanced CT scan (3-mm collimation) obtained through the trachea demonstrates a mass producing irregular thickening of the left wall of the trachea. Note the intraluminal extension of the tumor (arrow). (b) Bouin-fixed gross specimens of two discontinuous tracheal rings show tumor of the left cartilaginous wall with a minor exophytic component and a major infiltrating component. Scale is in centimeters.



Figure 7. Mucoepidermoid carcinoma of the trachea. (a) Contrast-enhanced CT scan (5-mm collimation) demonstrates irregular thickening of the left posterolateral tracheal wall with nodular intraluminal extension of tumor (arrow). No extension beyond the airway is seen. (b) Low-power photomicrograph (original magnification, $\times 2$; periodic acid–Schiff stain with diastase) of a tracheal ring open before fixation shows an exophytic mucoepidermoid tumor primarily involving the membranous trachea.

Less common tumors include mucoepidermoid (Fig 7) and carcinoid (Fig 8) tumors (1,5,8,10). Other tumors are rare (Fig 9). Because of the vascular nature of carcinoid tumors, they may show marked enhancement on CT scans obtained after intravenous administration of contrast material (Fig 7) (9,11).

• Secondary Malignant Neoplasms

The trachea and main-stem bronchi may be involved with malignancies arising elsewhere, either through direct invasion or, less commonly, through hematogenous metastasis to the mucosa. The most common malignancies that locally invade the trachea include cancers of the thyroid, esophagus, larynx, and lung. Hematogenous metastases are rare. The most common sources for hematogenous me-



Figure 8. Carcinoid tumor of the trachea. (a) CT scan (5-mm collimation) obtained through the trachea demonstrates a well-defined, rounded intraluminal mass arising from the left posterolateral wall of the trachea. No obvious extension of tumor beyond the airway is seen. (b) After an injection of contrast material, the mass enhances dramatically with no evidence of extension. (c) Nonfixed gross specimen demonstrates a polypoid erythematous tumor of the left lateral tracheal wall.





b.

July 1992



Figure 9. Giant cell carcinoma of the trachea. Contrast-enhanced CT scan (5-mm collimation) obtained just above the carina demonstrates a polypoid mass extending from the right tracheal wall into the tracheal lumen (arrow). Circumferential thickening of the tracheal wall is present, with no gross evidence of invasion of other mediastinal structures.



Figure 10. Metastatic melanoma of the trachea. Contrast-enhanced CT scan (5-mm collimation) demonstrates a well-defined, rounded intraluminal mass arising from the right posterolateral wall. No tracheal thickening or extraluminal extension is seen.



Figure 11. Tracheobronchomegaly. CT scans (1.5-mm collimation) obtained through the trachea and main-stem bronchi demonstrate diffuse dilatation of the trachea and both main-stem bronchi. The trachea measured 3.3 cm in diameter, and the right and left main-stem bronchi measured 3.0 and 3.4 cm, respectively.

tastases are melanomas (Fig 10), breast carcinomas, and malignancies of the genitourinary tract. On CT scans, the lesions usually appear as a polypoid soft-tissue mass that is generally solitary but may be multiple (1,12).

■ DIFFUSE DISEASE THAT PRO-DUCES AN INCREASED DIAMETER

Tracheobronchomegaly (Mounier-Kuhn disease) is a rare condition characterized by marked dilatation of the trachea and bronchi. The disease affects primarily men in their 4th and 5th decades. The cause is unknown. Clinically, the patient may not have any symptoms or may present with repeated respiratory tract infections and bronchitis. Recurrent infections may lead to bronchiectasis (1,2).

The CT scan demonstrates enlargement of the central airways (Fig 11). In adults, the diagnosis is established by a diameter of the trachea that is greater than 3.0 cm or by a diameter of the right or left main-stem bronchi greater than 2.4 or 2.3 cm, respectively (13). Tracheal diverticulosis may be present, producing an irregular corrugated or scalloped appearance of the trachea (1,14).







c.

Pathologically, the cartilaginous rings are dilated, and the muscular and elastic tissues are atrophied. Because of the underlying weakness of the tracheal wall, portions of the mucosa and submucosa may bulge outward between the tracheal rings, producing the tracheal diverticulosis seen on CT scans (2).

DIFFUSE DISEASES THAT PRODUCE A DECREASED DIAMETER

• Relapsing Polychondritis

Relapsing polychondritis is a systemic disease in which cartilage is affected diffusely by recurrent episodes of inflammation. The pinnal, nasal, laryngeal, and tracheal cartilages are most commonly involved. The findings may also include polyarthritis, inflammation of ocular structures, and arteritis (2). The disease is classified as an autoimmune connective tissue disorder. Patients present clinically **Figure 12.** Relapsing polychondritis. (a) Contrastenhanced CT scan (1.5-mm collimation) obtained through the trachea demonstrates mild circumferential thickening of the tracheal wall (arrow) with slight narrowing of the airway diameter. (b) CT scan obtained at the level of the main-stem bronchi shows more easily the narrowing of the airway diameter (arrows). (Reprinted, with permission, from reference 15.) (c) Low-power photomicrograph (original magnification, $\times 25$; hematoxylin-eosin stain) of tracheal biopsy specimen shows a chronic inflammatory infiltrate destroying the inner (straight arrows) and outer (curved arrows) perichondrium.

with episodic inflammation of the nose, ears, upper airways, and joints. Ear and nose deformities are common. The major airways are involved in greater than 50% of cases, and recurrent pneumonia is the most common cause of death in these patients (2,3).

On CT scans, a fixed narrowing of the tracheobronchial lumen with associated thickening of the wall is noted (Fig 12) (1,15). Gross destruction of the cartilaginous rings progressing to cicatricial fibrosis and stenosis may occur (16). Rarely, the trachea may become flaccid, likely representing a form of tracheomalacia (1). Pathologically, a dense inflammatory exudate surrounds the perichondrium (Fig 12). The mucosa, submucosa, and tracheal glands appear normal. Cartilage loses its basophilic staining properties and lacunar structure. Progressive dissolution and fragmentation and eventual replacement by fibrous tissue may follow (15).



14.

Figures 13, 14. (13) Amyloidosis. Contrast-enhanced CT scan (10-mm collimation) of the main bronchi demonstrates considerable narrowing of the left main-stem bronchus produced by an eccentric nodular mass along its posterolateral wall (arrow). (14) Tracheopathia osteoplastica. Contrast-enhanced CT scan (5-mm collimation) demonstrates irregular thickening of the tracheal cartilages. Multiple calcified nodular tumors extend into the lumen from the lateral walls (arrows). No involvement of the posterior wall is seen.

• Amyloidosis

Amyloidosis is a condition in which a fibrillar protein is deposited in various organ systems. The lungs and central airways may be involved in systemic disease or may be the only organ involved (2). Tracheobronchial involvement in amyloidosis localized to the respiratory tract most commonly takes the form of diffuse or multifocal submucosal infiltrates (1,2,15). Less commonly, there is a single submucosal masslike lesion. The overlying mucosa is usually intact. Calcification or ossification may occur.

On CT scans, the lumen appears narrowed, with a thickened wall (Fig 13). Calcification may be seen. The distribution of lesions may be focal or diffuse (1,2).

Sarcoidosis

The larynx and subglottic trachea are affected in 1%-3% of patients with sarcoidosis. Disease is usually present elsewhere, although rarely the proximal airway may be the initial site (2). Sarcoidosis may produce intrinsic granulomatous lesions of the trachea and main-stem bronchi or may cause extrinsic compression of these airways due to enlarged mediastinal lymph nodes or extensive mediastinal fibrosis. CT can help confirm the presence of extrinsic masses and enable an estimation of their extent. In cases in which the disease involves the tracheal submucosa, thickening of the tracheal wall may be demonstrated (9). Pathologically, the hallmark of sarcoidosis is the presence of noncaseating granulomas.

Wegener Granulomatosis

Wegener granulomatosis is characterized by a vasculitis with granulomatous inflammation primarily involving the lungs, kidneys, and upper and lower respiratory tract. Involvement of the trachea is uncommon and is typically a late manifestation. Rarely, tracheal stenosis may be the initial abnormality (17).

On CT scans, the major airways are narrowed with abnormal soft tissue within the laryngeal cartilages and tracheal rings. Pathologically, mucosal or submucosal granulomatous inflammation and vasculitis are present (9, 17).

Tracheopathia Osteoplastica

Tracheopathia osteoplastica is a rare benign condition involving the trachea and major bronchi with formation of multiple submucosal osteocartilaginous growths within the anterior and lateral walls. The posterior walls are spared, as they contain no cartilage. The cause is unknown (2,18).

On CT scans, the tracheal cartilages are thickened with irregular calcification (Fig 14). Multiple nodules with or without calcification may be seen protruding into the lumen from the anterior and lateral walls. This is considered to be pathognomonic for this condition. Typically, a long segment of the trachea is involved with possible extension to the mainstem bronchi (1,2,18).





d.



b.

a.

Figure 15. Tracheobronchitis associated with ulcerative colitis. (a) CT scan (5-mm collimation) obtained at the level of the trachea demonstrates circumferential thickening of the tracheal wall, with no extension beyond the cartilage (arrow). (b, c) CT scans obtained at the level of the carina show diffuse circumferential thickening of the bronchial walls (arrow) (b), with considerable narrowing of the left bronchial lumen (c). (Reprinted, with permission, from reference 19.) (d) Pathologic specimen (original magnification, $\times 2$; trichrome stain) of the trachea demonstrates circumferential submucosal sclerosing tracheitis and highlights submucosal fibrosis (arrows).

Pathologically, the masses are composed of submucosal islands of hyaline cartilage with areas of lamellar bone and occasional marrow elements. The mucosal surface is intact. A connection to the perichondrium is often evident, suggesting that the lesions arise from native cartilage (2).

Tracheobronchitis Associated with **Ulcerative** Colitis

Extraintestinal manifestations of ulcerative colitis include abnormalities of the airways. Changes range from predominantly large-airway disease with tracheobronchitis or bronchiectasis to small-airway disease with bronchiolitis obliterans. The pattern of disease is similar to that of sclerosing cholangitis, although no association with this abnormality has been noted (19).

On CT scans, the tracheobronchial walls are thickened, producing irregular narrowing (Fig 15). Bronchiectasis may be demonstrated. Pathologically, the airway lumen is narrowed by concentric fibrosis of the submucosa. The

Figure 16. Tracheomalacia. Contrast-enhanced CT scan (5-mm collimation) shows marked narrowing of the trachea adjacent to an aortic aneurysm. No thickening of the tracheal wall is demonstrated. At bronchoscopy, the trachea collapsed on expiration.

mucosa is inflamed and ulcerated. Both the membranous and cartilaginous portions are involved. The cartilaginous plates may be calcified but are not destroyed (19).

• Saber-Sheath Trachea

The sagittal diameter of the trachea is normally slightly greater than the coronal diameter. Occasionally, the coronal diameter is markedly reduced, resulting in a saber-sheath configuration (1,2). It is commonly associated with chronic obstructive pulmonary disease and almost exclusively occurs in male patients. The abnormal configuration of the trachea changes as it moves from the intrathoracic portion to the extrathoracic portion, at which point the coronal diameter abruptly widens and the sagittal diameter narrows. Tracheal ring calcification is common.



• Tracheomalacia and Bronchomalacia

Tracheomalacia and bronchomalacia refer to a weakness of the walls of the trachea and central bronchi, as well as their supporting structures. This leads to increased flaccidity and collapse of the major airways during forced expiration. A primary form of the disease may be found in children in whom the cartilage is congenitally deficient.

Tracheomalacia and bronchomalacia may be secondary to intubation, chronic obstructive airway disease, trauma, recurrent infections, or polychondritis. These patients are older and present clinically with shortness of breath and wheezing. At bronchoscopy, excessive collapsibility of the airways during a voluntary cough is noted (3).

On CT scans, tracheomalacia is considered to be present if the trachea collapses more than 50% on expiration (Fig 16). Use of ultrafast CT facilitates examining the patient during expiration, and cine CT allows a dynamic study that may help identify abnormalities of the trachea caliber, which may be missed on static images (3,20).

• Infectious Disorders

The major airways may be involved in viral, bacterial, or fungal disease processes. In North America, most cases of laryngotracheobronchitis are viral in nature (parainfluenza or respiratory syncytial viruses most commonly). In these cases, subglottic or laryngeal narrowing is common, but radiographically demonstrable tracheal narrowing is unusual. A membranous form of croup exists that may cause proximal tracheal irregularity. CT scans are rarely obtained for the assessment of these disorders (2,15).

SUMMARY

CT is a valuable tool in the investigation of focal and diffuse diseases of the trachea and main-stem bronchi. CT can demonstrate the location and extent of disease; help characterize abnormal tissues; help evaluate the thickness of the tracheal wall; and help determine the extent of extraluminal disease, including the presence of mediastinal extension and lymphadenopathy. CT findings in both focal and diffuse abnormalities correlate well with the pathologic findings.

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