

Pictorial Essay

Diffuse abnormalities of the trachea: computed tomography findings*

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Abstract

The aim of this pictorial essay was to present the main computed tomography findings seen in diffuse diseases of the trachea. The diseases studied included amyloidosis, tracheobronchopathia osteochondroplastica, tracheobronchomegaly, laryngotracheobronchial papillomatosis, lymphoma, neurofibromatosis, relapsing polychondritis, Wegener's granulomatosis, tuberculosis, paracoccidioidomycosis, and tracheobronchomalacia. The most common computed tomography finding was thickening of the walls of the trachea, with or without nodules, parietal calcifications, or involvement of the posterior wall. Although computed tomography allows the detection and characterization of diseases of the central airways, and the correlation with clinical data reduces the diagnostic possibilities, bronchoscopy with biopsy remains the most useful procedure for the diagnosis of diffuse lesions of the trachea.

Keywords: Thoracic diseases; Tracheal diseases; Tomography, X-ray computed; Trachea.

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Tracheal diseases are uncommon and can be classified as focal, when they are located in only one region of the trachea, or as diffuse, when there is involvement of various tracheal segments. The main causes of focal involvement are primary tracheal neoplasms, lesions of traumatic origin, and some infectious diseases. Diffuse involvement, however, is caused by a wide array of conditions, including amyloidosis, tracheobronchopathia osteochondroplastica, relapsing polychondritis, laryngotracheobronchial papillomatosis, tracheobronchomegaly, neurofibromatosis, tuberculosis and other granulomatous infections, such as Wegener's granulomatosis, as well as lymphomas.⁽¹⁻³⁾

Abnormalities of the trachea are rare in daily clinical practice and, although some patients present significant symptoms, abnormalities of the central airways are frequently unapparent or are not seen on chest X-rays. In cases of clinical or radiological suspicion of tracheobronchial alteration, further evaluation using computed tomography is of great importance.⁽³⁾ The aspect/location of the lesions, the presence/location of the calcifications, and the association with abnormalities of the pulmonary parenchyma, as well as the correlation with the clinical data, can allow the differential diagnosis to be made among the diffuse abnormalities of the trachea.

The spiral acquisition makes it possible to perform multiplane two- or three-dimensional reconstructions, which are especially useful for the evaluation of the extent of the disease and for surgical planning. Virtual bronchoscopy does not provide information additional to that provided by images obtained at the axial plane, nor do other three-dimensional reconstructions. However, it is useful for surgeons and pulmonologists who are already familiar with bronchoscopic images. In addition, it is less invasive and can be performed without general anesthesia. Furthermore, it allows the evaluation of the tracheal segments distal to the stenosis.^(2,3) Nevertheless, bronchoscopy with histopathological study remains the method of choice for the definitive diagnosis of diffuse abnormalities of the trachea.⁽³⁾

Tracheobronchial amyloidosis

Amyloidosis is characterized by local or systemic deposition of abnormal amyloid material in extracellular tissues and can involve multiple organs,

such as the heart, the kidneys, and the gastrointestinal tract.⁽³⁻⁵⁾ Primary respiratory amyloidosis presents three characteristic forms: nodular, diffuse parenchymal, and tracheobronchial.⁽³⁻⁶⁾ The most common is the tracheobronchial form. The most common symptoms are cough, dyspnea, hemoptysis, and hoarseness.⁽⁴⁾ The predominant radiographic finding is nodular, irregular narrowing of the trachea. Computed tomography scans can reveal nodules protruding into the tracheal lumen or, more often, diffuse parietal thickening, with multifocal submucosal plaques^(5,7) (Figure 1). In contrast to what is seen in tracheobronchopathia osteochondroplastica, the posterior wall of the trachea can be affected. The thickened wall can present calcifications.^(6,7) Parenchymal abnormalities secondary to obstruction caused by bronchial collapse can be seen, as can recurrent infectious consolidations, bronchiectasis, and obstructive hyperinflation.^(3,7)

Tracheobronchopathia osteochondroplastica

Tracheobronchopathia osteochondroplastica is a benign disease of the tracheobronchial tree, of unknown etiology, characterized by the presence of multiple submucosal osteocartilaginous nodules, which are primarily located in the anterior and lateral walls of the trachea and in the main bronchi. The posterior wall of the trachea is rarely



Figure 1 - Tracheobronchial amyloidosis. Mediastinal window slice showing diffuse nodular thickening of the tracheal wall.

affected.^(1,8-12) Many patients are asymptomatic, and the disease, in such cases, is diagnosed through routine tests.⁽³⁾ The most common symptoms are cough, dyspnea, wheezing, and, occasionally, hemoptysis, which is caused by the friction between nodules.⁽⁹⁾ Patients frequently present recurrent respiratory infections.^(8,9) Although tracheobronchopathia osteochondroplastica is a benign disease, it can evolve to severe tracheal stenosis.^(1,9) Computed tomography is very sensitive in the identification of nodule calcification, in the definition of the extent and distribution of the tracheobronchial stenosis, and in the characterization of complications, such as atelectasis, bronchiectasis, and postobstructive pneumonia.⁽¹⁰⁾ As can be seen in Figure 2, the most common tomographic finding is the presence of multiple submucosal nodules, calcified or not, located in the anterior and lateral walls of the trachea.^(1,3,9-12)

Tracheobronchomegaly

Tracheobronchomegaly, also known as Mounier-Kuhn syndrome, is characterized by marked dilatation of the trachea and main bronchi due to atrophy or to the absence of the longitudinal elastic fibers and the smooth muscles that form its walls.^(13,14) The main abnormality observed in the respiratory physiology of such patients is the total collapse of the

airways during expiration.^(2,14) With the impairment of the cough reflex and of the mucociliary defense mechanism, the airways are extremely widened and weakened, which causes mucus accumulation, recurrent pneumonia, and fibrosis.^(2,13,15) Therefore, the symptoms are less than specific and are related to recurrent infections, including excessive expectoration, hemoptysis, and dyspnea.⁽²⁾ Computed tomography scans show an increase in the diameter of the upper airways (Figure 3). A decrease in the thickness of the tracheal wall, together with bronchiectasis and diverticula, which usually originate from the posterior lateral wall, can also be observed.^(2,13-15)

Laryngotracheobronchial papillomatosis

Laryngotracheobronchial papillomatosis is an infection of the airways, caused by the human papillomavirus, and is capable of presenting malignant degeneration.^(2,3) The larynx is usually affected, whereas tracheal involvement is rare, occurring in only approximately 50% of patients with laryngeal papillomatosis.^(2,3,16) Papillomas can be single or multiple.⁽³⁾ The most common initial symptom is hoarseness caused by the impairment of the true vocal cords. With the dissemination of the disease, the patient can present varying degrees of airway obstruction, with wheezing, atelectasis, recurrent pneumonia, and bronchiectasis. Hemoptysis occurs frequently, and the infection is commonly confused with active tuberculosis.⁽¹⁶⁾ The radiographic findings include polypoid lesions, either sessile or pedunculated, located in the trachea and in the main bronchi. When there is distal involvement of the airways, computed tomography scans show nodules with a centrilobular distribution, often cavitated, predominantly located in the basal and posterior halves of the lungs (Figure 4). When other infections are superimposed, air-fluid levels within the cavities, areas of consolidations, and atelectasis can be seen.^(2,3,16,17)

Tracheal lymphoma

Lymphomas are primary neoplasms of the lymphoreticular system and are classified as Hodgkin's disease or non-Hodgkin's lymphoma. The primary tracheal form is extremely rare, and, when it occurs, it seems to be related to the mucosa-associated lymphoid tissue.⁽¹⁸⁾ The most common symptoms

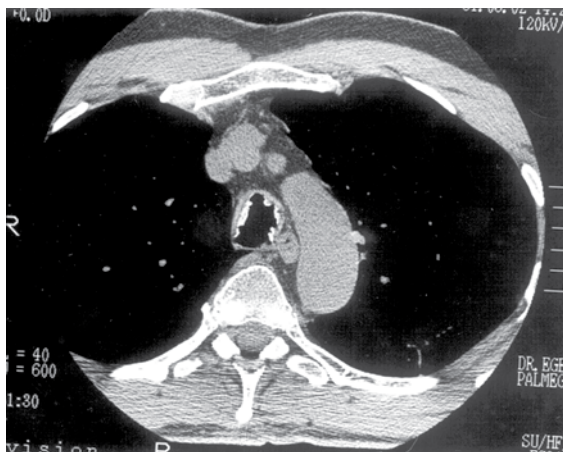


Figure 2 - Tracheobronchopathia osteochondroplastica. Mediastinal window slice, obtained through spiral acquisition, revealing calcified plaques, some with a nodular aspect, distributed throughout the cartilage rings, sparing the posterior portion of the trachea.

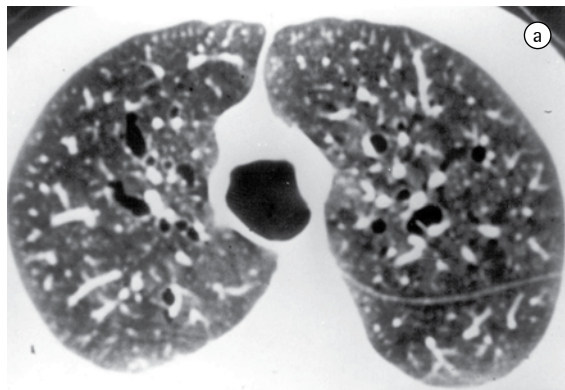


Figure 3 - a) Tracheobronchomegaly. Pulmonary parenchyma window slice showing irregular and diffuse dilation of the trachea. Note also the presence of bronchiectasis; and b) Tracheobronchomegaly. Three-dimensional reconstruction revealing the marked dilatation of the trachea and of the main bronchi.

of airway lymphoma are related to obstruction, and they often delay the initiation of treatment due to the diagnostic confusion with bronchial asthma.⁽¹⁸⁾ Among such symptoms are dyspnea, nonproductive cough, and wheezing. Hemoptysis can also occur, although it is not common, since, in most cases, the bronchial mucosa is intact.^(18,19) Computed tomography scans, such as the one displayed in Figure 5, can reveal focal tracheal narrowing and a solitary mass or polypoid thickening of the tracheobronchial

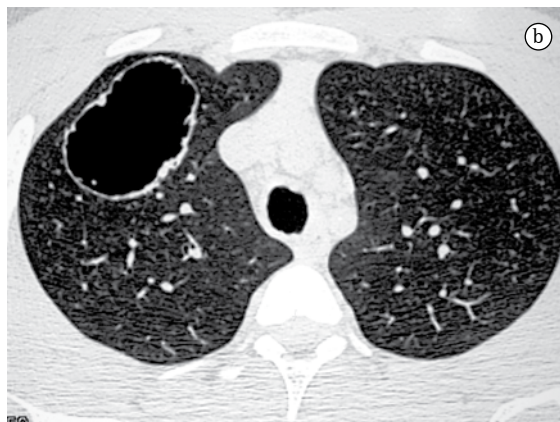


Figure 4 - a) Tracheobronchial papillomatosis. Mediastinal window slice revealing irregular thickening of the tracheal wall, with nodules and plaques protruding into the lumen; and b) Tracheobronchial papillomatosis. Pulmonary parenchyma window slice, at the level of the upper lobes, showing irregularities throughout the tracheal lumen, in addition to a thin-walled cavity in the upper lobe of the right lung.

walls due to the diffuse infiltration of the submucosa.^(18,19)

Tracheal neurofibromatosis

Neurofibromatosis is the most common phacomatosis and presents thoracic manifestations in 10 to

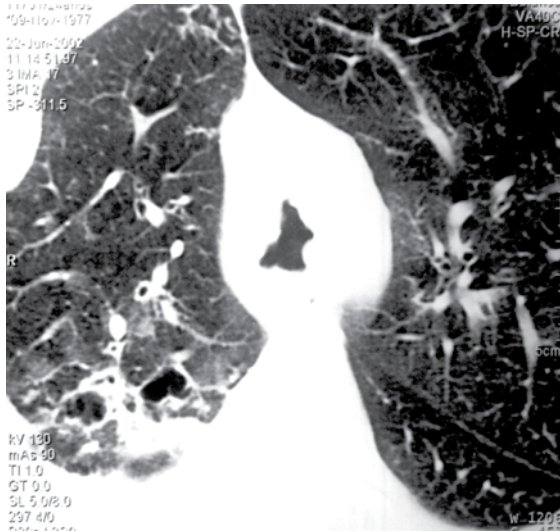


Figure 5 – Lymphoma. Computed tomography scan revealing nodulation and plaques in the tracheal lumen, which cause an irregular reduction in its caliber. Note also bronchiectasis and areas of parenchymal consolidations caused by an accompanying infectious process.



Figure 6 – Neurofibromatosis. Three-dimensional (volume rendering) reconstruction of the trachea and main bronchi in which one can identify, by transparency, three nodular polypoid images, with regular borders, located in the carina and in each of the main bronchi.

20% of the cases. As can be seen in Figure 6, upper airway involvement is revealed by the presence of intraluminal or extraluminal nodular lesions.⁽²⁰⁻²²⁾ The typical clinical profile consists of numerous circumscribed areas of skin hyperpigmentation accompanied by dermal and neural tumors of various types. Additional diagnostic criteria include freckles in the axillary region, iris hamartomas, bone dysplasia, multiple tumors of the central nervous system, such as optic nerve gliomas, and having a first-degree relative affected by the disease.⁽²¹⁾ Upper airway obstruction occurs due to the presence of intraluminal or extraluminal lesions that cause extrinsic compression of the tracheobronchial tree.⁽²⁰⁻²²⁾

Relapsing polychondritis

Relapsing polychondritis is a rare systemic disease characterized by recurrent episodes of cartilage inflammation involving the cartilage of the nose, ear, peripheral articulations, larynx, and tracheobronchial tree. In the early phase of the disease, laryngotracheobronchial involvement is present in approximately 10% of cases and indicates bad prognosis. Over the course of the disease, approximately half of all patients with polychondritis

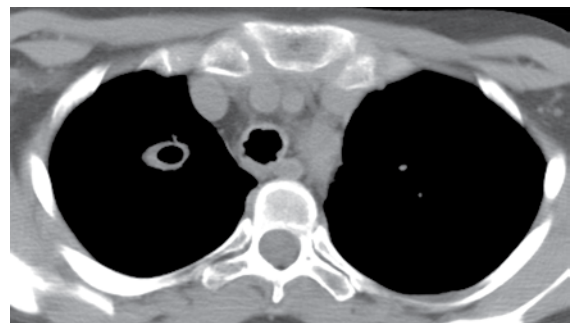


Figure 7 – Wegener's granulomatosis. Mediastinal window slice, in the supra-aortic region, showing irregularities and nodulation in the tracheal lumen, affecting its entire circumference, with thickening of the wall. Note the cavitated lesion in the right upper lobe.

present airway involvement, and the larynx and the upper portion of the trachea are the most common sites. The clinical criteria for the diagnosis include the presence of three or more of the following abnormalities: bilateral auricular chondritis, seronegative inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, and auditory-vestibular disorder. Confirmation

is made by immunohistochemistry and histological analysis of the cartilage samples analyzed. Computed tomography findings include thickening of and calcifications in the tracheal wall, which can evolve to fibrotic scarring and destruction of the cartilage rings, causing deformity and narrowing of the trachea and bronchi. The posterior wall of the trachea is usually spared. The images obtained during expiration can reveal airway collapse.^(3,23)

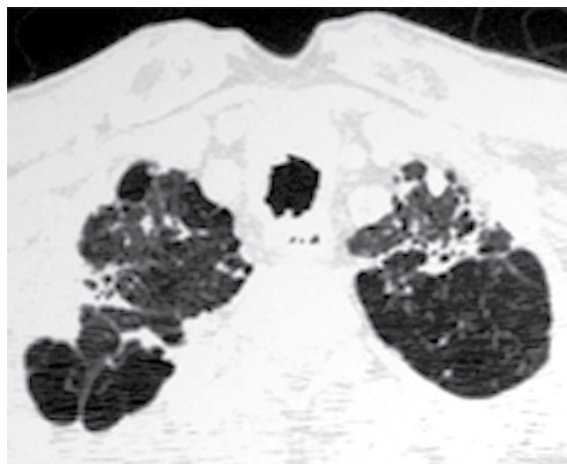


Figure 8 – Paracoccidioidomycosis. Pulmonary parenchyma window slice showing irregularities in the tracheal wall, in addition to consolidations and fibrotic lesions in the upper lobes, with emphysematous bullae and probable bronchiectasis.

Wegener's granulomatosis

Wegener's granulomatosis is a systemic autoimmune disease that primarily affects the kidneys, the upper airways, and the lower airways. It occurs more frequently in males and in middle-aged individuals. The most common symptoms are cough, hemoptysis, chest pain, and dyspnea. The principal histopathological abnormality is necrotizing granulomatous vasculitis of the small blood vessels.^(3,24,25) The principal computed tomography findings include subglottic stenosis, and circumferential mucosal thickening, as well as irregularity and ulceration of the tracheobronchial walls. Although involvement of the cartilage rings is less common, it can also result in deformity and narrowing of the trachea. As shown in Figure 7, Wegener's granulomatosis is often accompanied by pulmonary nodules, which can be cavitated, and by alveolar infiltration.^(3,24,25)

Tracheal tuberculosis and other granulomatous infections

The main finding in central airway tuberculosis is the narrowing of the lumen. Tuberculosis involving the central airways occasionally results in diffuse stenosis, which can lead to respiratory failure in the acute phase. On computed tomography scans, active tracheobronchial tuberculosis presents as irregular circumferential luminal narrowing and can be accompanied by mediastinitis. In the fibrotic

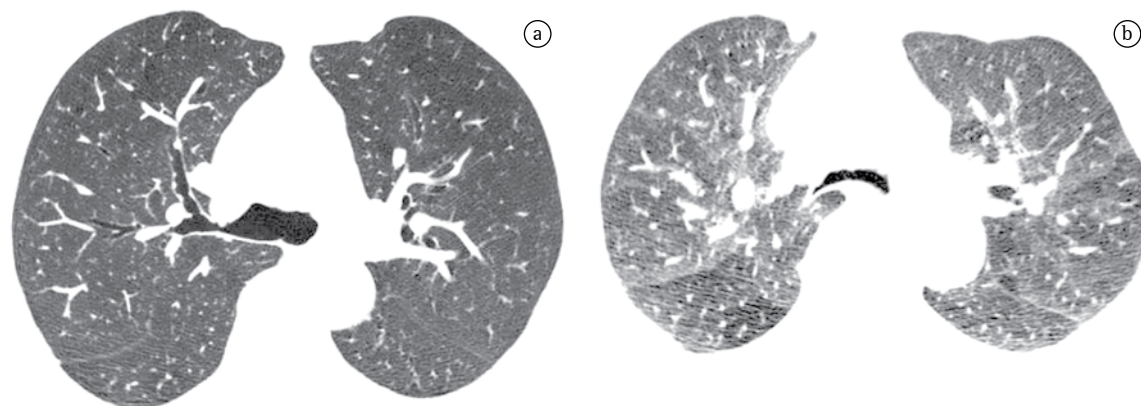


Figure 9 – a) Tracheobronchomalacia. Slice obtained during the inspiratory phase, at the level of the bronchial bifurcation, showing the main bronchi to be of normal caliber; and b) Tracheobronchomalacia. Slice at the level of the bronchial bifurcation, during the expiratory phase, in which marked collapse of the main bronchi can be seen.

form of the disease, the lumen is smoother, and the wall is not thickened. Lymph node enlargement is usually associated with the active form of the disease.⁽²⁶⁾ The disease can affect a large portion of the trachea, also affecting the bronchi,⁽²⁶⁾ or can involve only a small segment of the trachea or of one bronchus.^(2,27) Other granulomatous infections, such as histoplasmosis and paracoccidioidomycosis (Figure 8), can cause similar lesions.^(2,28)

Tracheobronchomalacia

Tracheobronchomalacia is characterized by flaccidity of the walls of the airways, which causes excessive collapse of the trachea and bronchi during expiration. The disease can be congenital or can be acquired secondary to a series of factors, such as selective intubation, trauma, infection, chronic extrinsic compression, and chronic obstructive pulmonary disease.^(29,30) Clinically, patients can present dry cough, dyspnea, and recurrent infections. The result of the tomographic examination obtained during the inspiratory phase is normal. As can be seen in Figure 9, the tomographic diagnosis is usually made based on slices obtained during expiration, or, in dynamic studies, when the inner area of the tracheal or bronchial lumen, on axial slices, suffers a greater than 50% reduction in comparison with the inspiratory phase.^(29,30)

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