Lesions of the tracheobronchial wall: radiological pearls

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Learning objectives

The purpose of this work is to describe the radiological features of the main non-neoplastic conditions of the tracheobronchial wall and their correlation with bronchoscopy images.
Background

Non-neoplastic lesions of the tracheobronchial wall are rare. However, they constitute a group of heterogeneous diseases that can cause severe respiratory symptoms and, sometimes, be associated with systemic disorders, which can have a significant impact on the quality of life of the patient. Therefore, it is of paramount importance for the general radiologist to be familiar with the imaging characteristics of these abnormalities.

In order to better understand these radiological features, it is important to remember some anatomical characteristics of the tracheobronchial wall:

- The trachea is composed of 16-22 incomplete cartilaginous rings disposed in axial plane, showing discontinuity of posterior part. The anterior side is composed of different layers of mucosa, submucosa, cartilage, muscle and adventitia, whereas the cartilaginous rings are closed on the posterior side by the thin trachealis muscle. For this reason, conditions affecting cartilage (such as recurrent polychondritis) will show radiological features different from other conditions such as vasculitis or granulomatous diseases.
- The diameter of the normal trachea varies according to gender. It ranges from 13-25 mm in coronal diameter and 13-27 mm in sagittal diameter in male; and from 10-21 mm in coronal diameter and 10-23 mm in sagittal diameter in female.

The protocol we use to study tracheobronchial wall conditions consists in end-inspiratory high-resolution computed tomographic (HRCT) and end-expiratory HRCT, using multiplanar reconstructions for a further understanding of the images.
Findings and procedure details

The non-neoplastic lesions of the tracheobronchial wall have different aetiologies. We will review the main radiological characteristics of the following abnormalities: relapsing polychondritis, granulomatosis with polyangiitis, osteochondroplastic tracheobronchopathy, tracheobronchial amyloidosis, papillomatosis, post-intubation stenosis, idiopathic stenosis, Mounier-Kuhn syndrome and bronchial anthracofibrosis.

1. Relapsing polychondritis

Rare auto-immune syndrome characterised by the destruction of the cartilage, especially from the auricle (90%), nose (50%), tracheobronchial wall (50%) and peripheral joints. The laryngotraceobronchial presentation is seen in 10% of the cases, but in approximately 50% of these, the disease will eventually progress affecting the tracheobronchial wall. When it affects the respiratory, it is associated with a poorer prognosis, being pneumonia the first cause of death in approximately 30% of patients.

It is more prevalent in Caucasian at the 40-60 years old. Despite it affects equally men and women, the tracheobronchial involment is more frequent in women.

It is characterized by recurrent episodes of cartilage inflammation with subsequent degeneration, loss of structure and fibrosis. It usually affects the larynx and the upper trachea, although it can progress to the subsegmentary bronchi.

It can be diagnosed when three or more of the following are observed: bilateral auricular chondritis, seronegative inflammatory polyarthritis, nasal chondritis, ocular inflammation, chondritis of the respiratory tract, and audio-vestibular lesions.

Conventional chest radiograph might show diffuse or focal airway affectation. However, CT scan is the main tool (Fig. 1 on page 10), which may present:

- Increased airway wall attenuation (calcium) and smooth thickening of the anterolateral tracheal wall with loss of structure of the cartilaginous rings and sparing of the posterior wall.
- Focal stenosis due to cartilage destruction and fibrosis.
- Collapse in the end-expiratory HRCT scan.

As for the bronchoscopy images, diffuse airway narrowing, mucosal oedema and erythema may be appreciated.
2. Granulomatosis with polyangiitis

Granulomatosis with polyangiitis, previously known as Wegener granulomatosis, is a systemic necrotising non-caseating granulomatous vasculitis with unknown pathogenesis.

It usually occurs in middle-aged men.

Although it can affect any system and organ, it has predilection for respiratory system and kidneys. There is also a form of the disease affecting the lung and the lower airway, without altering the upper airway or the kidneys. In the classic granulomatosis, larynx and endobronchial implication are usually late complications.

Conventional chest radiograph might present diffuse or focal airway affectation, and the HRCT scans show (Fig. 2 on page 10):

- Circumferential and irregular thickening of the mucosa with ulceration.
- Rare affectation of the tracheal rings, which can lead to stenosis. If the peripheral bronchi wall is affected, it can cause obstruction and atelectasis.
- Pulmonary nodular areas of increased attenuation that can cavitate.
- Ground-glass opacities.
- Pleural effusion is a rare presentation.

Findings at bronchoscopy are very variable. Inflammatory ulcerations are the most frequent, yellow-green plaques are related to active illness and, therefore, are the site of greater profitability on a biopsy; subglottic stenosis, granulomatous inflammatory pseudotumors, purulent secretions or lobar haemorrhage would be other presentations.

3. Osteochondroplastic tracheobronchopathy

Rare idiopathic benign disease that affects the lower 2/3 of the trachea and major bronchi. It is characterised by multiple submucosal osteocartilaginous nodules, which can be focal or diffuse. Men around 50 years old are more likely to suffer from osteochondroplastic tracheobronchopathy than women, with a 3:1 ratio.

It is believed that this disease has a higher prevalence than estimated due to its asymptomatic course. It is usually diagnosed after a difficult intubation or during bronchoscopy. Cough, shortness of breath, recurrent infections, wheeze and, sometimes, haemoptysis can be present.
Conventional chest radiograph shows nodular irregularity or asymmetric irregular stenosis in moderate-severe disease. The HCRT scan shows (Fig. 3 on page 11):

- Thickening tracheal cartilage with irregular calcified nodules, which spare the posterior tracheal wall, as a pathognomonic feature.
- Pneumonia or atelectasis as possible complications.

Bronchoscopy shows multiple smooth, white, cartilaginous and hard nodules distributed in the anterolateral tracheal wall.

4. Tracheobronchial amyloidosis

Disease characterized by abnormal protein deposits (amyloid) in the extracellular tissue, it can be of idiopathic, hereditary, or inflammatory/neoplastic related aetiology. Tracheobronchial amyloidosis can be found in two different scenarios: (i) as a part of a systemic process or, (ii) rarely, involving the airway and pulmonary parenchyma exclusively. This latter form is usually asymptomatic, and can be found in three forms: diffuse interstitial lung deposits, single or multiple nodules, or submucosal tracheobronchial deposits.

Conventional chest radiograph might show a nodular appearance with irregular narrowing of the tracheal lumen. If there is endobronchial occlusion, lobar atelectasis can be seen.

HCRT scan shows (Fig. 4 on page 12):

- Diffuse, nodular and concentric thickening of the trachea and main bronchi wall.
- Sometimes nodular calcified lesions can be seen. In this case, differential diagnosis include osteochondroplastic tracheobronchopathy and relapsing polychondritis. Whereas tracheobronchial amyloidosis has a concentric involvement of the lumen, osteochondroplastic tracheobronchopathy and relapsing polychondritis spare the posterior tracheal wall.

Bronchoscopy shows white-grey flat plaques of amyloid deposit in the tracheal and main bronchi wall.

5. Laryngotracheobronchial papillomatosis

Infection of the upper respiratory tract by the human papillomavirus (HPV). It is more frequent in children, after vaginal delivery infection. Nonetheless, it can also occur in adults, probably due to oral contact with infected genitals. Papillomatous lesions are usually
found in the larynx, but the viral infection can be spread all through the tracheal and bronchial lumen, and even reach the lung parenchyma.

Radiological features are better viewed in the HRCT scan (Fig. 5 on page 13), with:

- Diffuse nodular thickening of the airway wall, or multiple nodules projected to the airway lumen.
- When it reaches the lung, multiple and bilateral cysts and cavitated nodules can be seen.
- Atelectasis, air trapping, infections or bronchiectasis can also be seen when airway obstruction happens.

Bronchoscopy shows white polypoid appearance in the larynx, trachea or main bronchi wall.

6. Post-intubation stenosis

Among others, post-intubation stenosis is caused by the high pressure of the endotracheal tube balloon against the tracheal wall, with mucosal necrosis and secondary scar and stenosis outcomes. It occurs in the subglottic region, typically 3-4 cm below the cricoid cartilage, whereas stenosis in patients with history of tracheostomy occurs at the stoma site. Symptoms are variable and can include shortness of breath or stridor.

Conventional chest radiograph and HRCT scan show a symmetric, focal subglottic stenosis with an hourglass shape (Fig. 6 on page 14).

7. Idiopathic laryngotracheal stenosis

Idiopathic stenosis consists in the narrowing of the upper airway due to an unknown cause, although gastroesophageal reflux or autoimmune diseases have been suggested as aetiology.

It is a rare and slowly progressive disease more prevalent in women than men. It usually affects the subglottic region, but it can also involve the upper trachea.

Symptoms are variable, similar to those present in post-intubation stenosis, including dysphonia or life-threatening airway obstruction, with a long duration of 2 years on average.
Radiological features are heterogeneous, although the most frequent appearance is a smoothed hourglass shaped stenosis (Fig. 7 on page 15).

8. Mounier-Kuhn syndrome

Mounier-Kuhn syndrome or tracheobronchomegaly is characterised by the important and diffuse dilatation of the trachea and the main bronchi due to atrophy of smooth muscle and elastic tissue, especially in middle aged men. This disease has an unknown cause, although a congenital predisposition has been hypothesised. This condition can be an accidental finding in a chest X-ray of an asymptomatic patient undergoing imaging for other indications, but the most frequent symptom is recurrent airway infection.

HRCT scan shows an increased diameter, more than 3 cm, of the trachea and more than 2.4 cm in main bronchi. Further, airway dilatation with multiple diverticula projected between the cartilaginous rings, and bronchiectasia. (Fig. 8 on page 16).

9. Bronchial anthracofibrosis

Bronchial anthracofibrosis is characterised by bronchial lumen narrowing associated with anthracotic pigmentation without a significant exposure to smoking or pneumoconiosis. It is described mostly in women in their sixties and it can be associated with tuberculosis in around 50% of the cases, as well as with chronic exposure to biomass smoke. Symptoms are very variable, from chronic productive cough and dyspnoea, to chest pain or obstructive pattern in pulmonary functional tests.

Conventional chest radiograph is not specific and does not show luminal stenosis, but can show atelectasis, and a reticular or reticulonodular pattern, and sometimes a mass lesion.

HRCT scan shows (Fig. 9 on page 17):

- Mediastinal or hilar lymphadenopathies, with calcified nodes adjacent to bronchi.
- Multifocal thickening of the bronchial wall causing smooth narrowing, with a predilection for the upper and middle right lobe bronchi.
- Collapse in distal lung portions to the site of airway stenosis.
- Fibrotic bands.
- Nodules.
- Bronchiectasis.

Bronchoscopy shows a darkened bronchial mucosa.
**Fig. 1:** Relapsing polychondritis. Chest X-ray (a) in a 59-year-old man shows diffuse narrowing of the tracheal lumen. Sagittal (b) and coronal (c) CT images show the anterolateral tracheal and main bronchi wall thickening, sparing the posterior wall, and conditioning collapse of the tracheal lumen in the end-expiratory image (e). A small calcification can also be seen in the sagittal image. Bronchoscopy image (f) shows loss of structure of the tracheal wall and reveals the presence of whitish deposits.

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**Fig. 2:** Granulomatosis with polyangiitis. CT images of a 51-year-old woman with granulomatosis with polyangiitis. It shows cavitated masses in the right lung (a, d) and circumferential soft-tissue thickening and narrowing of the main right bronchi and intermediate bronchi (b). Segmentary and subsegmentary bronchi of the upper right lobe are also affected, as it can be seen in the axial CT image and in the 9,1 mm minIP coronal image.

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**Fig. 3:** Osteochondroplastic tracheobronchopathy. Chest X-ray (a) in a 48-year-old man shows stenosis of the trachea and left main bronchus, and partial collapse of the middle right lobe. CT images (b, c) show multiple calcified nodules in the anterolateral tracheal and main bronchi wall. Virtual bronchoscopy (d) confirm the irregular narrowing of the tracheal lumen and conventional bronchoscopy (e) reveals the presence of multiple white nodules.

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Fig. 4: Tracheobronchial amyloidosis. CT scan in a 48-year-old woman (a, b, c) shows a concentric thickening of the trachea wall, more evident in bronchi, with calcifications. Bronchoscopy image (c) reveals the presence of multiple flat deposits that cause narrowing of the airway lumen.

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**Fig. 5:** Laryngotracheobronchial papillomatosis. Conventional radiograph (a) in a 20-year-old patient shows bilateral pulmonary involvement with multiple cavity masses. The CT scan reveals polypoidal lesions in the interior of pulmonary cavitations (b, d, e), as well as a circumferential thickening of the tracheal wall (c) with a papilloma in the extrathoracic trachea.

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Fig. 6: Post-tracheostomy stenosis. Focal stenosis of the upper trachea was shown in CT scan scout and 37.4 mm minIP coronal reconstruction (a, b) of a 63-year-old woman with a history of orotracheal intubation and tracheostomy 2 years before. Axial CT image (d) shows the tracheal stenosis at the tracheostomy level. Bronchoscopy image (c) confirms the radiological finding.

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Fig. 7: Idiopathic laryngotracheal stenosis. CT scan (a) in a 69-year-old woman shows a circumferential thickening of the extrathoracic trachea. Volume rendering reconstructions (e, f) reveal focal stenosis of the upper trachea. Images of virtual (b) and conventional bronchoscopy (d) confirm the tracheal stenosis.

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Fig. 8: Mounier-Kuhn syndrome. Conventional radiograph (a, b) of a 54-year-old patient in study of recurrent pneumonia shows opacification of the right middle lobe and increased calibre of the trachea and left main bronchus with irregularity of the posterior tracheal stripe. The images of CT scan (c, d, e) show multiple tracheal diverticula throughout the intrathoracic trachea and large bronchi with extensive varicose bronchiectasis.

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Fig. 9: Bronchial anthracofibrosis. Conventional radiography (a) in a 60-year-old male shows opacification of the left lower lobe. Smooth stenosis and diffuse bronchial wall thickening in the CT images (b, d), with enlarged and partially calcified lymph nodes. Bronchoscopy image (c) reveals bronchial stenosis with an anthracofibrosis stain in bronchial bifurcation.

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Conclusion

The morphological characteristics of the tracheobronchial abnormalities (stenosis or enlargement of the trachea, smooth or nodular wall thickening, calcification, lung parenchymal involvement...) and their location (anterior or posterior tracheal wall, upper and/or lower respiratory tract, focal or diffuse) are important features for an adequate differential diagnosis. For example, sparing of the posterior tracheal wall narrows the diagnosis into two possible conditions: relapsing polychondritis or osteochondroplastic tracheobronchopathy. Other tools such as multiplanar, volume rendering or virtual bronchoscopic reconstructions of the trachea might have an important role in the evaluation of more complex cases, and combined, can help get a better diagnosis.