Pathologies of the Large Airways: tumors and tumorslike

Poster No.: P-0094
Congress: ESTI 2019
Type: Educational Poster
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Keywords: Education, Image manipulation / Reconstruction, CT-High Resolution, Thorax, Respiratory system, Lung, Chronic obstructive airways disease

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Learning objectives

To describe the imaging findings of tumors involving the tracheobronchial tree and the main differential diagnoses.
Background

Tumors of trachea and bronchi are uncommon. Although tracheobronchial tumors represent only 0.6% of all pulmonary tumors, they are clinically significant. Chest radiography is often the first test performed in patients with suspected large airway disease, but CT is the modality of choice in suspected large-airway diseases. Chest CT allow multiplanar reconstruction images that are mandatory to assess the craniocaudal extent of disease and to plan bronchoscopic or surgical therapies. Large airway diseases may be congenital or acquired lesions which can be further subdivided into focal and diffuse disease, benign and neoplastic. Primary malignant tumors commonly originate from the surface epithelium or the salivary glands, whereas most benign tumors arise from the mesenchymal tissue; the predominant types of malignant are squamous cell. Secondary malignant tumors occur as a result of either hematogenous metastasis or direct invasion by a malignancy from an adjacent structure. Benign tumors of the tracheobronchial tree are quite rare and constitute 2% of all lung tumors. Histologically, the most frequent benign tumors are hamartomas and papillomas. The acquired lesions can occur in a wide number of conditions such as foreign body, bronchocele, tracheobronchomalacia, post-intubation and post-infectious stenosis, systemic diseases (sarcoidosis, Behcet), Wegener Granulomatosis, papillomatosis.
Imaging findings OR procedure details

ACQUIRED FOCAL LESIONS

*Squamous Cell Carcinoma*

Primary neoplasms of the central airways are rare and make up approximately 5% of all lung malignancies. The majority are squamous cell carcinomas. Squamous cell carcinoma is highly associated with cigarette smoking and it is more common in men. It is commonly asymptomatic; when symptomatic, the tumor often occludes more than 50% of the airway diameter. Presenting symptoms of most large-airway tumors are caused by the mass itself and include cough, hoarseness, dyspnea, hemoptysis, and wheeze. Hemoptysis is more common with SCC because of erosion or ulceration of the respiratory epithelium. CT demonstrates a small sessile or polypoid lesion in the lower third portion of the trachea. Because squamous cell carcinoma arises from the surface epithelium, the mucosal surface is typically irregular. It may invade the mediastinum by direct extension or lymph node metastasis and uncommonly spreads hematogenously.

*Adenoid Cystic Carcinoma*

Adenoid cystic carcinoma is the second most common tracheal tumor and is the most common salivary-type tumor of the large airways. As with other focal tracheal tumors, the most common symptoms are caused by mass effect and include cough, hoarseness, dyspnea, and wheeze, hemoptysis can also occur. Because of its submucosal origin, it tends to have an intact mucosa and a smooth contour, often with circumferential or longitudinal submucosal extension.

On CT, adenoid cystic carcinoma appears as a smooth, focal mass in the trachea or main bronchi (*Fig. 1*); it may involve more than half of the airway circumference. The longitudinal extent is typically greater than the cross-sectional area, and it may even involve the entire trachea and extend into the main bronchi. Lymphadenopathy and distant metastasis are uncommon; most recurrence is local.

*Endobronchial carcinoid*

Bronchial carcinoids account for less than 5% of all primary lung tumors in adults and represent 25% of all carcinoids, but are the most common primary pulmonary neoplasms in the pediatric population. Typical carcinoids are low-grade neuroendocrine tumors that have an excellent prognosis. They usually present with symptoms of airway obstruction.

Radiologic findings depend on tumor size and location; CT illustrates a well-defined rounded nodule with a slightly lobulated border. Carcinoid tumors usually show avid contrast enhancement due to their high vascularity (*Fig 2B-C*). Approximately 80% of
bronchial carcinoids are located in the main, lobar or segmental bronchi. The other 20% present as peripheral lung nodules. Depending on size, carcinoids can present as endobronchial nodules to large perihilar masses. Eccentric calcifications can be seen within the tumor in addition to hilar and mediastinal lymphadenopathy.

While imaging cannot differentiate between typical and atypical bronchial carcinoids, atypical carcinoids generally are most frequent in older patients and much more aggressive behavior.

Adenoma

Pedunculated adenomas of the tracheobronchial tree arise from bronchial mucous glands. CT demonstrates a solitary, polypoid mass.

Endobronchial lipoma

Among benign lesions, endobronchial lipoma is a very rare disease entity, accounting for only 0.1%-0.4% of all bronchial tumors. Endobronchial lipomas arise from the adipose tissue in the submucosal layer of the bronchial wall. CT examination shows fat-density (HU of -40 to -120) lesion smooth margins located in the bronchus; it can be associated with lung collapse (Fig. 4) or distal airway dilatation, occasionally with infiltration or consolidation.

Foreign body

Foreign body aspiration is more common in children than adults. In adults, the foreign body is usually aspirated food. Chest radiographs often show normal findings (Fig. 7A), and in children, up to 80% of tracheal foreign body are radiographically imperceptible. Aspirated foreign bodies most commonly are lodged in the right main bronchus and lower lobe bronchus. CT is useful in stable patients if the diagnosis is unclear to identify and localize foreign bodies to guide bronchoscopy (Fig. 7B).

ACQUIRED DIFFUSE LESIONS

Tracheobronchomalacia

Tracheobronchomalacia is defined as the loss of cartilaginous support of the trachea or major bronchi, leading to complete or near-complete collapse of the airway lumen during respiration. It may be congenital or secondary to trauma, infection, chronic external compression or chronic inflammation. Expiratory CT imaging (Fig. 8) is required for diagnosis with a 50% expiratory reduction in the cross-sectional luminal area of the
airway, this cut-off is, widely considered diagnostic although recent studies have shown that this may occur in healthy volunteers and a 70% cut off may be more appropriate.

**Post intubation stenosis**

Post intubation stenosis can occur as a result of endotracheal intubation or tracheostomy tube placement; the site of stenosis is usually at the cuff in the subglottic region in endotracheal cases or at the site of the stoma post tracheostomy.

The cuff-pressure of endotracheal tubes play an important role on the development of tracheal damage: at a cuff pressure of >30 mmHg, there is an increase in mucosal capillary perfusion pressure, which leads to mucosal ischemia and consequent inflammation of the tracheal cartilages. These pathological changes may eventually lead to fibrosis of circumferential lesions, resulting in progressive tracheal stenosis. At CT the typical features are a soft tissue thickening (eccentric or concentric) with luminal stenosis on axial images (*Fig. 9*) and a short hourglass shape narrowing on coronal reconstructions (*Fig. 10*). The outer tracheal wall has a normal appearance without evidence of deformity of narrowing. Expiratory CT shows little change in tracheal diameter.

**Post-infectious stenosis**

The major airways may be involved in viral, bacterial, or fungal infections. Most bronchitis are viral; parainfluenza or respiratory syncytial viruses are the most common causes. Among infectious causes there is *Mycobacterium tuberculosis* (*Fig. 11*). Tuberculosis related stenosis has been described in up to 90% of patients with pulmonary tuberculosis, despite appropriate therapy. At CT, postinfectious airway strictures are characteristically multifocal usually with normal airway between strictures and may be complicated by external lymph node compression. Other specific pathogens that can affect the large airways include *Coccidioides immitis, Histoplasma capsulatum, Aspergillus* species, mucormycosis, and *Klebsiella rhinoscleromatis*.

**Papillomatosis**

Respiratory papillomatosis results from infection of the upper respiratory tract by the HPV. The infection usually occurs during birth, as the infant passes through the infected birth canal. Patients usually present with voice changes or stridor, other symptoms include cough, recurrent pneumonia, dyspnea, and acute respiratory distress. Papillomatosis predominately affects the larynx but may spread into the trachea, bronchi and even lung. The virtual bronchoscopy can show the endoluminal nodules and map the complete extent of the disease without the risk of spreading the virus downstream, as may occur with bronchoscopy. A rare late complication of respiratory papillomatosis is malignant transformation of a papilloma to SCC (HPV11 subtype). Radiographic findings include
nodular narrowing of the airway that may be either focal or diffuse. Nodules arise from the mucosal surface, and their intraluminal extent is better evaluated with CT. Airway obstruction may lead to atelectasis, air trapping, postobstructive infection, or bronchiectasis. Distal spread may lead to parenchymal nodules that usually cavitate.

**Granulomatosis with polyangiitis (Wegener Granulomatosis)**

Granulomatosis with polyangiitis is a uncommon multisystem necrotizing non-caseating granulomatous c-ANCA positive vasculitis affecting small to medium sized arteries, capillaries and veins, with a predilection for the respiratory system and kidneys. The tracheobronchial tree is the second most commonly affected area in the thorax and is due to mucosal inflammation. Tracheal involvement (present in 16%-23% of cases) can be segmental, unifocal, or multifocal but is usually focal, involving a 2-4-cm span of the trachea. The subglottic region is most commonly affected. CT images showing circumferential (smooth or nodular) tracheal wall thickening and luminal narrowing. Involvement of the posterior membrane of the trachea is the rule, thereby helping distinguish Wegener granulomatosis from other entities such as relapsing polychondritis and tracheobronchopathia osteochondroplastica, both of which characteristically spare this area. Stenosis may be seen in up to 18% of patients with airway involvement.

**Amyloidosis**

Amyloidosis is a rare diverse condition caused by the pathologic extracellular deposition of abnormal insoluble proteins throughout the body. It may exist as a primary disease or, more commonly, may be secondary to a wide variety of pathologic processes ranging from chronic infection or inflammation to malignancy. Hereditary forms also exist. Lung involvement may manifest as diffuse reticulonodular interstitial thickening, consolidations, or solitary or multiple round or oval parenchymal nodules that may calcify (approximately 50%), cavitate, and slowly enlarge. Pleural involvement most commonly manifests as pleural effusions. Tracheobronchial involvement may exhibit concentric airway thickening (nodular or irregular), mural and intraluminal nodules, nodular and irregular narrowing of the tracheal lumen (Fig.13). Lobar or segmental collapse may be seen with endobronchial obstruction due to amyloid deposition. In certain cases of diffuse involvement, there is a significant component of calcification and ossification of the lesions. This finding leads to a second diagnosis of tracheopathia osteoplastica. In presence of calcification, differentiation is made on the basis of posterior membrane involvement.

**Tracheobronchomegaly**

Tracheobronchomegaly is a rare condition characterized by irreversible dilatation of the trachea and proximal bronchi. The cause of this condition is unknown initially thought to be a congenital anomaly but now presumed to be due to chronic irritation and inflammation. Most cases present with chronic cough or repeated respiratory tract
infections. CT is the best imaging modality for diagnosis and also demonstrates the scalloped appearances (diverticulosis).

**Tracheobronchopathia osteochondroplastica**

Tracheobronchopathia osteochondroplastica is an idiopathic non-malignant disease of large airways featured by submucosal cartilaginous to osseous nodules overlying the cartilaginous rings, which may be focal or diffuse. It is a rare benign disease, which is usually asymptomatic but may cause cough, wheezing and dyspnea on exertion. The disease is characterised by diffuse nodularities, or polyps, consisting of cartilaginous and/or osseous metaplastic tissue involving the anterior and lateral walls of the tracheobronchial tree and sparing the *pars membranacea*. The nodules are 1 to 3 mm in diameter and may cause narrowing and rigidity of the trachea. CT showed tiny, subcentimeter, submucosal, sessile nodules, some of them calcified, at the different levels of the trachea, with predominance in the two lower thirds of the trachea, and also in the right main bronchus (*Fig. 13*). No significant luminal narrowing was observed and, typically, the posterior membranous wall of the trachea was spared.

**Secondary tracheal malignancy**

The central airways may be subject to direct invasion by primary malignancies of the thyroid, oesophagus, lung and larynx. CT will show the primary neoplasm with evidence of an associated cartilage destruction and an endoluminal mass (*Fig. 14*).

**Mucus**

Mucus is seen as filling defects within the trachea or bronchi on CT. These are usually easily distinguishable as they have bubbles of air within them or are low density strands (*Fig. 15*).
Fig. 1: Adenoid cystic carcinoma. Axial (A, B) and coronal (C) images shows focal mass in wall of the trachea.

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Fig. 2: Endobronchial carcinoid. CT image in soft-tissue window shows round endo-esobronchial lesion (A). This mass showed enhancement on the post contrast images (B-C).
Fig. 3: Endobronchial carcinoid. Biopsy revealed a carcinoid (A-B).

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Fig. 4: Endobronchial lipoma. CT axial and coronal images in lung and soft-tissue windows showing round lesion with fat density (HU -109) in right lower lobe bronchus (arrows).

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Fig. 5: Endobronchial lipoma. Biopsy revealed a lipoma.

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Fig. 6: Foreign body. 2-year-old girl who presented with shortness of breath and wheezing. Anteroposterior radiograph (A) shows hyperinflation and resultant apparent paucity of vessels within right lung; nonmetallic foreign body is not detectable. Coronal CT images (B) in lung window shows foreign body (arrow) in bronchus intermedius and right upper lobe bronchus and hyperinflation of the right lower lobe. Bronchoscopy revealed peanuts's pieces.

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Fig. 7: Foreign body. Axial CT images in soft tissue window shows a tooth (arrow) in left lower bronchus.

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Fig. 8: Tracheobronchomalacia. Axial images shows narrowing of trachea and main bronchi. Marked narrowing of the antero-posterior diameter of the carina and right and left main bronchi due to collapse of the tracheal and bronchial cartilage during end expiration (ESP).

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Fig. 9: Post intubation stenosis. CT axial soft-tissue window demonstrate focal subglottic stenosis (B-C) that resulted from prolonged intubation.

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Fig. 11: Post-tuberculous stenosis. CT coronal lung-tissue window (A, arrows) and VRT image (B) show some focal eccentric stricture in distal right main bronchus and bronchus intermedius.

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Fig. 10: Post intubation stenosis. Reformatted coronal CT image (Fig 10) obtained in the same patient shows marked subglottic narrowing (arrow).

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Fig. 12: Amyloidosis. Axial and coronal MIP reconstruction images using soft-tissue windows setting demonstrates thickening and submucosal calcification of tracheobronchial tree.

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Fig. 13: Fig. 13 Tracheobronchopathia osteochondroplastica. Coronal images using soft and lung tissue windows shows submucosal, sessile nodules of trachea wall.

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**Fig. 14:** Endobronchial metastasis from lung carcinoma in 65-year-old man. Axial CT image shows enhancing oval nodule (arrow in C) intimately associated with bronchus.

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**Fig. 15:** Endotracheal mucus. Axial CT image shows rounded tiny nodule that is adherent to anterior wall aspect of tracheas.

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Conclusion

Large-airway tumors and tumorlike conditions are rare. Knowledge of the characteristic CT findings of tracheobronchial tumors can help in diagnosis and treatment planning.
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