Tracheal tumors:
Radiologic-pathologic correlation of tumors and mimics

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Tracheal tumors

Teaching points

- Review cross sectional findings of tracheal tumors and tumor-like entities
- Comparison of imaging findings with histologic pathology findings
- Discuss pearls and pitfalls in accurately diagnosing and classifying tumors and mimics

Table of contents/outline

- Classification of tracheal tumors
- Describe the cross sectional features of tracheal tumors and their mimics with pathologic correlates
- Identify distinguishing imaging and clinical features of each tumor
- Discuss mimics, diagnostic pitfalls, and management of the described entities
Tracheal lesions

Focal lesions

- **Primary malignant tumors:**
  - Bronchogenic adenocarcinoma
  - Squamous cell carcinoma
  - Small cell carcinoma
  - Carcinoid
  - Mucoepidermoid carcinoma

- **Endobronchial metastases**

- **Benign**
  - Endobronchial hamartoma
  - Tracheal leiomyoma
  - Lipoma
  - Pleomorphic adenoma
  - Granular cell tumors

Diffuse lesions

- Relapsing polychondritis
- Wegner’s Granulomatosis
- Infection
- Metastasis
- Amyloidosis
- Papillomatosis
- Tracheobronchopathia osteochondroplastica
Normal anatomy of trachea

- Extends from lower border of the larynx (2 cm below the vocal cord) to the carina.
- Average length: 10-12 cm
- Normal angle of bifurcation: 70+/−20 degrees
- Four layers: mucosa, submucosa, cartilage, muscle
- 18-22 semicircular incomplete rings of cartilage connected by annular ligaments of fibroconnective tissue anteriorly and laterally
- Posterior wall consists of trachealis muscle and fibrous connective tissue
- Wall thickness: 1-3 mm
- Normal transverse internal diameter: 15-25 mm in male and 10-21 mm in female
- Distally, bronchi are differentiated from bronchioles by the presence of cartilage in their wall
- Blood supply: inferior thyroid, bronchial and intercostal arteries
- Nerve supply: Recurrent laryngeal nerve
TUMORS
Epidemiology of malignant tracheal tumors

• < 0.4% of all tumors
• 0.1% of cancer deaths
• Mostly malignant which arise from surface epithelium or salivary gland.
• Imaging modality of choice: CT with 3-D reconstructions, PET/CT

• Metastasis to trachea:
  1. Direct invasion from: Thyroid, lung, esophagus, larynx
  2. Hematogenous: Kidney, breast, colon, melanoma
Squamous cell carcinoma

- Most common tracheal tumor
- Male >> Female
- Highly associated with smoking
- Becomes symptomatic if obstructs more than half of the cross sectional area: Cough, hemoptysis, dyspnea

- Imaging:
  - Most common in lower third of trachea
  - Polypoid mass
  - Direct invasion into mediastinum
  - Lymph node metastasis
  - Distant metastases are uncommon
A&B: Mass centered in the superior trachea with posterior extension to the esophagus
C: U/S- guided Supraclavicular lymph node biopsy was performed
D: Infiltration by large atypical neoplastic cells with ample amount of eosinophilic cytoplasm and prominent nucleoli. Numerous mitosis are identified.
Adenoid cystic carcinoma

- 2nd most common tracheal tumor
- M=F; younger predilection
- Submucosal tumor
- Intact mucosa and smooth contour
- Cough, hoarseness, dyspnea, wheeze
- Treatment: Surgical resection

- CT: smooth, focal mass in the trachea or main bronchi
- Longitudinal involvement more than cross sectional
- 3D reconstruction helpful in evaluation of extent of involvement
- Lymphadenopathy and distant metastasis uncommon
**A&B:** Intra-luminal mass within the proximal right mainstem bronchus with smooth contours

**C:** Avid and homogenous FDG uptake within the tumor suggestive of intermediate to high grade.

**D:** ill defined firm white-tan mass enveloping the trachea measuring at least 3.6 x 5 x 0.8 cm and extending into the right superior lobar bronchus. 10 X image with H&E demonstrates respiratory mucosa at top with nests of tumor below.
Lung cancer

- Leading cause of cancer mortality.
- 29% and 26% of all cancer deaths in men and women.
- Main cell types: Adenocarcinoma, Small cell, Large cell, and SCC
- Most common in trachea: SCC and small cell
- Risk factor: Smoking

- SCC: Lobar collapse (differential contrast enhancement), distant metastasis rare
- Small cell: primary endobronchial lesion usually invisible, extensive mediastinal and hilar lymphadenopathy, distant metastasis more common
A&B&C: Large infiltrative mediastinal mass causing left upper lobe left lower lobe collapse with invasion of mediastinal structures.

D: The section reveals diffuse infiltration by neoplastic cells with high nucleo-cytoplasmic ratio, nuclear molding, salt and pepper chromatin, scant cytoplasm and numerous mitoses.
A: Large opacity in the right lower lung
B & C: Lesion within the distal main bronchus causing post obstructive pneumonia
D: diffuse infiltration by poorly differentiated large neoplastic cells with nuclear pleomorphism, numerous mitoses and areas of necrosis
Endobronchial carcinoid

- 1-2% all pulmonary neoplasms
- 25% of all carcinoid tumors
- Most common in adolescent and young adults
- Central bronchi
- Cough, hemoptysis, post-obstructive pneumonia, wheezing
- May present as carcinoid syndrome (less than 5%)
- Well defined spherical or oval mass with lobulated borders
- Intense contrast enhancement
- 1/4th calcified
- Can exhibit minimal or no uptake on FDG-PET
- 68-Ga DOTATATE PET/CT: Approved by FDA in June 2016, higher resolution than 111-In pentetreotide SPECT-CT for tumor localization
- Associated with right heart valvular disease
- May metastasize to liver
A: Avidly enhancing well defined ovoid nodule in the proximal left main bronchus.

B: Lower than expected FDG uptake for a malignant tumor on PET/CT

C&D: 1.6 x 1.0 x 1.0 cm well circumscribed, endobronchial tan-white nodule arising from the right upper lobe bronchus, within 0.5 cm of the inked specimen edge, and 0.5 cm to the inked bronchial specimen edge. 20 X H&E shows small nests of uniform cells representing a typical carcinoid.
Endobronchial lymphoma

- Rare manifestation of Hodgkin's or Non-Hodgkin’s lymphoma
- Arises de novo from bronchus associated lymphoid tissue
- Airway obstruction, coughing, wheezing
- No specific radiologic findings
- Diagnosis based on biopsy
- Treatment with chemotherapy
A&B: Large intraluminal mass extending into the right main and right intermediates bronchus.
C: Intense uptake corresponding to the intraluminal soft tissue mass and subcarinal lymphadenopathy
D: Sections show bronchial mucosa with a lymphoid infiltrate expanding the submucosa, composed of predominantly small sized cells with condensed chromatin and irregular nuclei. The findings are consistent with bronchial involvement by lymphoma.
Endobronchial metastases

- Direct invasion or hematogenous spread
- Direct invasion far more common
- Direct invasion: Lung, esophagus, thyroid
- Hematogenous metastases: RCC, breast, colon, HCC, melanoma
- Solitary or multiple nodules
- Rare entity
A&B: Enhancing nodule in the left main bronchus which has resulted in partial collapse of the left upper lobe in a patient with known history of lymphoma.

C: Fragment of respiratory mucosa

D: Fragment of metastatic melanoma
TUMOR LIKE LESIONS AND MIMICS
Granulomatosi with polyangiitis (Wegner Granulomatosis)

- Systemic necrotizing granulomatous vasculitis mainly affecting respiratory system
- 90% of patients present with pulmonary involvement
- ANCA +
- Concurrent glomerulonephritis
- CT: Tracheal wall thickening with ulceration
- Involves posterior wall
- Predilection for subglottic trachea which results in subglottic stenosis
- Pulmonary findings: Cavitary nodules, consolidations and GG opacities
**A&B:** Diffuse tracheal wall thickening producing bronchial stenosis.

**C:** Acute and chronic inflammation, follicular bronchiolitis, dense interstitial lymphoplasmacytic infiltrate and blood vessels with granulomatous inflammation and fibrosis.
Infection

- Viral: parainfluenza or RSV; subglottic or laryngeal narrowing
- Tuberculosis: 10-40% of patients with TB exhibit features of endobronchial involvement. Thickened and irregular wall with active disease, smooth stenosis after treatment
- Rhinoscleroma: Chronic granulomatous condition of the nose and upper airway structure; result of K. rhinoscleromatis; F>M; 10-30 y; chronic course with 25% chance of relapse; treatment with antibiotics and surgery if results in obstruction
A&B: Diffuse tracheal wall thickening with mild nodularity without calcification compatible with Rhinoscleroma
C: Mucoid impaction within the lobar bronchus (finger in glove sign), a nonspecific finding but suggestive of fungal infections such as aspergillosis
Amyloidosis

- Rare disorder of deposition of abnormal proteinaceous material in trachea
- CT: diffuse nodular thickening vs short segment stenosis
- The subglottic larynx and adjacent cervical trachea are most commonly involved
- May calcify and resemble tracheopathia osteochondroplastica
- It involves the posterior wall
- Present with dyspnea, wheeze, cough, hemoptysis, and recurrent pneumonia
A&B&C: Partially calcified soft tissue nodule in the trachea, involving the posterior membrane.

D: Low magnification image demonstrates eosinophilic amorphous material compatible with amyloid deposition.
Relapsing Polychondritis

- Recurrent inflammation involving cartilage in the body
- Incidence: 1 in 285000
- Involves trachea in half of the cases
- Mortality often due to pneumonia
- Diffuse smooth tracheal and bronchial wall thickening sparing posterior wall
- Other: subglottic stenosis, air trapping, tracheal cartilage calcification
- Treatment: Medical (corticosteroid, azathioprine) versus surgical with limited indications (limited involvement or to maintain airway patency)

Diffuse smooth tracheal wall thickening sparing the posterior wall
Respiratory Papillomatosis

- Infection by HPV
- At time of birth versus oral-genital sexual transmission
- Voice change and stridor, cough, recurrent pneumonia, dyspnea
- Lung: multiple bilateral thin walled cysts and nodules
- Trachea: endo-luminal nodules
- Treatment: Cryotherapy
- Potential malignant transformation to SCC
Tracheobronchopathia Osteochondroplastica (TPO)

- Idiopathic and benign.
- Submucosal osteocartilagenous nodules
- $M >>> F; > 50$ y
- Often incidental
- Lower 2/3 of trachea and main bronchi
- Spares the posterior wall
- No malignant transformation potential
Foreign body/Mucus plug

- Children > adult
- Ipsilateral air trapping in expiratory image
- Post obstructive collapse / atelectasis
- CT and virtual bronchoscopy best at localization
Conclusion

- Tumors and tumor like conditions of the airways are rare
- Majority of airway tumors are malignant
- CT is the modality of choice for anatomic localization
- FDG PET/CT provides assessment of tumor metabolic activity
- Combining clinical, imaging, and pathology data can facilitate appropriate management strategy
References

Thank You!

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