Rare Airway Tumors - Malignant

Marwan Saoud MD, Kassem Harris MD, FCCP

Co-Chair: WABIP Rare Lung, Pleura & Airway Disorders
Rare Airway Tumors (RATs)

- Tracheobronchial tumors that have not been extensively studied in literature due to limited diagnostic feasibility

- They represent 0.1% of all primary lung tumors

- Occur anywhere from the subglottus to the segmental bronchioles

- Often misdiagnosed in early stages as obstructive lung disease
RATs Cell Type

• Mesenchymal Cell
• Salivary Gland
• Epithelial Cell
• Miscellaneous
Mesenchymal Cell RATs

**Malignant**

- Fibrosarcoma
- Chondrosarcoma
- T-cell Lymphoma
Salivary Gland RATs

Malignant

Myoepithelial Carcinoma
Mucoepidermoid Carcinoma
Adenoid Cystic Carcinoma
Malignant

- Carcinoid Tumor
Fibrosarcoma

- Malignant mesenchymal cell tumor
- Prevalent in children and young adults, males>females
- Associated with exposure to radiation
- Manifest as atelectasis or post-obstructive pneumonitis on x-ray and as smooth, lobular nodules or masses on CT scan
- Appears as a multi-nodular mass on bronchoscopy
- Biopsy is the definitive diagnosis and reveal spindle cells in herringbone pattern
- Bronchoscopic resection is the preferred modality of treatment
Chondrosarcoma

- Malignant mesenchymal cell tumor
- Mean age 30-60 years with male:female ratio of 1.3:1
- Characteristic CT findings including bone and soft-tissue involvement with scattered areas of calcification
- Appears as a polypoid mass on bronchoscopy
- Biopsy is the definitive diagnosis and reveal cartilaginous and binuclear cells with open chromatin
- Treatment options include:
  1. Surgical resection
  2. Adjuvant chemotherapy and/or radiation therapy for extensive tumors

Radiographics 2002; 22(3): 621-37
T-cell Lymphoma

- Malignant mesenchymal cell tumor
- Prevalent in adults age 40-60 years old, females>males
- Associated with tobacco smoking
- Variable size lesions on radiological imaging as well as bronchoscopy
- Biopsy is the definitive diagnosis
- Tissue stains positive for CD3, CD4, and CD5
- Treatment options include:
  1. Chemotherapy: pirarubicin, cyclophosphamide, vincristine and steroids
  2. Surgical resection after chemotherapy

Respirol Case Rep 2015; 3(2): 44-7
(A) Computed tomography (CT) revealed chronic pyothorax with calcified foci on the right and a mass inside the bronchus intermedius.

(B) Flexible bronchoscopy identified an endobronchial tumor obstructing the bronchus intermedius.

(C) Positron emission tomography with [18F] fluoro-2-deoxyglucose and CT revealed uptake at the endobronchial tumor.

(D) CT after the chemotherapy demonstrated that the endobronchial tumor markedly diminished.
Myoepithelial Carcinoma

- Malignant salivary gland tumor
- 20 cases reported, Male:Female ratio of 1:1
- Detected as opaque shadows with defined borders on x-ray and CT scans
- Appears as a smooth, vascular mass with defined borders on bronchoscopy
- Biopsy is the definitive diagnosis
- Histology consistent with glandular differentiation with dual epithelial and myoepithelial cell population; occasional atypia and increased mitotic figures seen
- Tissue stains positive for p-53 and c-Kit (CD117)
- Surgical resection is the preferred treatment modality

*Arch Pathol Lab Med* 2004; **128**(1): 92-4
Mucoepidermoid Carcinoma

- Malignant salivary gland tumor
- Reported in younger population (<30 years of age), equal in males and females
- Appears as ovoid or lobulated polypoid nodules on x-ray
- Have the characteristic "pneumonic consolidation" and "punctuate calcifications" on CT scan
- Appears a glossy, non-vascular mass on bronchoscopy
- Biopsy is the definitive diagnosis
- Histology consistent with mucus-secreting cells, squamous cells and intermediate cells
- Treatment options include:
  1. Bronchoscopic resection
  2. Surgical resection

Arch Pathol Lab Med 2007; 131(9): 1400-4
Mod Pathol 2014; 27(11): 1479-88
Left main stem completely occluded with mucoepidermoid tumor
Axial chest CT showing a highly vascularized left main stem occlusive mucoepidermoid tumor

Coronal chest CT showing a complete obstruction of the left main stem with mucoepidermoid tumor
Mucoepidermoid Carcinoma

Neoplastic tissue composed of round to oval epithelioid cells and occasional goblet cells punctuated by mucin containing cystic spaces
Adenoid Cystic Carcinoma

- Malignant salivary gland tumor
- Equal prevalence in males and females, mean age of 46 years
- Detectable on x-ray and CT as well as positive uptake on PET scan
- Appears a nodular, vascular lesion with characteristic “ice-berg” appearance on bronchoscopy
- Biopsy is the definitive diagnosis with 3 histological cell subtypes: Tubular, Cribriform and Solid (most aggressive)
- Tissue stains positive for keratin, CK7, CD117S-100, and SMA
- Treatment options include:
  1. Surgical resection
  2. Bronchoscopic resection
  3. Pneumonectomy if there is extensive bronchial involvement

*Clin Oncol (R Coll Radiol)* 2015; 27(12): 732-40
*Oncol Lett* 2015; 9(3): 1475-81
Carcinoid Tumor

- Malignant epithelial cell tumor
- Prevalent in younger population (<35 years of age)
- Appears as spherical or ovoid nodules on radiological imaging with vascular enhancement on CT scan
- Appears as a large polypoid lesion with narrow stalk arising from the lumen on bronchoscopy
- Biopsy is the definitive diagnosis with intra-cytoplasmic granules on electron microscope
- Tissue stains positive for chromogranin and synaptophysin
- Treatment options include:
  - Surgical resection
  - Bronchoscopic ablation

*Case Rep Pulmonol* 2014; **2014**: 349707
Carcinoid Tumor

Carcinoid tumor of the right middle lobe

Radial Endobronchial Ultrasound showing the highly vascularized tumor (arrows).
Outcomes

- RATs prognosis depend on multiple factors:
  - Tumor malignant potential
  - Tumor location
  - Patient’s co-morbidities
  - Risks of treatment modality

- Benign tumors are usually localized and amendable to resection with no or minimal risk of recurrence

- Outcome of malignant tumors depend mainly on lymph node and adjacent tissue metastasis

- Tumors found on the carina have poor prognosis due to the high risk of surgical resection attributed to the anatomical feasibility


*Intern Med* 2013; 52(18): 2113-6
This presentation was prepared by Marwan Saoud MD and Kassem Harris MD, FCCP and reviewed for accuracy and content by members of the WABIP Rare Lung, Pleura and Airway Disorders section.