Rare Lung, Pleura, and Airway Disorders

Rare Airway Tumors - Malignant

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Background

Rare Airway Tumors (RATs)

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

Lancet Oncol 2006; **7**(1): 83-91

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

Epithelial Cell RATs

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

Am J Otolaryngol 2015; **36**(2): 287-9

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

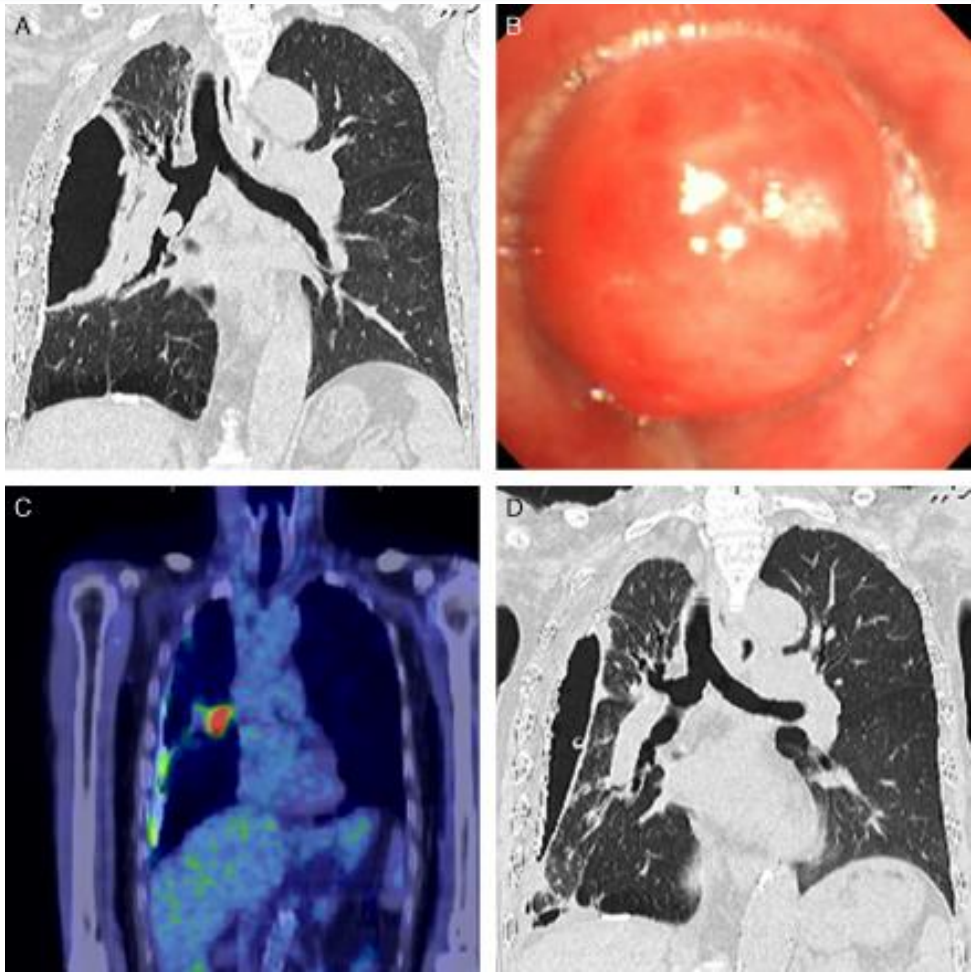
T-cell Lymphoma

- 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

T-cell Lymphoma

(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.



Matsumoto et al. *Respirol Case Rep* 2015; **3**(2): 44-7

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

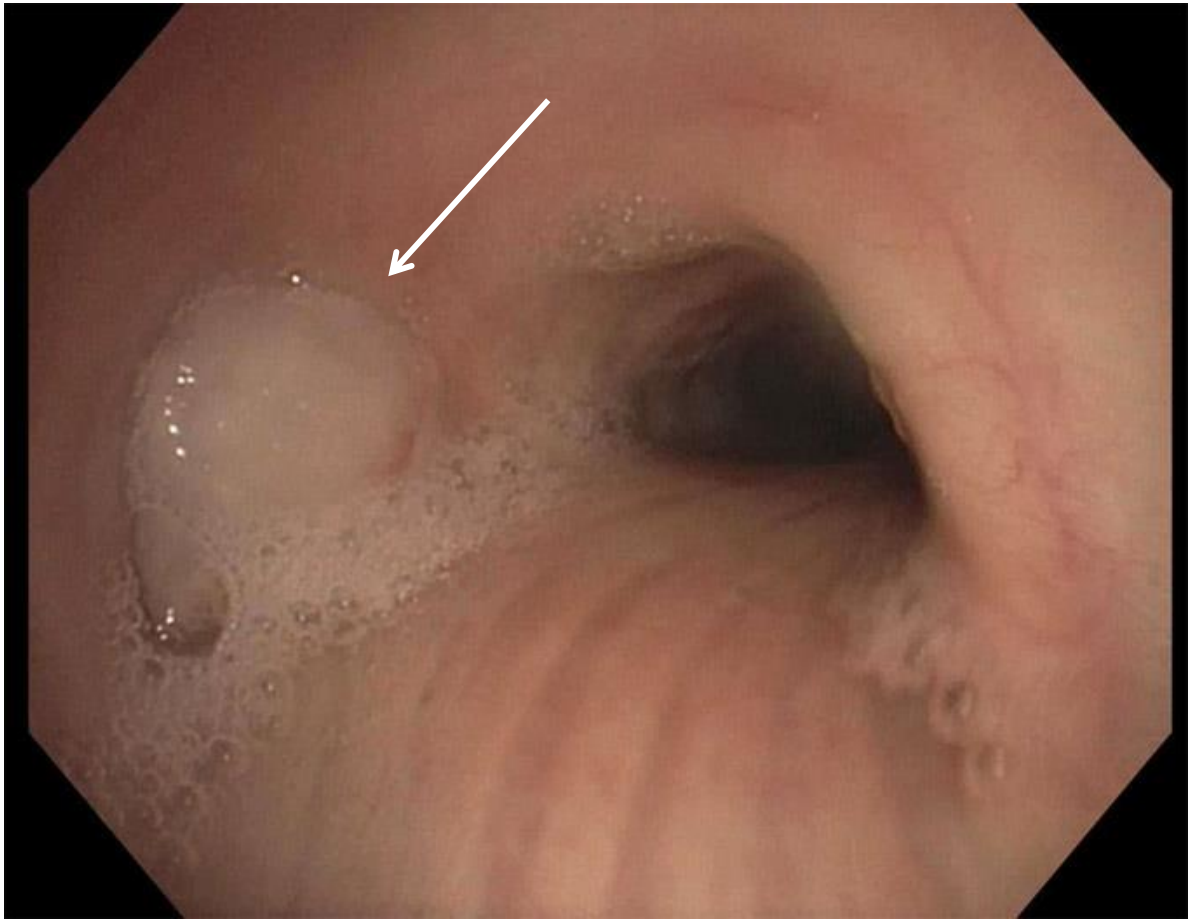
- Malignant salivary gland tumor
- Reported in younger population (<30 years of age), equal in males and females
- Appears as ovoid or lobulated polypoid nodules on x-ray
- Have the characteristic "***pneumonic consolidation***" and "***punctuate calcifications***" on CT scan
- Appears a glossy, non-vascular 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

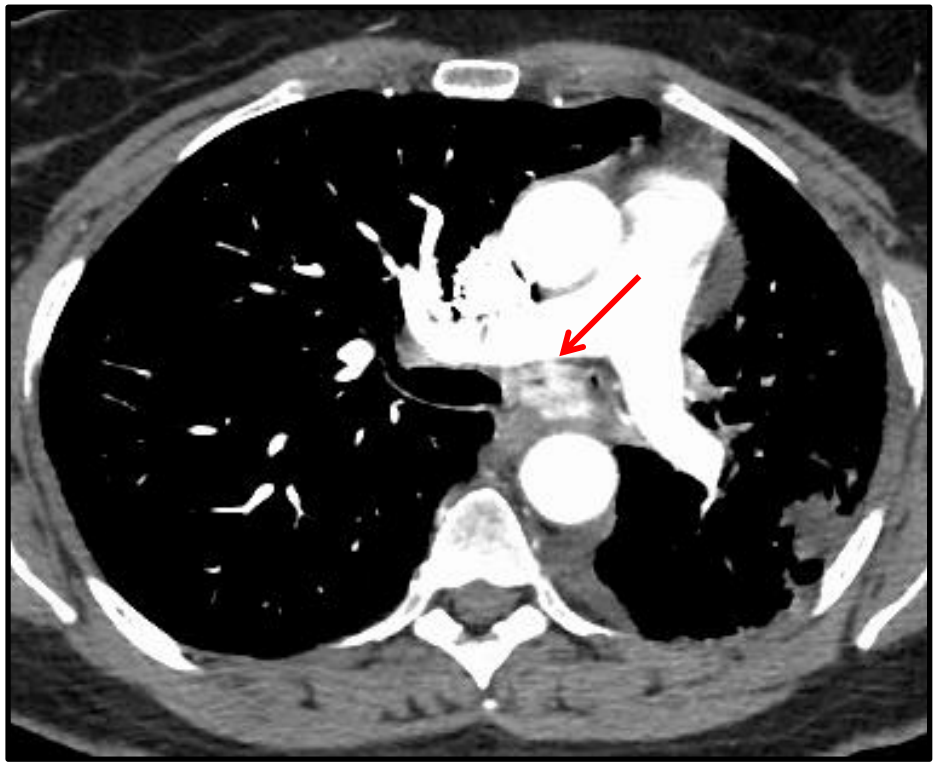
Mod Pathol 2014; **27**(11): 1479-88

Mucoepidermoid Carcinoma

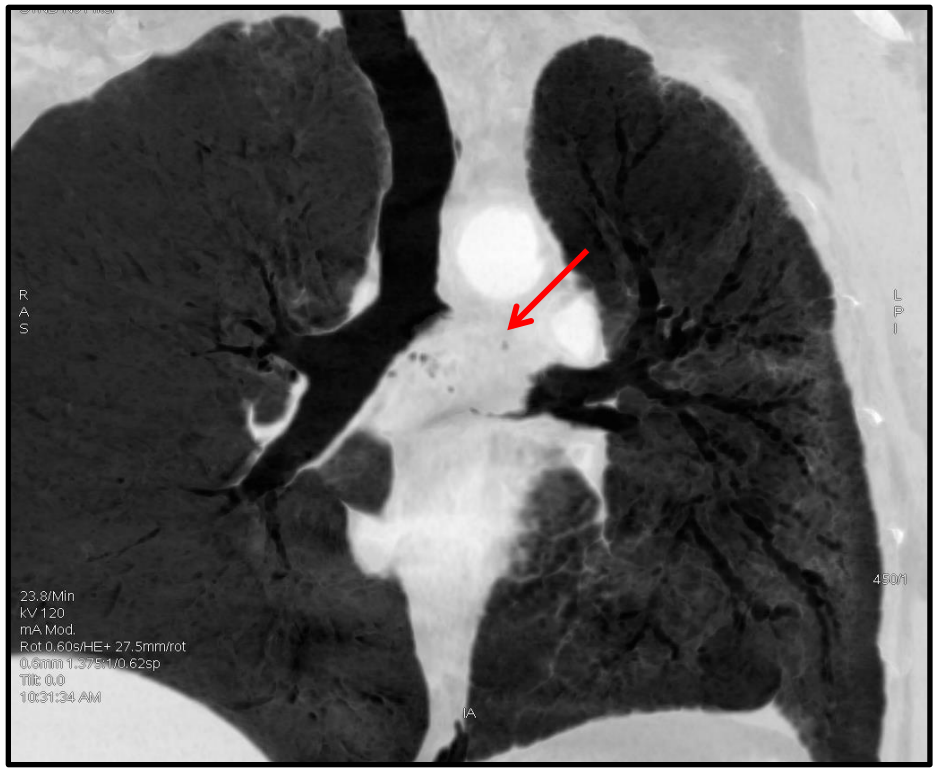
Left main stem completely occluded with mucoepidermoid tumor



Mucoepidermoid Carcinoma



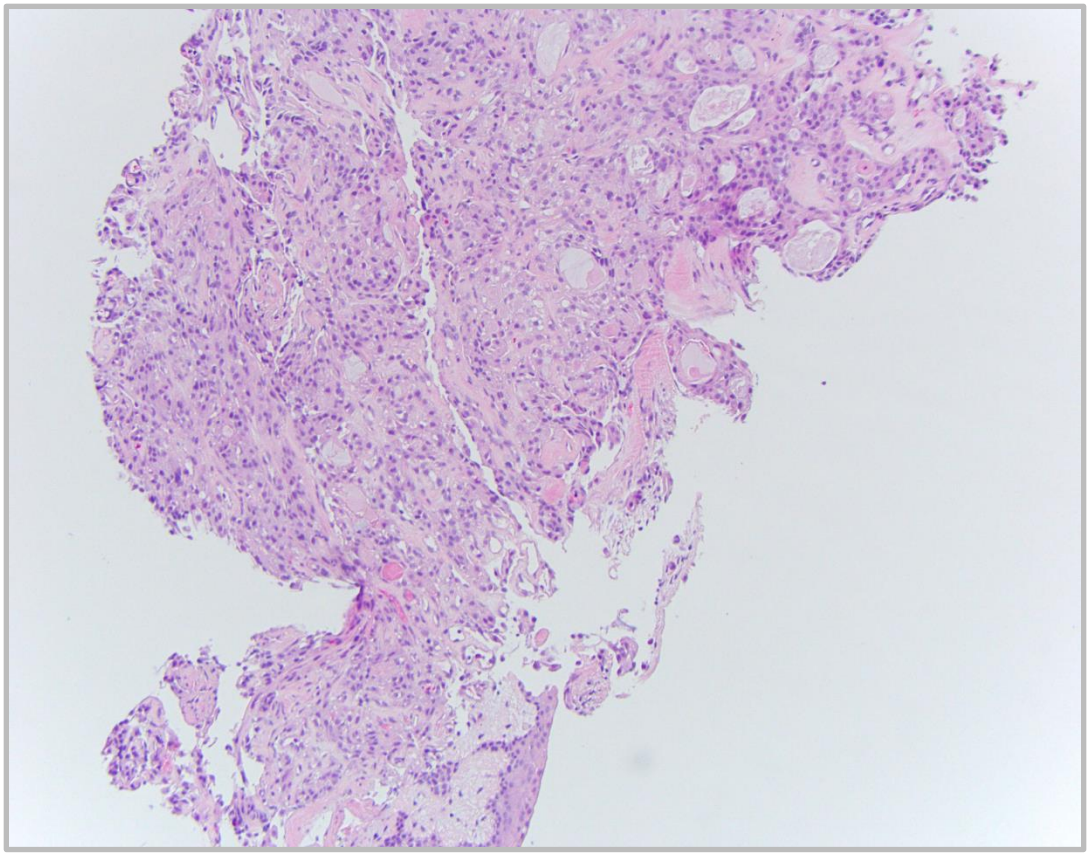
Axial chest CT showing a highly vascularized left main stem occlusive mucoepidermoid tumor



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

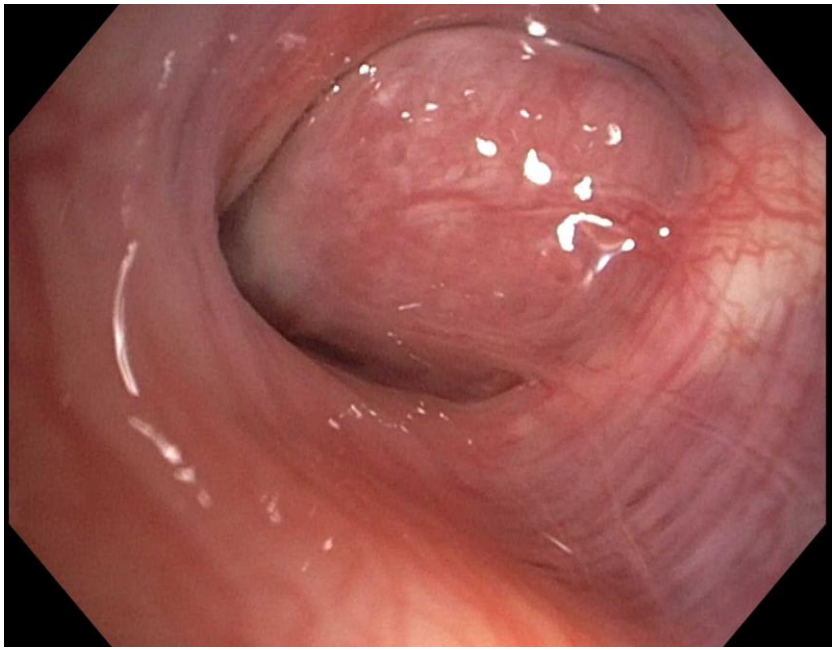
Carcinoid Tumor

- 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

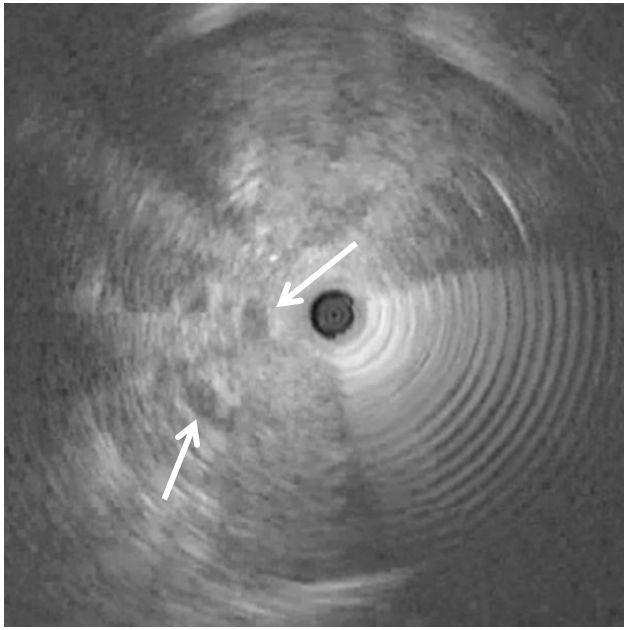
Mayo Clin Proc 1993; **68**(8): 795-803

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

Lancet Oncol 2006; **7**(1): 83-91

Intern Med 2013; **52**(18): 2113-6

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and reviewed for accuracy and content by members of the

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