



Rare Lung, Pleura, and Airway Disorders



# Rare Airway Tumors - Benign

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# Rare Airway Tumors (RATs) Cell Type

- Mesenchymal Cell
- Salivary Gland
- Epithelial Cell
- Miscellaneous



# Mesenchymal Cell RATs

## Benign

- Lipoma
- Schwannoma
- Leiomyoma
- Hemangioma
- Hamartoma
- Chondroma
- Glomus Tumor
- Granular Cell Tumor



# Salivary Gland RATs

## Benign

- Mucus Gland Adenoma
- Oncocytoma



# Epithelial Cell RATs

## **Benign**

- Papilloma



# Miscellaneous RATs

## Benign

- Inflammatory Myofibroblastic Tumor
- Extramedullary Plasmacytoma



# Lipoma

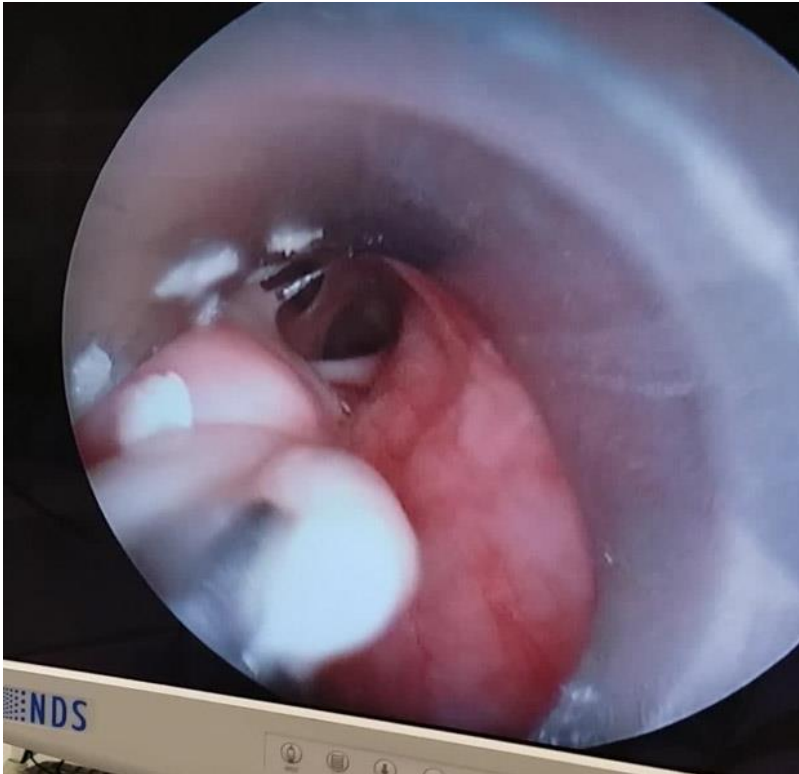
- Benign mesenchymal cell tumor
- Prevalent in adults age 50-60 years old, males>females
- Associated with smoking and obesity
- Appears as a hilar shadow or atelectasis on x-ray
- Appears as a soft, pale, glistening, poorly vascular mass on bronchoscopy
- Definitive diagnosis via biopsy which is difficult due to firm, fibrous sheath around the tumor
- Treatment options include :
  1. Bronchoscopic resection
  2. Surgical resection if there is atypical features
  3. Lobectomy or pneumonectomy if there is parenchymal involvement

*BMC Pulm Med* 2009; **9**: 40

*Pathol Int* 2011; **61**(4): 252-8



# Lipoma



Endobronchial lipoma originating from the superior segment of the right lower lobe



Post mechanical debulking of the endobronchial lipoma





# Schwannoma

- Benign mesenchymal cell tumor
- 51 cases reported in the literature with Male:Female ratio of 2:3
- Appears as heterogenous spherical lesions on CT or MRI
- Appears as a smooth, round, white, non-vascular mass on bronchoscopy
- Definitive diagnosis via biopsy as confirmed by the presence of bipolar and spindle cells within a myxoid matrix
- Tissue stains positive for S-100
- Treatment options include:
  1. Surgical resection
  2. Endoscopic excision for more localized tumors and/or poor pulmonary function



# Leiomyoma

- Benign mesenchymal cell tumor
- Prevalent in adults with mean of age 35-40 years, males>females
- Can be detected on x-ray and CT scans
- Appears as a smooth, pink, polypoid mass with smooth contours and broad base on bronchoscopy
- Biopsy is the definitive diagnosis
- Histology consistent with pseudostratified columnar epithelium with spindle-shaped cells and acidophilic cytoplasm
- Tissue stains positive for caldesmon and desmin
- Treatment options include:
  1. Surgical resection via *carinal resection*, *tracheal sleeve resection*, or *segmental tracheal resection*
  2. Bronchoscopic resection

*Eur J Cardiothorac Surg* 2012; **41**(1): 41-5  
*Lung India* 2013; **30**(1): 57-60



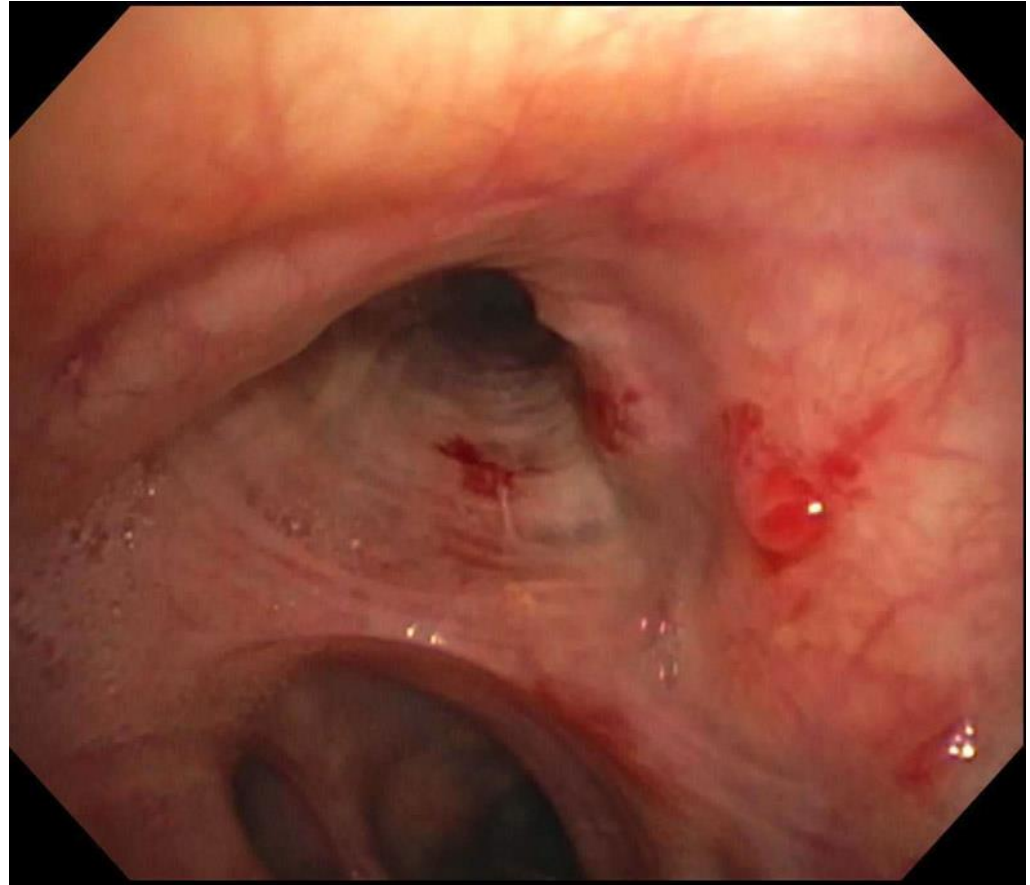
# Hemangioma

- Benign mesenchymal cell tumor
- 10 reported cases in infants
- Rarely captured on plain radiological imaging techniques
- Appears as a vascular/hemorrhagic polypoid lesion on bronchoscopy
- Biopsy is the definitive diagnosis
- Histology consistent with lobular capillary hemangioma
- Treatment options include:
  1. Bronchoscopic resection
  2. Bronchial artery embolization by IR
  3. Medical therapy with high-dose corticosteroids in children



# Hemangioma

## Left Main Stem Hemangioma





# Hemartoma

- Benign mesenchymal cell tumor
- 43 cases reported in adults with mean age of 62 years
- Male:Female ratio of approximately 6:1
- Resembles fatty or calcified lesions on CT scan
- Appears as a smooth, round, pink, non-vascular lesion on bronchoscopy
- Biopsy is the definitive diagnosis
- Collection of fat, cartilage, fibrous tissue and epithelial cells are seen on histological analysis
- Treatment options include:
  1. Bronchoscopic resection
  2. Surgical resection



# Chondroma

- Benign mesenchymal cell tumor
- Prevalent in adults with mean age of 46 years
- Male:Female ratio of 3:2
- Appears as a lobar consolidation on x-ray and pedunculated, vascularized lesion on bronchoscopy
- Biopsy is the definitive diagnosis and tissue analysis consistent with characteristic chondromatous tissue
- Bronchoscopic excision is the treatment of choice



# Glomus Tumor

- Benign mesenchymal cell tumor
- 31 cases reported, mean age of 52 years
- Male:Female ratio of 2:1
- Characterized by marked contrast enhancement on CT scan
- Appears as a vascularized, hyperemic mass with solid surface on bronchoscopy
- Biopsy is the definitive diagnosis and reveals sheets of cells around the capillaries in an eosinophilic cytoplasm
- Tissue stains positive for vimentin and smooth muscle actin
- Treatment options include:
  1. Sleeve resection with primary reconstruction of the trachea
  2. Bronchoscopic resection for intra-luminal tumors



# Granular Cell Tumor

- Benign mesenchymal cell tumor
- 31 cases have been reported in adults
- Can be detected on x-ray or CT scan
- Appears as pedunculated polypoid lesions on bronchoscopy
- Biopsy is the definitive diagnosis and tissue stains positive for S-100
- Surgical resection is the preferred treatment modality





# Mucous Gland Adenoma

- Benign salivary gland tumor
- More common in females than males
- Not easily detectable on x-ray or CT scans
- Appears as solitary, polypoid nodules on bronchoscopy
- Biopsy is the definitive diagnosis and tissue analysis reveals cystic mucus-filled glands
- Treatment options include:
  1. Bronchoscopic resection
  2. Surgical resection



# Oncocytoma

- Benign salivary gland tumor
- 10 cases reported, males>females
- Associated with tobacco smoking
- Not easily detected on x-ray or CT scans
- Appears as polypoid nodules on bronchoscopy
- Biopsy is the definitive diagnosis and tissue analysis reveals polygonal cells forming sheets, trabeculae and acinar structures
- Treatment options include:
  - Surgical resection
  - Bronchoscopic resection

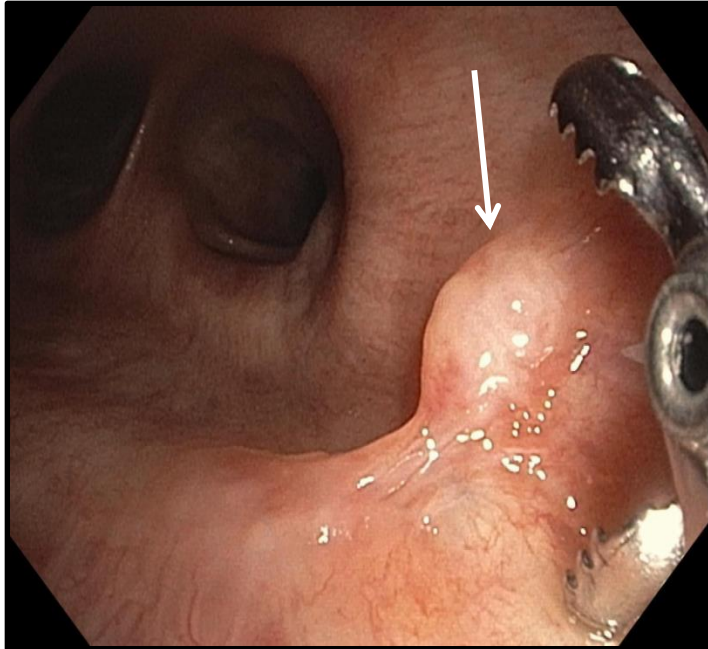


# Papilloma

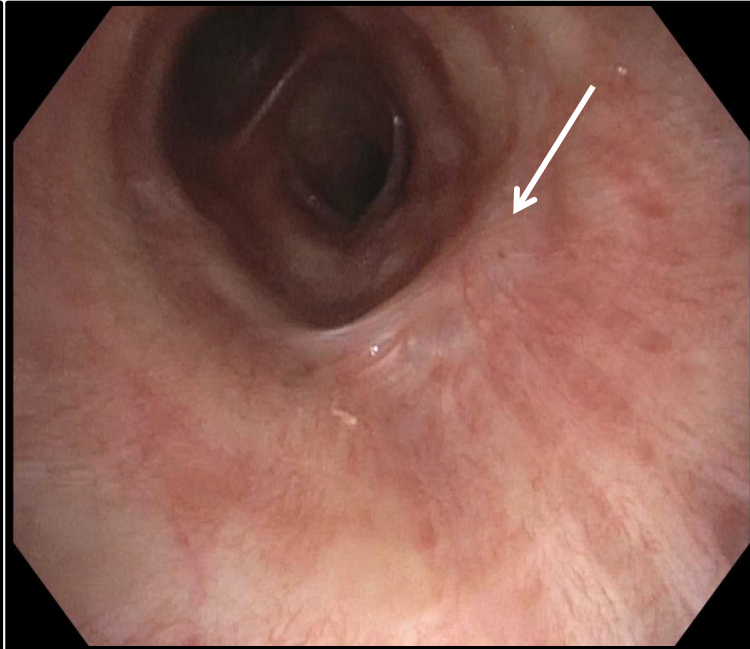
- Benign epithelial cell tumor
- 50 cases reported, mean age of 54-68 years, males>females
- Association with Human Papilloma Virus (HPV)
- Focal bronchiectasis is commonly seen on x-ray
- Appears as polypoid or pedunculated, glistening, non-vascular lesions with tan to red color on bronchoscopy
- Biopsy is the definitive diagnosis
- 3 cell subtypes: *squamous*, *glandular* and *mixed*
- Characteristic histological feature is intracellular mucin
- Tissue stains positive for MUC5AC which is expressed in goblet cells
- Bronchoscopic resection is treatment modality of choice



# Papilloma



**Papilloma of the bronchus  
intermedius**



**Bronchoscopic image 6 months after  
ablation using argon plasma  
coagulation showing minimal scarring  
and no recurrence of the papilloma**

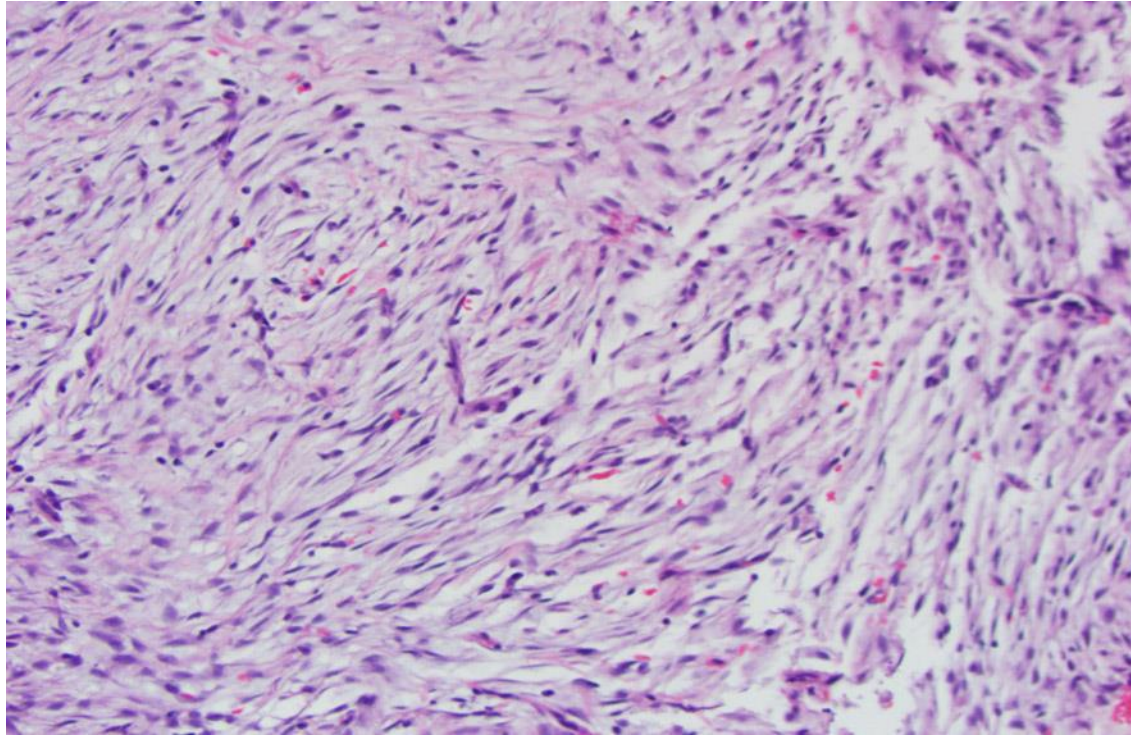


# Inflammatory Myofibroblastic Tumor

- Benign miscellaneous tumor
- Rare tumor reported in young population (<16 years of age)
- Non-specific lesions on x-ray and CT scan
- Appears as a vascular, smooth lobulated mass on bronchoscopy
- Biopsy is the definitive diagnosis
- Histology consistent with myofibroblastic spindle cells, inflammatory plasma cells, lymphocytes and eosinophils
- Tissue stains positive for vimentin, muscle-specific actin, SMA and ALK-1
- Treatment options include:
  1. Surgical resection
  2. Bronchoscopic resection
  3. Medical therapy with NSAIDs, steroids and ALK-inhibitors



# Inflammatory Myofibroblastic Tumor



**Microscopic image of inflammatory myofibroblastic tumor. The image shows bland spindle cells without necrosis or mitoses, fibromyxoid stroma and scattered plasma cells (blue arrow). Hematoxylin and Eosin stain.**



# Extramedullary Plasmacytoma

- Benign miscellaneous tumor
- 5 cases reported, mean age of 56 years, male:female ratio of 4:1
- Appears as a smooth-surface mass on bronchoscopy
- Biopsy is the definitive diagnosis with 5 criteria:
  - Single extramedullary mass of clonal plasma cells
  - Histologically normal bone marrow
  - Lack or decrease in serum or urinary level of monoclonal immunoglobulin
  - Absence of anemia
  - Normal skeletal survey
- Characteristic histological feature is the kappa chain-type and lambda chain-type cells
- Treatment options include:
  1. Argon plasma coagulation via rigid bronchoscopy for narrow-base tumors
  2. Surgical resection followed by radiotherapy for wide-base tumors



- 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





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This presentation was prepared by  
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