Carcinoid Tumors (Bronchial/lung)

Key Points:
- Carcinoid tumors are classified by location and by histology and have variable presentations
- Carcinoid syndrome varies in incidence based on location (common in small bowel, rare in lung)
- Typical bronchial carcinoid has a better prognosis than other primary lung cancers when respectable
- Somatostatin analogues can treat symptoms of syndrome and stabilize tumor, but there are limited options for tumor regression

Background
- Neuroendocrine cells with granules (epithelial, enterochromaffin-like, subepithelial)
- Secrete hormones and biogenic amines: serotonin, corticotropin, histamine, dopamine, substance P, neurotensin, prostaglandins, kallikrein
- Classification by location
  - Foregut (lungs, bronchi, stomach)
  - Midgut (small intestine, appendix, proximal large bowel)
  - Hindgut (distal colon, rectum)
- Classification by differentiation (well or poorly differentiated)
- Incidence: 1-2 per 100,000 (may be higher because of indolence)
- Carcinoid syndrome:
  - Diarrhea, wheezing, flushing
  - Right-sided valvular heart disease (fibrous endocardial thickening -> TR, TS, PR, PS; unknown mechanism)
  - Associated more with small bowel tumors than other subtypes

Lung and bronchial carcinoid
- 2% of primary lung tumors
- Derived from Kulchitsky cells
- Typical well-differentiated neuroendocrine:
  - Presents in 5th decade
  - Symptoms of recurrent pneumonia, cough, hemoptysis, or chest pain
  - Cushing’s due to corticotrophin, less common acromegaly due to growth hormone-releasing factor
  - Carcinoid syndrome (serotonin) in less than 5%
  - Perihilar, indolent with metastases in <15%
  - Histology: epithelial, minor atypia with rare mitoses
  - 5 year survival >90% with resection
- Apytical well-differentiated neuroendocrine: (1/3)
  - Older patients
  - Larger, more peripheral
  - Aggressive, 30-50% metastases
  - Histology: epithelial, atypical, increased mitoses, areas of necrosis

See review article for carcinoids at other locations

Metastatic carcinoid tumors
- Evaluation
  - Abdominal CT to r/o metastases (LFTS not sensitive) – before and after IV contrast (hypervascular and isodense after contrast)
  - 24 hour 5-HIAA (hydroxyindoleacetic acid)
- Clinical course: variable

Treatment
- Somatostatin analogues (octreotide/lanreotide)
  - Receptors on 80% of tumors (G-coupled receptor) -> relieve symptoms of carcinoid syndrome
  - Tumor stabilization, but rarely lead to tumor regression
- Interferon: under study, tumor stabilization
- Chemotherapy: poor response
- Embolization and chemoembolization for liver metastases