Metastatic, Poorly-Differentiated Neuroendocrine Tumor of the Esophagus: Case Report and Literature Review of a "Rare Rarity"!

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Introduction:

- Neuroendocrine tumors (NET) most commonly develop in the lungs, appendix, bowel and pancreas
- Esophageal NETs are extremely rare, comprising < 2% of all NETs
- Most common symptom is dysphagia although patients may be asymptomatic
- We present a case of metastatic esophageal NET with literature review

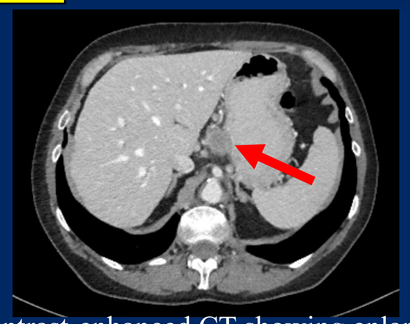
Case Report:

- 61yo female presents with worsening chest tightness, weight loss and diarrhea x 5 months
- Abdominal CT scan revealed enlarged gastrohepatic lymph nodes (Image 1)
- EGD showed a 3cm ulcerated lesion at the gastroesophageal junction (GEJ). Biopsies suggested high-grade NET
- Patient was referred to our center for further management

Case Report (continued):

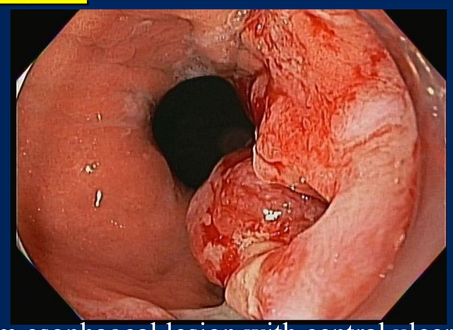
- PET/CT confirmed a 3.0 x 2.3cm ill-defined, hypermetabolic, soft tissue mass at the GEJ (SUV max 9.0) and several hypermetabolic, enlarged gastrohepatic LNs, largest measuring 2.8 x 2.0cm (SUV max 8)
- Repeat EGD revealed a 3cm distal esophageal lesion with central ulceration extending across the GEJ into the high cardia (Image 2)
- On endoscopic ultrasound (EUS), a hypoechoic mass is seen with involvement of the muscularis propria (T2). Peri-gastric lymphadenopathy was present. Transgastric fine needle aspiration (FNA) of the largest LN confirmed metastatic neuroendocrine tumor (T2N1)
- Repeat endoluminal biopsies from the esophageal mass confirmed poorly-differentiated NET, CD56 & synaptophysin positive, Ki-67 > 90%
- Multidisciplinary tumor board recommended chemotherapy with carboplatin and etoposide
- Restaging CT scan after 6 treatment cycles revealed a new hypermetabolic focus in the left hepatic lobe consistent with metastasis; plan to start 2nd line immunotherapy with ipilimumab + nivolumab

Image 1:



Contrast-enhanced CT showing enlarged gastrohepatic lymph node

Image 2:



3cm esophageal lesion with central ulceration at GEJ/high cardia

Conclusion:

- Metastatic poorly-differentiated high-grade NET of the esophagus is very rare
- First-line therapy is chemotherapy
- Average prognosis with treatment is 12-18 months
- Immunotherapy may provide additional survival benefit; however, data is extremely limited, so this is typically reserved for second-line therapy (if progression seen on chemotherapy)
- Given the rarity and aggressiveness of these lesions, accurate pathological diagnosis and multidisciplinary discussion is critical for optimal management of these patients

References:

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2. Lee C, Lim Y, Park S, et al. The clinical features and treatment modality of esophageal neuroendocrine tumors: a multicenter study in Korea. BMC Cancer. 2014; 14:569