ESOPHAGEAL HIGH GRADE NEUROENDOCRINE CARCINOMA

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68 year old male with past medical history of anemia, chronic kidney disease, renal osteodystrophy.
Initial abdomen MRI for evaluation of renal cyst showed multiple hepatic lesions
CA19-9: 22
CEA: 3.3
PET/CT scan was requested for further evaluation
MRI Abdomen without contrast:
Multiple T2 hyperintense lesions throughout the hepatic parenchyma. The lesions demonstrate restricted diffusion.
FDG PET/CT:
Intense uptake in the distal Esophagus with adjacent hypermetabolic paraesophageal lymph nodes
FDG PET/CT:
Numerous hypermetabolic metastatic lesions diffusely involving the liver, left gluteus muscle, and L5 vertebral body.
**Abdomen CT:**
Diffuse soft tissue thickening of the distal aspect the esophagus

**MRI:**
Enhancing lesion in the left brainstem
GI ENDOSCOPY

- A large fungating mass with bleeding in the lower third of the lower third of the esophagus.
- The mass is partially obstructing and circumferential.
SURGICAL PATHOLOGY

- Poorly differentiated carcinoma, with evidence of mixed differentiation
- Predominantly poorly differentiated neuroendocrine carcinoma (Ki-67 index approximately 80%)

Monomorphic tumors cells with poorly differentiated neuroendocrine carcinoma
Tumor cells with amphophilic cytoplasm with nucleoli
Synaptophysin positive (protein made by specialized neuroendocrine cells)
Neuroendocrine tumor of the esophagus is very rare and represent only 0.05% of all gastrointestinal NETs. Most NET patients are male in the sixth and seventh decade of life. Typically, the cases are discovered as a polypoid mass causing obstructive symptoms or as an incidental finding in the setting of adenocarcinoma. Mostly located in the distal half of the esophagus and may be associated with adenocarcinoma and Barrett esophagitis. Usually large size (>4cm). Lesions are typically large, polypoid, and confined to the submucosa or lamina propria. Rarely secrete hormones, and their clinical manifestations are similar to those of other types of esophageal cancer.
DISCUSSION

• The prognosis of the tumor depends on the depth or invasion and stage of the tumor.
• Grading is based on histology and proliferative activity and is divided into G1, G2, and G3:

<table>
<thead>
<tr>
<th>Grading</th>
<th>Mitotic figures (n/10 HPF)</th>
<th>Ki-67 positive index (%)</th>
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<tbody>
<tr>
<td>G1</td>
<td>&lt;2</td>
<td>≤ 2</td>
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<tr>
<td>G2</td>
<td>2-20</td>
<td>3-20</td>
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<tr>
<td>G3</td>
<td>&gt;20</td>
<td>&gt;20</td>
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• CT: Irregular thickening or a polypoid lesion in the esophagus that may be confirmed by endoscopic biopsy.
• Differential: Other more common esophageal malignancies, such as esophageal carcinoma, and benign lesions like leiomyoma, fibrovascular polyps, and hemangioma.
References: