Neuroendocrine Carcinoma of the Esophagus: Presentation and Outcomes of a Rare Histology

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INTRODUCTION

- Extremely rare: <0.04% of neuroendocrine carcinoma tumors
- Primary symptoms: dysphagia and chest pain
- Diagnose with barium swallow, CT scan, EGD
  - Confirmed by immunohistochemical staining
- Currently no treatment guidelines
  - Operative treatment the only cure
  - Chemoradiotherapy also commonly used
- Purpose: characterize presentation and examine treatment strategies, including the role of surgery

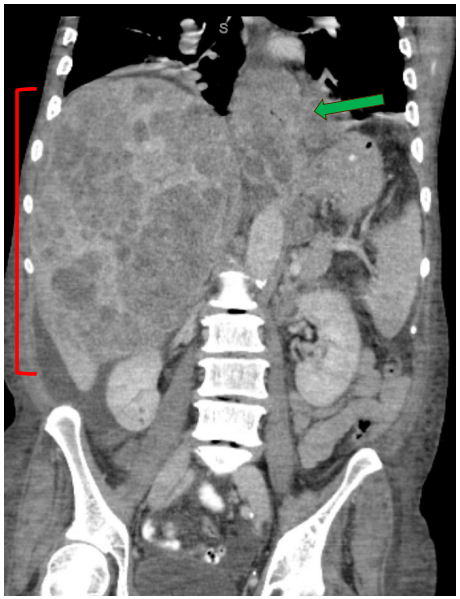

Figure 1: CT scan demonstrating primary esophageal neuroendocrine carcinoma tumor (green arrow) and extensive liver metastases (red bracket).

METHODS

- Retrospective chart review
- 2003-2024
- Work-up included CT Scan and EGD in all patients
  - Some patients had barium swallow and/or PET scan
- Surgical candidates had Minimally Invasive Ivor Lewis Esophagectomy

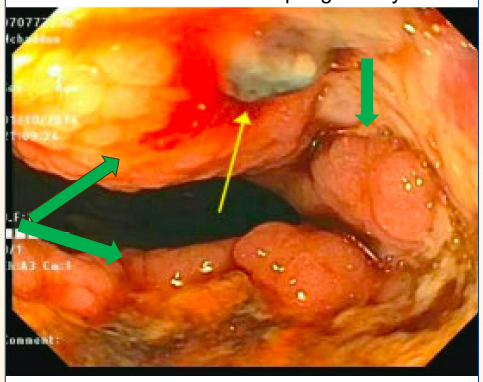

Figure 2: EGD demonstrating and ulcerated neuroendocrine tumor (green arrows) and exposed vessel (yellow arrow).

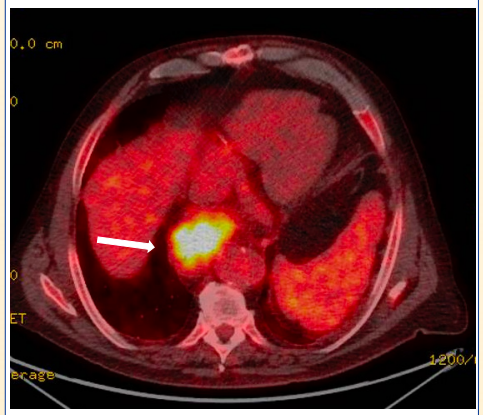

Figure 3: Pre-operative PET scan showing hypermetabolic distal esophageal lesion (arrow).

RESULTS

- 6 patients
- Age[1]: 56 years old
- Symptoms
  - Dysphagia (3 patients)
  - Weight loss (2 patients)
  - Night sweats
  - Hoarseness
  - Chest pain
  - Regurgitation
  - Hematemesis/syncope
- EGD
  - All masses were in the distal 1/3 of esophagus
  - 5/6 masses crossed the GEJ and extended onto cardia of the stomach
- Neoadjuvant therapy: 5 patients
- Surgical patients: 3
- Surgical Outcomes
  - Respiratory failure requiring tracheostomy: 3
  - Anastomotic leak: 2
  - Hospital length of stay: 43, 94, and 38 days
- Nonsurgical patients: 3
  - All had liver metastasis at the time of diagnosis

[1]Reported as a median

Table 1: Esophageal NEC Patients' Presentation and Treatments

Patient	Age (years)	Presenting symptoms	Surgery (yes/no)	Neoadjuvant chemoXRT (yes/no)	Adjuvant chemoXRT (yes/no)	Overall survival (months)
1.	55	Dysphagia	Yes	Yes	Yes	20
2.	74	Chest pain, nausea	Yes	Yes	Yes	8
3.	53	Syncope and hematemesis	Yes	No	No	14
4.	36	Dysphagia to solids, weight loss	No	Yes	N/A	8
5.	65	Night sweats, weight loss, fever	No	Yes	N/A	1
6.	58	Dysphagia to soft solids, hoarseness, regurgitation	No	Yes	N/A	8

CONCLUSIONS

- Esophageal neuroendocrine tumors are rare with heterogeneous presentation
  - Dysphagia and weight loss are most common
- Aggressive and patients often already have extensive M1 disease
  - Commonly in the liver
- Surgical candidates have high morbidity, but do recover to receive adjuvant treatment
  - Leak, pneumonia, respiratory failure
- Imperative to carefully consider and determine surgical candidacy prior to offering operative intervention.
- Further studies may elucidate risk factors for surgical complications.

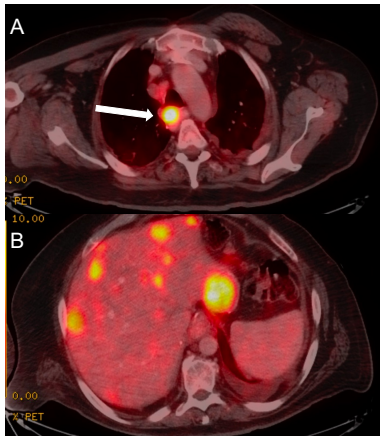

Figure 4. A: Postoperative PET scan demonstrating an FDG-avid paraesophageal lymph node (arrow) B: Postoperative PET scan demonstrating extensive FDG-avid liver metastases

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