

Small Cell Neuroendocrine Esophageal Carcinoma: A Rare Cause of Dysphagia

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Introduction

- New onset dysphagia warrants endoscopic evaluation to rule out structural etiologies, including malignancy.
- The vast majority (>95%) of esophageal cancers are adenocarcinoma or squamous cell carcinoma.
- We report a rare case of esophageal small-cell neuroendocrine carcinoma (NEC) presenting with dysphagia.

Patient Presentation

- A 76-year-old African American female with a history of left lung non-small cell neoplasm status post left lower lobectomy and adjuvant chemo-radiation presented to an outside hospital with dysphagia to solids and unintentional weight loss over the previous month.
- She denied history of smoking.
- Her BP was 163/81, pulse 81, temperature 36.7 and respiratory rate 16.
- Rest of the physical exam was normal.
- Labs showed mild normocytic anemia (hemoglobin 11.2 g/dL).
- Chest CT scan revealed a soft tissue mass at junction of the cervical and thoracic esophagus (Figure 1A).
- Due to the proximity of the mass to the airway, she was transferred to our hospital for evaluation.
- EGD confirmed an esophageal mass located at 16 cm from incisors, just distal to the upper esophageal sphincter, causing near complete esophageal obstruction (Figures 1B-C). Using a pediatric gastroscope, the mass was traversed and noted to extend to 25 cm from the incisors. Multiple biopsies were obtained with histology showing small cell NEC (Figures 1D-F).
- Positron emission tomography revealed lymph nodes metastases above and below the diaphragm, liver and multiple bones.
- Brain magnetic resonance imaging was negative for metastases.
- Patient was not a candidate for surgical resection or stenting given the proximal esophageal location of the tumor and required gastrostomy tube placement for nutrition by interventional radiology.
- Palliative chemoradiation (carboplatin, etoposide, atezolizumab) has started and the patient is currently tolerating therapy for 2 months since diagnosis.

Images

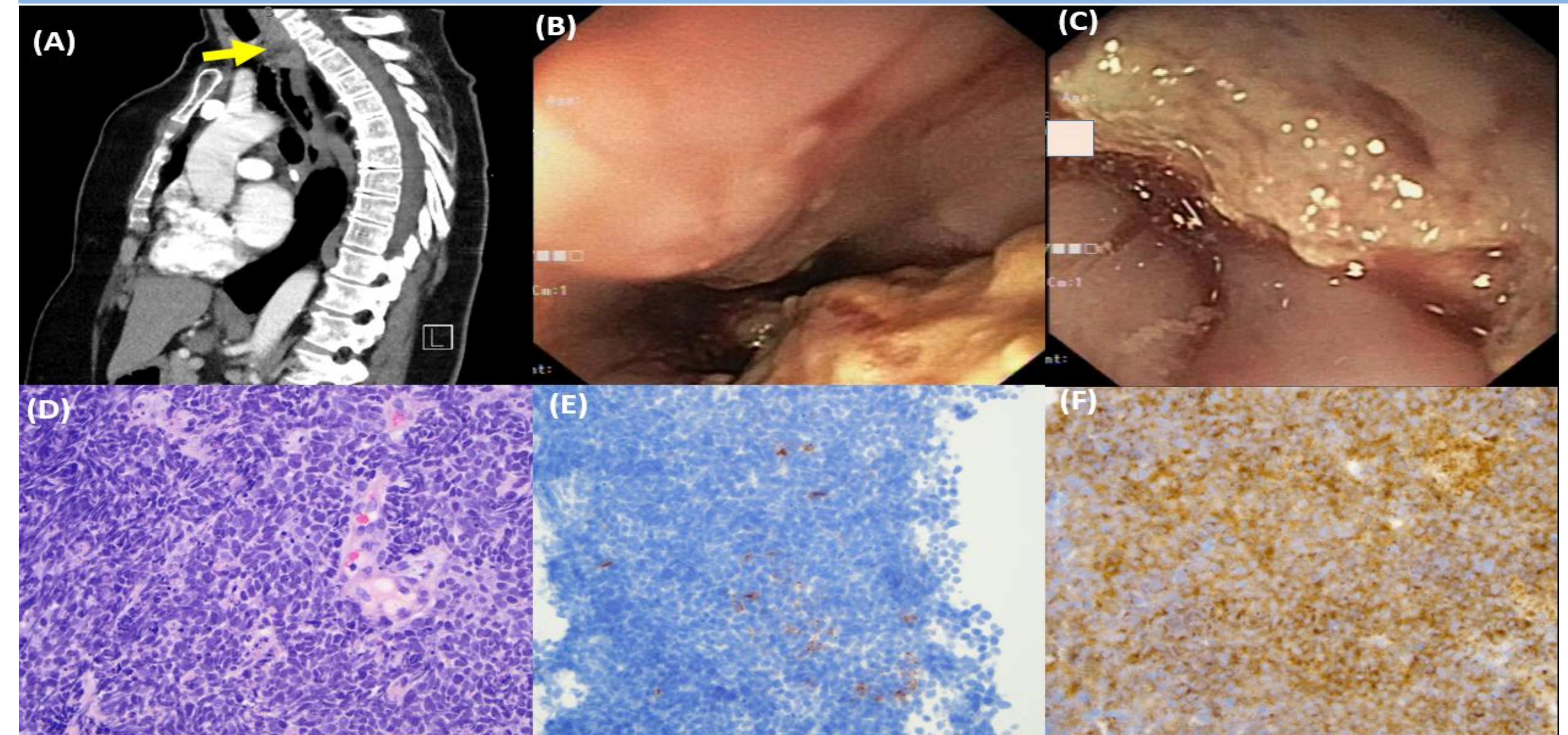


Figure 1A: CT scan of the chest (sagittal view) showing a soft tissue mass at the junction of the cervical and thoracic esophagus (arrow). Figure 1B-C: Esophagogastroduodenoscopy showing an esophageal mass located at 16 cm from incisors causing near complete obstruction of the esophagus. Figure 1D: A high power (400x, H&E) view shows a highly cellular tumor comprised of small cells with scant cytoplasm, high nuclear/cytoplasmic ratio, salt-and-pepper chromatin, and nuclear molding with crush artifact. There are abundant apoptotic cells and frequent mitotic figures, indicating a highly proliferative tumor. Figure 1E: 400x, chromogranin immunostaining, showing focally positive cells, consistent with neuroendocrine differentiation. Figure 1F: 400x, synaptophysin immunostaining, showing diffuse positivity in tumor cells, consistent with neuroendocrine differentiation.

Discussion

- Esophageal small-cell NEC is a rare esophageal malignancy, accounting for 0.4-2.0% of esophageal cancers.
- Esophageal small-cell NEC are typically highly aggressive, frequently have metastasized by the time of diagnosis, and associated with poor prognosis.
- While surgical resection may be attempted for limited disease, those with metastases are typically managed with chemotherapy, with or without radiotherapy.
- Gastroenterologists should include rare esophageal malignancies, including small cell NEC, on their differential for new onset dysphagia to solids.