

Icahn School of Medicine at **Mount** Sinai

Small Cell Neuroendocrine Carcinoma of the Esophagus Presenting as Acute Hematemesis

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Introduction

Esophageal cancer often presents as dysphagia and weight loss and is diagnosed with the help of imaging and endoscopy. Esophageal carcinomas are most commonly squamous cell carcinoma or esophageal adenocarcinoma.

Gastrointestinal small cell neuroendocrine carcinomas account for 0.1 to 1% of all gastrointestinal malignancies and are most commonly noted in the small intestine, appendix, pancreas, and rectum.

We present a case of an 85 year old male with decreasing appetite and 1 day of hematemesis found to have high grade small cell neuroendocrine carcinoma of the esophagus with necrosis.

Case

An 85-year old male with a medical history of hypertension, dementia, and gastrointestinal bleed 5 years ago in the setting of NSAID use with an unremarkable EGD, presented to the hospital with 4 episodes of coughing up blood. Patient had a productive cough with white sputum for 1 week prior with decreased appetite and progressive weakness.

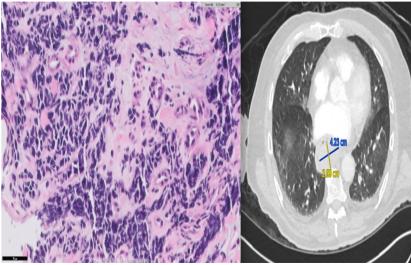


Image 1: Left: H&E stain of esophageal mass biopsy revealing tumor cells that are small with scant cytoplasm, salt and pepper chromatin, inconspicuous nucleoli, nuclear molding and smudging.

Right: CT chest revealing for 4.2 x 3.0 cm distal esophageal mass. There is circumferential wall thickening involving the mid and lower esophagus.

Case Continued

Labs were significant for down-trending hemoglobin from 12.4 to 10.7 on repeat labs.

CT pulmonary angiography and CT chest showed soft tissue thickening and a mass involving the proximal stomach and the gastroesophageal junction with thickening of the distal esophagus and large amount of fluid and debris (Figure 1). Enlarged surrounding lymph nodes and thickening of the distal aspect of the stomach were concerning for malignancy and metastatic disease.

Endoscopy showed a large partially obstructing bleeding esophageal mass with food debris proximal to the mass. Biopsy revealed high grade small cell neuroendocrine carcinoma with necrosis involving fibromuscular tissue (Figure 1).

Discussion

High grade neuroendocrine tumors can occur throughout the body, more commonly seen in the lungs, appendix, and small intestine. They can present as hemoptysis in patients with pulmonary involvement.

Hematemesis is an unusual presentation of esophageal cancer, and esophageal small cell neuroendocrine carcinomas are extremely rare with a poor prognosis.

Treatment modalities including chemotherapy, surgery, and radiation are selected based on staging of the disease.