

CASE REPORTS

Esophageal neuroendocrine carcinoma presenting as a rare cause of dysphagia

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ABSTRACT

A 60-year-old male with prior history of laryngeal carcinoma and active smoking presented with six months of solid food dysphagia. Endoscopy showed a large, friable gastroesophageal junction mass. Biopsies revealed a high-grade, poorly-differentiated neuroendocrine carcinoma. He was subsequently started on platinum based chemotherapy and radiation therapy and his tumor decreased dramatically in size. This case is unique as neuroendocrine carcinomas (NECs) are rarely found in the esophagus and usually have a poor prognosis at time of diagnosis.

Key Words: Neuroendocrine carcinoma, Endoscopy, Esophagus, Dysphagia

1. INTRODUCTION

Neuroendocrine carcinomas (NECs) of the esophagus are rare, aggressive, poorly-understood malignancies that may present as a rare cause of dysphagia. The prognosis is generally poor as the tumor is usually advanced at the time of presentation and diagnosis.

2. CASE PRESENTATION

A 60-year-old male presented with a six-month history of daily solid food dysphagia and a 10-pound unintentional weight loss. His past medical history was significant for stage 1 squamous cell carcinoma of the larynx diagnosed four years prior and treated with chemotherapy. He had a 30 pack-year smoking history and continued to smoke half a pack of cigarettes daily. His only medication was an ipratropium/albuterol 20-100 mg inhaler for chronic obstructive pulmonary disease. Endoscopy demonstrated a large villous

tumor occupying approximately 50% of the circumference of the distal third of the esophagus extending 3 cm proximal to the Z-line (see Figure 1). There was a pseudo-lumen within the tumor that led into the stomach, and on retroflexed view there was a 5 cm portion of ulcerated tumor extending into the cardia (see Figure 2). Biopsy results showed squamous and oxyntic-gastric mucosa containing sheets and packets of pleomorphic cells (see Figure 3). The cells demonstrated immunohistochemical positivity for synaptophysin, chromogranin and CD56. Immunohistochemistry for Ki-67 revealed a proliferation index of greater than 90%, classifying the tumor as a grade 3 or high-grade neuroendocrine carcinoma. Per the current National Comprehensive Cancer Network guidelines, this denotes a poor prognosis.^[1] Subsequent computed tomography revealed findings of a 7.1 cm × 6.8 cm × 4.8 cm gastroesophageal junction mass with prominent regional lymph nodes and without radiologic ev-

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idence of metastatic disease. The patient was treated with neoadjuvant chemotherapy consisting of cisplatin, etoposide, and dexamethasone and concurrent radiation therapy prior to consideration of surgical resection. Follow up endoscopy six months later showed dramatic reduction in the size of the tumor with near resolution (see Figures 4 and 5).

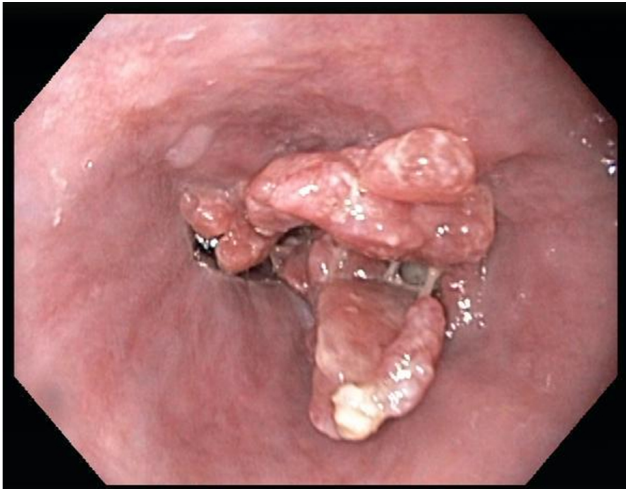


Figure 1. Villous tumor 3 cm proximal to z-line

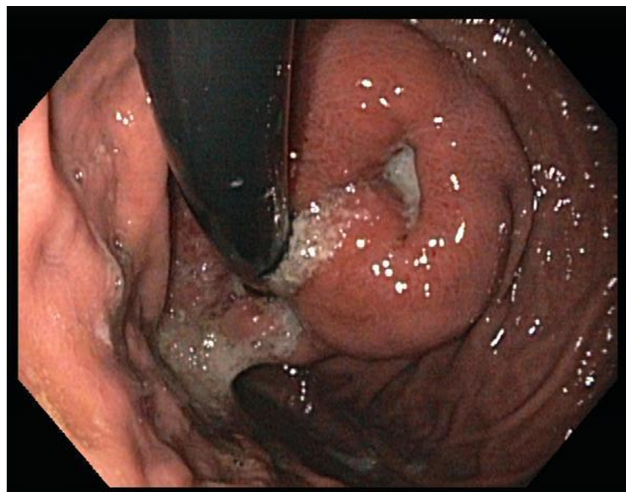


Figure 2. 5 cm ulcerated tumor extending into the cardia on retroflexed view

3. DISCUSSION

NECs are differentiated into two major categories: well-differentiated and poorly differentiated. Well-differentiated NECs are commonly known as carcinoids while poorly differentiated NECs typically appear in the lung as small or large cell carcinomas. Esophageal NECs are a very rare cause of dysphagia and represent only 0.4%-2.0% of all esophageal malignancies.^[2] Additional uncommon locations are the jejunum, ileum, bladder, cervix, and prostate.^[3]

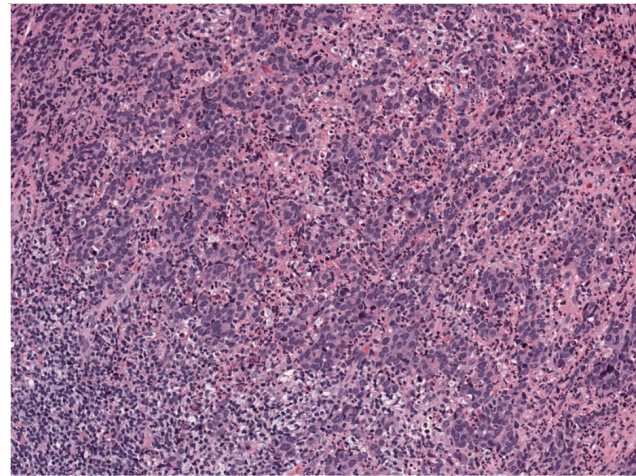


Figure 3. H&E stain demonstrating pleomorphic cells

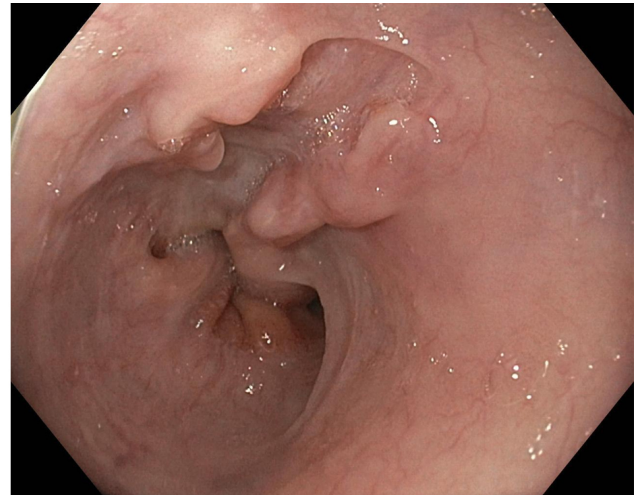


Figure 4. Marked reduction of tumor on upper endoscopy 6 months later

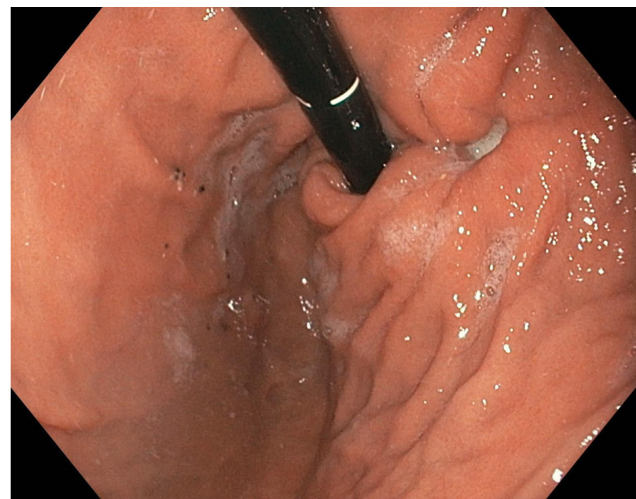


Figure 5. Marked reduction of tumor on upper endoscopy 6 months later on retroflexed view

Diagnostic criteria to classify a NECs as poorly-differentiated per the European Neuroendocrine Tumor Society along with the World Health Organization is defined as: positive endocrine markers such as chromogranin A, synaptophysin, CD56, > 20 per 10 high powered field mitotic figures, > 20% Ki-67% proliferation index.^[2,4]

At the time of diagnosis, the prognosis is considered poor, with median survivals of 34, 14, and 5 months for those with localized, regional, and distant disease respectively.^[5] Treatment is subsequently guided based on the classification of limited disease (LD) versus extended disease (ED).^[6] Those with LD are generally treated based on small cell lung carcinoma paradigm with platinum-based chemotherapy, cisplatin and etoposide. Chemoradiation has been found to improve efficacy of therapy in those with small cell lung cancer, but has yet to be thoroughly studied in those with esophageal NECs.^[7] However, there are those that would extrapolate utilizing chemotherapy with concurrent radiation in the setting of a highly aggressive malignancy. Surgical excision is rarely effective as a sole therapy, but has been used in combination with subsequent chemotherapy.^[8]

Esophageal NECs are still being characterized in their pathophysiology, presentation, and treatment; thus prompt recognition of symptoms is essential for diagnosis in this high mortality condition. Our patient responded to the current guideline directed therapy of chemotherapy and radiation based on small cell lung cancer practices and represents a valuable case for further understanding of a rare presentation.

CONFLICTS OF INTEREST DISCLOSURE

The authors declare that they have no competing interests.

CONSENT

Patient consent was obtained for case publication.

DISCLOSURES

The view(s) expressed herein are those of the author(s) and do not reflect the official policy or position of Brooke Army Medical Center, the U.S. Army Medical Department, the U.S. Army Office of the Surgeon General, the Department of the Army, the Department of the Air Force and Department of Defense or the U.S. Government.

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