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IMAGE | ESOPHAGUS

Primary Large Cell Neuroendocrine Carcinoma of the Esophagus Disguised as a Food Impaction

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CASE REPORT

A 50-year-old white man with a medical history of tobacco dependence presented with complaints of solid food dysphagia for 1.5 months with associated unintentional weight loss of 15 lbs and intractable hiccups. On arrival, he was hemodynamically stable. Physical examination was unremarkable. Laboratory testing revealed mild normocytic anemia of 12.3 g/dL and was otherwise unremarkable. Esophagogastroduodenoscopy initially revealed a large amount of meat in the midesophagus; however, after further manipulation, there appeared to be bleeding from the surface (Figure 1). A mass originating from the lateral esophageal wall was identified, caked with food particulate giving it the appearance of meat. The mass was traversed with no evidence of distal extension. Thoracic computed tomography (CT) showed a large heterogenous esophageal mass (8.1 × 5.7 cm) extending from the midesophagus to the gastroesophageal junction with no evidence of direct invasion (Figure 2). Biopsies returned positive for large cell neuroendocrine carcinoma (NEC) with diffuse synaptophysin and Ki67 positivity (>90%) (chromogranin/CD56 negative) (Figure 3). Outpatient positron emission tomography CT showed prominent perigastric lymphadenopathy and no other metastatic disease. Attempted endoscopic ultrasound failed due to technical difficulty. CT surgery subsequently performed a McKeown transthoracic esophagectomy for definitive therapy. Gross pathologic examination revealed a poorly differentiated NEC with positive lateral margins with invasion into

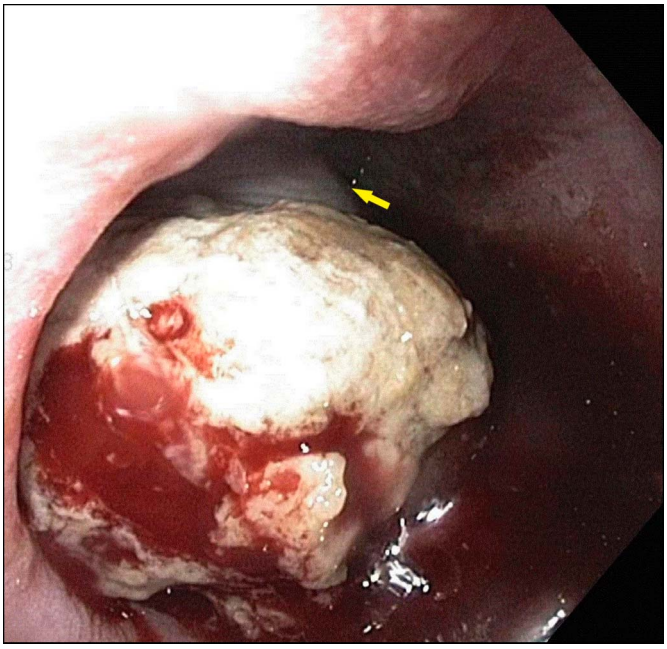

Figure 1. Endoscopic image of the esophageal lesion which appears like a food bolus with surface oozing originating from the lateral esophageal wall (arrow).

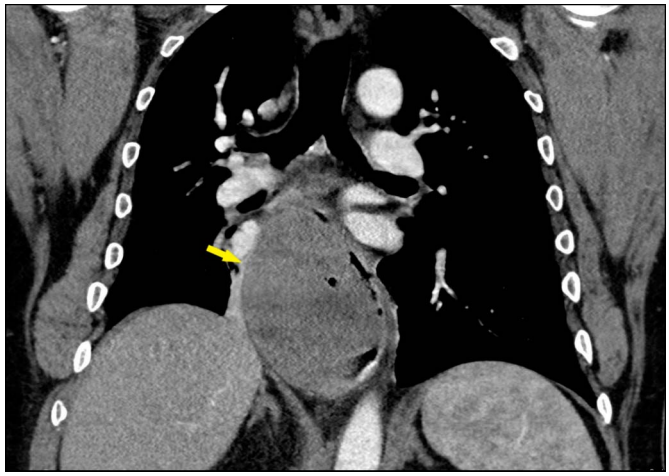

Figure 2. Coronal computed tomography showing a large heterogenous esophageal mass extending from the midesophagus to the gastroesophageal junction with no evidence of direct invasion (arrow).

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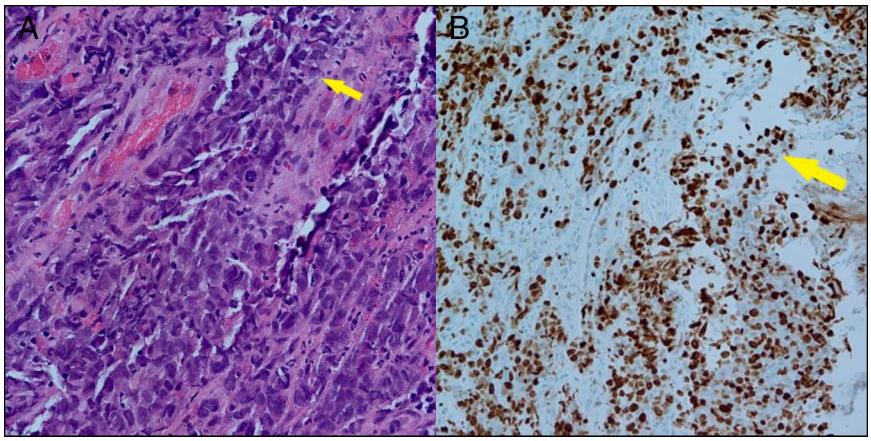

Figure 3. High-power histology image of the esophageal neuroendocrine carcinoma demonstrating (A) sheets and nest of highly atypical cells with intermediate to large nuclei, high nuclear-cytoplasmic ratios, variable chromatin with numerous mitoses, and abundant necrosis and apoptosis (arrow). (B) Histology image with Ki67 staining demonstrating a high proliferation rate (arrow).

the adventitia and to 4 of the 22 lymph nodes (pT3N2M0). He underwent adjuvant chemoradiotherapy with good response and no evidence of recurrent disease on surveillance imaging obtained 8 months postoperatively.

Esophageal NEC is a rare entity with a generally dismal prognosis when poorly differentiated and metastatic. It is reported that esophageal NECs represent only 0.4%–1.4% of the gastrointestinal lesions, most of which arise from the midesophagus.[1,2] A study of a large-volume, tertiary center reported approximately 40 cases over a 20-year period.[3] In a retrospective study of 14 patients with esophageal NEC, 11 of these cases showed protruding or localized type, 7 of the cases had a distant metastasis (primarily to the liver or bone) on presentation, 9 of the patients died within 17 months of receiving a combination of chemotherapy and/or radiotherapy, and 2 patients were noted to be alive for more than 12 months—both had limited disease and underwent surgery and/or chemotherapy.[4] There are limited data on the endoscopic and phenotypic appearance of large cell NEC of the esophagus, with few cases highlighting the inconsistent response to chemoradiotherapy.[5] This case is an example of an esophageal NEC with a unique endoscopic appearance and aggressive disease phenotype which has responded well to treatment.

DISCLOSURES

Author contributions: HD Patel wrote the manuscript, is a major contributor in writing the manuscript, and reviewed the literature. J. Beck wrote the manuscript and provided the images. A. Kataria reviewed the literature. GW Gross revised the manuscript for intellectual content. J. Echavarria edited and approved the final manuscript and is the article guarantor.

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