

Esophageal Neuroendocrine Carcinoma Presenting After Definitive Chemoradiation of Squamous Cell Carcinoma in the Same Location

Zarian Prenatt, DO, Hammad Liaquat, MD, Brittney Shupp DO, Lisa Stoll, MD, Yechezkel Schneider, MD – St. Luke's University Health Network, Bethlehem, PA

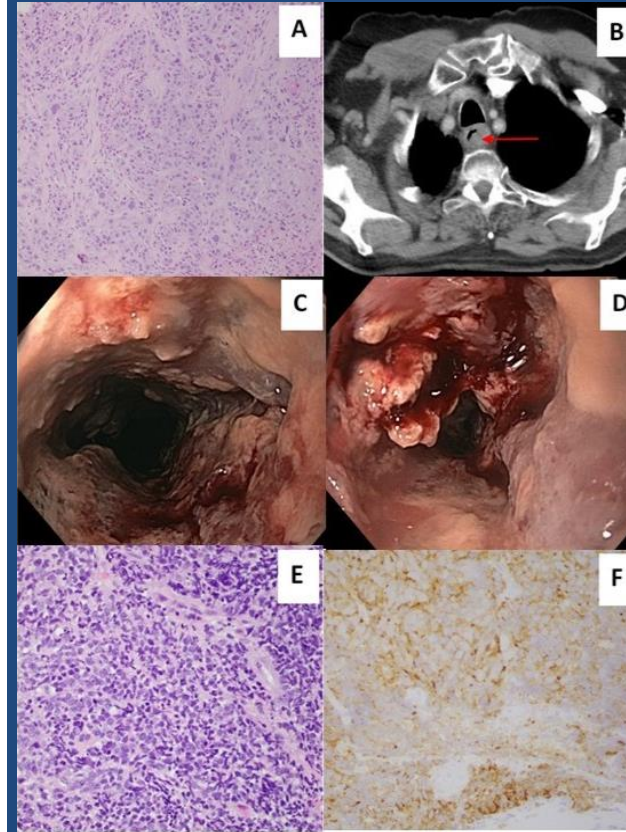
Introduction

- Esophageal neuroendocrine carcinoma (ENEC) is an extremely rare and highly aggressive malignancy that carries a poor prognosis.
- We present a rare case of ENEC presenting years after chemoradiation of squamous cell carcinoma (SCC) in the same location.

Case

- An 85-year-old male presented with difficulty swallowing.
- He had a history of SCC of the upper esophagus (A), for which he received chemoradiation with subsequent 3-year remission confirmed by serial imaging and endoscopy.
- A CT scan (B) showed soft tissue thickening in the upper esophagus at the location of prior SCC.
- EGD showed severe friable and ulcerated mucosa in the upper third of the esophagus (C & D).
- Biopsy revealed high-grade malignancy consistent with poorly differentiated neuroendocrine carcinoma, small cell type (E).
- Immunohistochemistry staining was positive for synaptophysin (F), pan-CK AE1/3, TTF1, and CD56. The Ki-67 mitotic proliferation index was 80-90%.
- He received chemotherapy with etoposide and carboplatin, but unfortunately had progression of his disease on imaging.
- He was found to have metastatic lesions in the scalp and passed away in hospice 14 months after diagnosis.

Images



(A) H&E stain showing moderately differentiated SCC. (B) CT chest showing soft tissue thickening (red arrow) in the proximal third of the esophagus compatible with recurrent tumor. (C, D) EGD showing friable, hemorrhagic, and ulcerated mucosa in the upper third of the esophagus. (E) Sheets of neuroendocrine carcinoma showing small blue cells with scant cytoplasm and nuclear molding. (F) Synaptophysin immunohistochemical stain showing diffuse cytoplasmic staining confirming neuroendocrine nature of the tumor.

Discussion

- To our knowledge, occurrence of ENEC in a site of prior esophageal SCC is a rarity.
- It could represent a case of radiation-induced malignancy (RIM) based on the temporal relation to his previous SCC treatment.
- Approximately 8% of second solid cancers may be related to radiation treatment, developing years after initial diagnosis and treatment of the first cancer.
- Unfortunately, management of ENEC is challenging because treatment strategies have not been well established due to the small number of cases reported in the literature.
- Patients and providers should discuss the possibility of developing secondary cancer from radiotherapy, and patients who have received radiation should be followed closely for RIM.

References

- Li Z, Hu J, Chen P, Zeng Z. Incidence, treatment, and survival analysis in esophageal neuroendocrine carcinoma population. *Transl Cancer Res.* 2020 Jul;9(7):4317-4329.
- Berrington de Gonzalez A, Curtis RE, Kry SF, Gilbert E, Lamart S, Berg CD, Stovall M, Ron E. Proportion of second cancers attributable to radiotherapy treatment in adults: a cohort study in the US SEER cancer registries. *Lancet Oncol.* 2011 Apr;12(4):353-60.