

De Novo Esophageal Small Cell Neuroendocrine Carcinoma in A Patient with Achalasia: A Novel Case Report

Rami Musallam, MD, Noor Aldeen Abdel Jalil, MS, Asem Abdeljalil, MD, Ala Abdel Jalil, MD

Background:

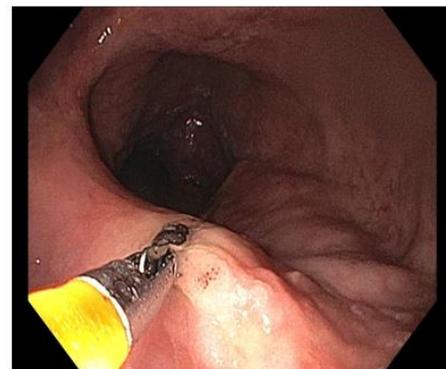
- Neuroendocrine carcinoma (NEC) of the esophagus is a rare and highly aggressive disease.
- Achalasia is a well-known condition associated with > 50 times higher risk of presenting with esophageal cancer than the general population.
- We present a novel case of small-cell NEC of the esophagus in a patient with longstanding achalasia who was treated using chemotherapy alone.

Case Description:

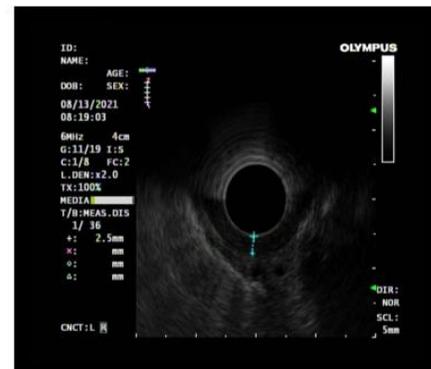
- An 81-year-old female with a history of longstanding achalasia presented for evaluation of progressive solid-food dysphagia for two months.
- EGD showed a partially obstructive fungating mass in the mid-esophagus.
- Mucosal biopsies demonstrated neoplastic cells with necrosis. IHC was consistent with high-grade neuroendocrine tumor, small cell subtype.
- A Chest CT scan revealed dilated distal esophagus with mass thickening along the left lateral side of the esophagus with an extension outside the esophagus.



EGD image shows a fungating mass in the mid esophagus, along with an abnormally dilated esophageal lumen from longstanding achalasia.



Repeat EGD shows resolution of the mid esophageal tumor post-treatment with chemotherapy.



An endoscopic ultrasound image shows an esophageal wall thickness of 2.5 mm post-treatment with chemotherapy.

Patient course:

- The patient received eight cycles of platinum plus etoposide chemotherapy.
- Four months later, the PET scan showed no evidence of hypermetabolic activity.
- The repeat endoscopy showed macroscopic resolution of the tumor, with histology showing some foci of high-grade dysplasia.
- Endosonography showed an esophageal wall thickness of 2.5 mm post-treatment.
- The patient's dysphagia resolved completely.

Conclusion:

- Neuroendocrine carcinoma (NEC) of the esophagus is rare.
- Achalasia is associated with an increased risk for malignancy, usually squamous cell type.
- To our knowledge, small cell NEC in the setting of achalasia has not been reported previously in the literature.
- Due to a paucity of cases of esophageal NEC, more studies are needed to understand the biological features of this aggressive cancer and its diagnostic and therapeutic approaches.