

Esophageal Manifestations of Paraneoplastic Pemphigus Vulgaris associated with Gastric Neuroendocrine Tumor

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Introduction:

- Pemphigus vulgaris (PV) typically affects the epidermis and oral mucosa, esophageal manifestations are uncommon.
- The primary symptoms accompanying this manifestation are dysphagia, odynophagia, and hematemesis.
- In this case report we examine the occurrence of esophageal pemphigus vulgaris associated with a gastric neuroendocrine tumor and note the endoscopic findings of esophagitis desiccans superficialis (EDS) within the esophagus.

Case Presentation:

- A 63-year-old female with a history of P-ANCA vasculitis presented for evaluation of dyspnea, hypotension, fatigue, rash [Figure 1a-c], and dysphagia.
- The patient was initially considered to have an infectious cause of these signs and symptoms, however, given a lack of response to antibiotic and antifungal therapy within 48 hours the differential was broadened, to include rheumatologic disease.
- The patient was treated with intravenous methylprednisolone and continued to experience dysphagia. When she failed to improve with the addition of corticosteroids gastroenterology was consulted.



Figure 1: Cutaneous manifestations of pemphigus vulgaris. a) Left Upper Extremity b) Right Upper Extremity c) Left Inguinal Region

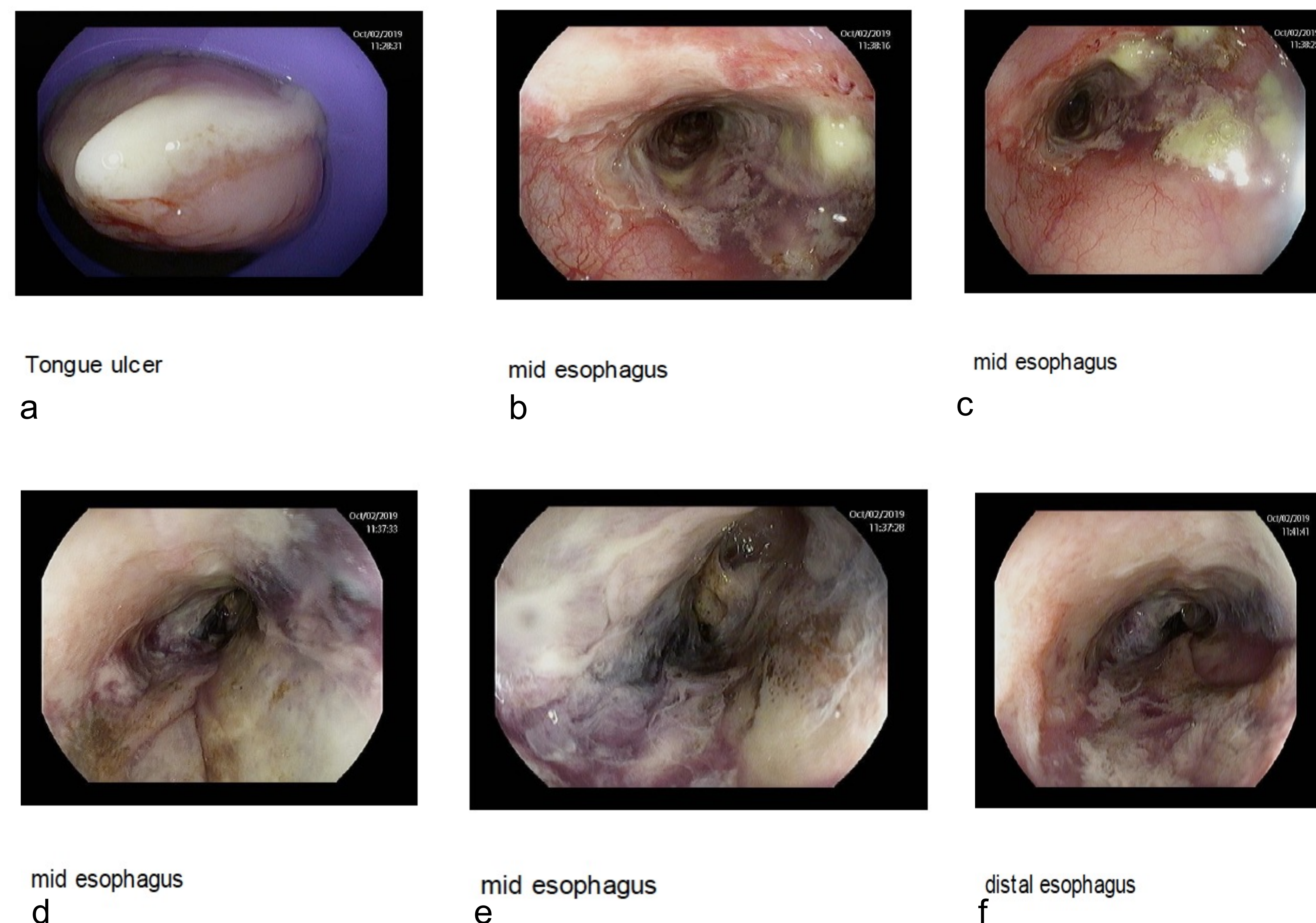


Figure 2[a-f]: EGD Findings with pseudomembranous appearance and subepithelial hemorrhage consistent with esophagitis desiccans.

Case Continued:

- EGD was performed and found tongue ulceration and desquamation in the distal esophagus with a pseudomembranous appearance [Figure 2a-f]. Extensive subepithelial hemorrhages were observed in the esophagus [Figure 2a-f], however, the gastric and duodenal mucosa were intact.
- Esophageal and gastric biopsies demonstrated staining for IgG reactive to desmoglein 3 consistent with PV. In addition, a well-differentiated neuroendocrine tumor (NET) in the stomach was identified.
- The patient was subsequently treated with octreotide for her neuroendocrine tumor, as well as solumedrol and dapsone for her skin lesions. She also received a loading dose of rituximab.

Discussion:

- PV involves autoimmune destruction of desmosomes via antibodies to desmoglein, causing suprabasal blistering, erosions, and ulcerations of epithelial and oral mucosa.
- Our patient demonstrated esophagitis desiccans, sloughing of the superficial esophageal mucosa, similar to the Nikolsky sign.