CLINICAL IMAGE

Esophagitis dissecans superficialis as the first manifestation of rectal adenocarcinoma

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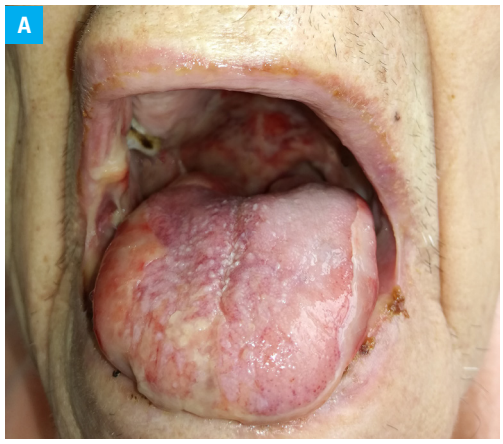
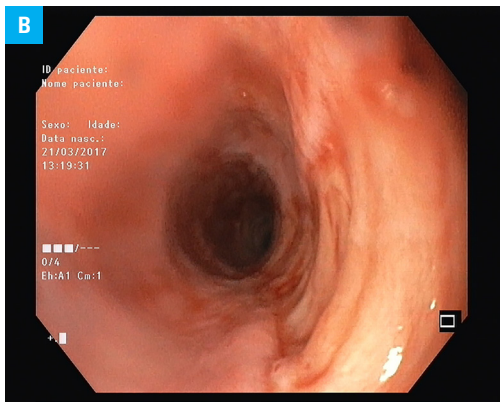
A 91-year-old woman presented with buccal bleeding, odynophagia, and epigastric pain aggravated by food intake. She reported an unintentional weight loss of 8% in the previous months but no other symptoms. Oral examination revealed hemorrhagic erosions and areas of desquamation in the tongue and buccal mucosa (FIGURE 1A). No ocular, cutaneous, or genital lesions were observed, and there was no history of caustic substance ingestion or the use of medication other than simvastatin and clopidogrel. The patient underwent esophagogastroscopy (FIGURE 1B and 1C), which showed erosions and sheets of sloughed mucosa from the incisors to the gastroesophageal junction, suggestive of esophagitis dissecans superficialis (EDS), later confirmed on histopathological examination (FIGURE 1D). Her medication was discontinued, and she was treated with pantoprazole, dexamethasone, and sucralfate. Direct immunofluorescence (DIF) staining was negative, with no observed reactants to immunoglobulins IgA, IgG, and IgM or complement C3. There was no clinical improvement and parenteral nutrition was necessary. Repeated endoscopy findings were identical; viral and fungal esophagitis were ruled out. A colonoscopy, prompted by acute hematochezia, was performed. A rectal mass was found, confirmed to be adenocarcinoma on pathological examination. Palliative care was provided. The patient died in the hospital, without regaining oral feeding.

EDS is a rare endoscopic finding characterized by sloughing fragments of necrotic esophageal





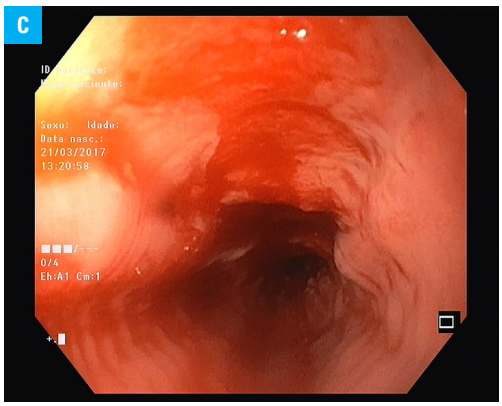


FIGURE 1 **A** – erosions and shallow ulcerations of the oral, soft palate, and tongue mucosa, extending to the vermillion surface of the lips, some covered by a yellowish pseudomembrane; **B** and **C** – esophagitis dissecans superficialis with an extensive reddish erosion of the entire esophagus and whitish sheets of sloughed mucosa

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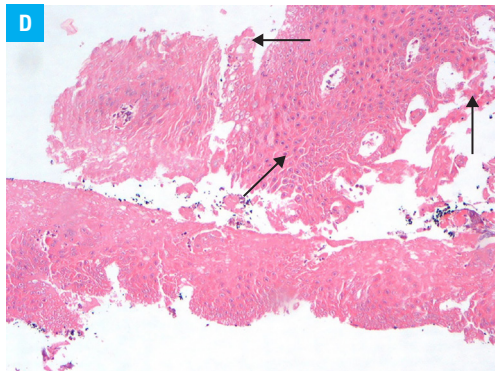

FIGURE 1
D – fragments of parakeratotic (horizontal arrow) and necrotic detached squamous esophageal epithelium with acantholysis (vertical arrow), spongiosis (diagonal arrow), and minimal lymphocytic inflammation (hematoxylin and eosin staining; magnification × 300)

epithelium.[1-3] It is frequently idiopathic; however, it has been associated with collagen diseases, physical and chemical trauma, autoimmune bullous dermatoses, and medications, such as bisphosphonates, clopidogrel, and anti-inflammatory drugs.[3] Paraneoplastic pemphigus (PNP) is an autoimmune mucocutaneous blistering disease associated with an underlying malignancy.[2] It manifests as diffuse erosions and shallow ulcerations of the oral mucosa.[4,5] The pharynx and esophagus may be affected, and there may be no cutaneous involvement.[4] Endoscopic features range from local erythema to EDS.[3] Adequate biopsy specimens are crucial as the prognosis relies on prompt diagnosis and treatment.

Most PNP cases are associated with hematologic malignancies, and only a few have been described in association with colorectal cancer.[4] Systemic corticosteroids are the first-line treatment. However, PNP rarely shows good response to steroids alone, and combination therapy with other immunosuppressants is often needed.[1,4] The prognosis is usually poor, not only due to the underlying malignant tumor but also due to infectious complications that frequently occur in these immunocompromised patients.[3-5]

Here, we report EDS as the first manifestation of rectal adenocarcinoma, presumably due to PNP. This association has been reported only a few times before. The patient fulfilled 2 major criteria (polymorphic mucosal eruption and concurrent neoplasia) and 1 minor criterion (acantholysis) for the diagnosis of PNP, as outlined by Camisa and Helm.[5] The other minor criterion, the deposition of immunoglobulins shown by DIF, was not met. Kaplan et al[4] reported DIF presentation months after clinical manifestation of the disease, so PNP cannot be excluded. Therefore, in the absence of a clear diagnosis for persistent oral or esophageal lesions in patients with malignancy, both hematologic and nonhematologic, a high index of suspicion is warranted and repeated tests may be needed to establish the final diagnosis.

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