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Visual Diagnosis in Emergency Medicine

PEMPHIGUS VULGARIS

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INTRODUCTION

We present the case of a 44-year-old woman with pemphigus vulgaris (PV) of 2 months' duration. She presented to an emergency department (ED) for evaluation of her condition where she was diagnosed with acute necrotizing ulcerative gingivitis (ANUG).

CASE REPORT

A 44-year-old woman presented to an oral medicine clinic for evaluation of oral lesions of 2 months' duration. After routine dental cleaning, she reported onset of painful, bleeding gingiva and mucosal ulcers accompanied by weight loss attributed to oral pain. She also reported cutaneous lesions on her neck and abdomen. The patient was evaluated in an ED for this complaint, diagnosed with ANUG, and was discharged without treatment with instructions to follow-up with her dentist. An incisional biopsy of the right buccal mucosa demonstrated acantholytic cells on routine analysis, suggestive of PV. Subsequently, she was prescribed a methylprednisolone dose pack and topical anesthetic rinse with temporary benefit.

Clinical examination revealed desquamation (peeling) of the gingiva (Figure 1) and erosive lesions affecting the buccal mucosa (Figure 2), ventral tongue, and soft palate. Erosions were also observed on the patient's neck (Figure 3). All findings were consistent with PV. The patient was prescribed dexamethasone solution 0.5 mg/5 mL, 10 mL swish/dwell/spit four times daily, Nystatin rinse 100,000 U, 5 mL swish/dwell/spit three times a day, and recommended to continue with topical anesthetic rinse, 10 mL swish/dwell/spit every 2 h as needed for symptom relief, with mild improvement of oral lesions. Serology revealed anti-desmoglein (DSG)-3 enzyme-linked immunosorbent assay (ELISA) 165 U and anti-DSG-1 ELISA 26 U, confirming a diagnosis of mucocutaneous PV. The patient consulted a dermatologist and rituximab was recommended for management due to worsening disease activity.

DISCUSSION

Pemphigus is a group of life-threatening, chronic, immune-mediated blistering diseases composed of five major entities: PV, pemphigus foliaceus, immunoglobulin A (IgA) pemphigus, paraneoplastic pemphigus, and drug-induced pemphigus (1). PV is the most common variant, with an incidence of 0.1 to 0.5 per 100,000 people per year, most often occurring in patients between 40 and 60 years of age, with a slight female predilection (2,3). In 80% of PV cases, the first signs of the disease are observed on the oral mucosa and lesions are frequently

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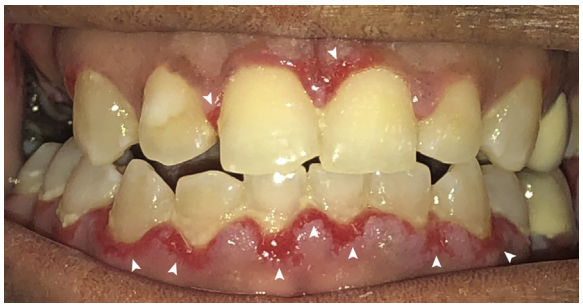

Figure 1. Epithelial desquamation affecting the maxillary and mandibular gingiva (arrows).

located on the gingiva, tongue, buccal mucosa, and palate. Desquamation of the gingiva may be the only clinical feature of PV and intraoral blisters often rupture quickly, leaving hemorrhagic, painful erosions with minimal inflammation (1). About 75% of PV patients also develop cutaneous lesions subsequent to oral manifestations, which may appear as erosions, vesicles, or bullae on normal-appearing or erythematous skin (1,4). A positive Nikolsky sign is associated with PV, which refers to blister formation or lateral extension of a blister with minor pressure or trauma (1,2). PV results from autoantibodies (IgG) directed against DSG, which is a desmosomal adhesion molecule, resulting in loss of cell-to-cell adhesion (1). DSG-1 is found in all layers of the epidermis, while DSG-3 is expressed in the parabasal and basal layers, therefore, patients with mucocutaneous PV have antibodies targeted against DSG-1 and DSG-3,

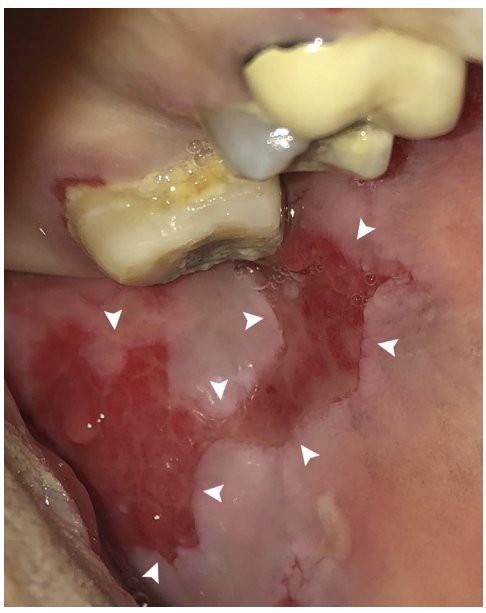

Figure 2. Superficial erosive lesions affecting the left buccal mucosa (arrows).

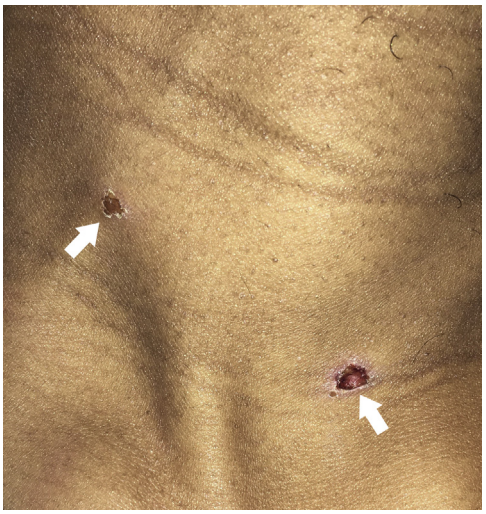

Figure 3. Erosions affecting the cutaneous surface of the neck (arrows).

whereas patients with PV limited to the mucosa have antibodies directed toward DSG-3 only (1).

Diagnosis of PV is based on clinical signs, histopathology, immunopathology, or serology. Characteristic histopathology of PV consists of intraepithelial separation, acantholysis, and a single layer of basal keratinocytes along the basement membrane, known as "tombstoning" (1). Direct immunofluorescence (DIF) analysis reveals intercellular deposition of IgG and occasionally Complement 3 (C3) between epithelial cells, commonly referred to as a "chicken-wire" pattern, in patients with PV (1,3). This distinguishes PV from other conditions with similar clinical presentations, such as mucous membrane pemphigoid and erosive lichen planus, as these conditions demonstrate different staining patterns on DIF analysis. Indirect immunofluorescence and ELISA are serologic studies used to detect circulating autoantibodies against epithelial cell surface antigens and may be used to establish diagnosis, guide therapy and determine prognosis of PV (2).

Management of PV depends on severity and distribution of the disease. Treatment for oral disease includes use of high and ultrahigh potency topical corticosteroids, intralesional corticosteroid injections, and topical tacrolimus (1,3). Severe mucous membrane involvement and concurrent cutaneous involvement may be more effectively managed with systemic immunomodulators, such as systemic corticosteroids (e.g., prednisone) and or steroid-sparing agents (e.g., mycophenolate mofetil) (1). Rituximab, a monoclonal antibody to $CD20^{+}$ B lymphocytes, has become a Food and Drug Administration–approved first-line treatment for PV (1). Topical anesthetic rinses, including viscous lidocaine and magic mouthwash are often effective for temporary symptom relief (3).

In this case, ANUG was considered as a diagnosis most likely due to the common clinical feature of painful, bleeding gingiva. ANUG is a form of periodontal disease confined to the gingiva with an acute clinical presentation characterized by rapid-onset interdental gingival necrosis, often described as “punched out” lesions, gingival pain, bleeding, and halitosis (5). Treatment of ANUG includes periodontal debridement, local oxygen therapy, and systemic antimicrobials to prevent sequelae and craters in soft tissues that will lead to new relapses (5). It is important for emergency physicians to distinguish PV from periodontal diseases to appropriately diagnose and manage the condition(s).

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