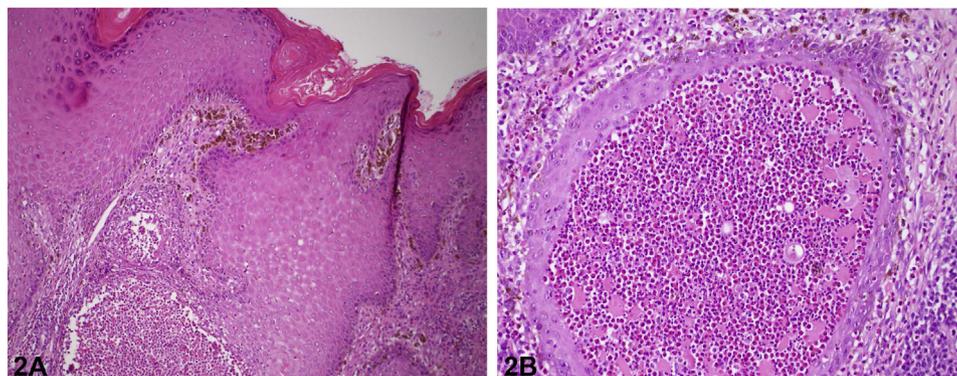


Dysphagia and history of vegetative plaques



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Key words: pemphigus vulgaris; pemphigus vegetans; desmoglein.



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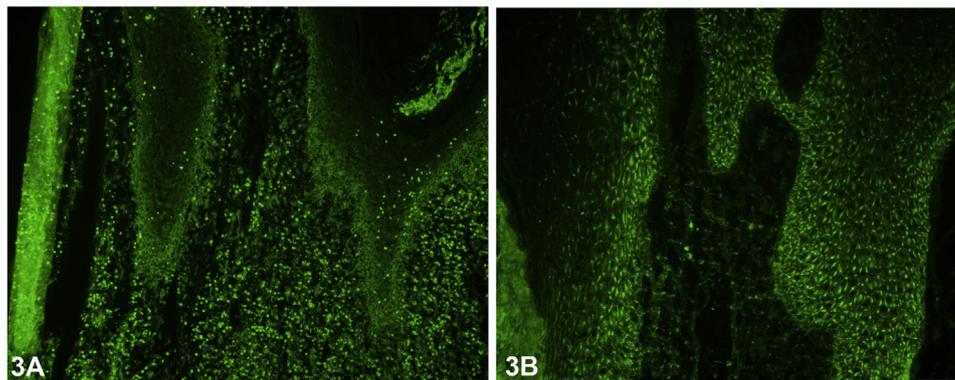
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A 40-year-old African-American woman with a documented history of hidradenitis suppurativa presented with hoarseness, dysphagia, and cough productive of blood and skin for 2 weeks. She describes a 12-year history of throat pain followed by vegetative plaques to the bilateral axilla (Fig 1) for which she had frequented the emergency department for pain management. Previous treatments include surgical excision, bleach baths, and antibiotics with minimal improvement. Trials of prednisone had intermittently alleviated her symptoms. Physical examination revealed oral mucosa without ulceration and hypertrophic surgical scars to the bilateral axilla. Laryngoscopy revealed inflammation of the vocal cord mucosa.

Question 1: What is the most likely diagnosis?

- A. Vegetative pyoderma gangrenosum (VPG)
- B. Keloids
- C. Hidradenitis Suppurativa (HS)
- D. Pemphigus vegetans
- E. Inverse psoriasis (IP)

Answers:

A. VPG — Incorrect. VPG is a rare pyoderma gangrenosum variant commonly found on the trunk as erythematous, ulcerated, vegetative plaques. Although vegetative, our patient’s prior lesions lack ulceration and were located on intertriginous areas. Additionally, VPG would not explain her mucosal involvement.

B. Keloids — Incorrect. Keloids are caused by cutaneous injuries such as trauma, insect bites, and piercings. Our patient’s hypertrophic scarring from surgery complicates her presentation, but her laryngeal involvement and previous vegetative intertriginous plaques suggest another diagnosis.

C. HS — Incorrect. HS is a chronic condition characterized by inflammation of the follicular epithelium, causing abscess, scar, and sinus tract formation. Our patient was unfortunately misdiagnosed and surgically treated for HS. The patient’s laryngeal involvement and history of vegetative skin lesions suggest another diagnosis.

D. Pemphigus vegetans — Correct. Pemphigus vegetans is the rarest (1% to 2%)¹ subtype of pemphigus vulgaris and is distinguished by the presence of papillomatous, vegetating plaques in flexural surfaces. It initially manifests with oral ulceration in 50% to 70% of cases, and 90% of patients ultimately develop oral involvement.² Although our patient previously presented with intertriginous vegetative plaques, she presented to us with isolated laryngeal involvement. Histology of previously biopsied axillary lesions revealed epidermal hyperplasia and acantholytic keratinocytes present within eosinophil-rich microabscesses (Fig 2).

E. IP — Incorrect. IP is a form of plaque psoriasis that involves axillary, anogenital, and inframammary body folds. Although our patient’s lesions are in intertriginous areas, they lack the well-demarcated, erythematous patches seen in IP.

Question 2: Biopsy and enzyme-linked immunosorbent assay panel testing confirm the diagnosis. Direct immunofluorescence (DIF) would most likely reveal...

- A. IgG ± C3 in netlike distribution in the lower epidermis
- B. IgA in linear distribution along DEJ
- C. IgA in granular deposition in dermal papillae
- D. C3 > IgG in linear distribution at DEJ

E. IgM, IgG, IgA, and C3 in linear distribution at DEJ

Answers:

A. IgG ± C3 in netlike distribution in the lower epidermis — Correct. In pemphigus vulgaris/vegetans, IgG (predominantly IgG4) autoantibodies against desmoglein 1 and/or 3 disrupt intraepidermal adhesion and lead to vesicle, blister, and erosion formation on the skin and mucous membranes.³ The mucocutaneous variant has antibodies to Dsg3. Our patient's laboratory findings revealed significant desmoglein-3 antibodies (titer, 179.7). Antibody titer correlates with disease activity and titers are expected to decrease with clinical improvement. Our patient's DIF stained positive for IgG4 and C3 in a netlike distribution (Fig 3).

B. IgA in linear distribution along DEJ — Incorrect. This DIF staining pattern is seen in linear IgA bullous dermatosis, not pemphigus vegetans.³

C. IgA in granular deposition in dermal papillae — Incorrect. This DIF staining pattern is seen in Dermatitis herpetiformis, not pemphigus vegetans.³

D. C3 > IgG in linear distribution at DEJ — Incorrect. This DIF staining pattern is seen in herpes gestationis, not pemphigus vegetans.³

E. IgM, IgG, IgA, and C3 in linear distribution at DEJ — Incorrect. This DIF staining pattern is seen in bullous lupus, not pemphigus vegetans.³

Question 3: What feature of this patient's presentation is least expected for her diagnosis?

- A.** Improvement with corticosteroids
- B.** Intertriginous involvement
- C.** Isolated throat involvement
- D.** Presentation in a 40-year-old woman
- E.** Mucosal involvement

Answers:

A. Improvement with corticosteroids — Incorrect. First-line therapy for pemphigus vegetans consists of systemic corticosteroids and immunosuppressant agents (ie, rituximab). Without treatment, epidermal barrier function is compromised leading to fluid and protein loss, malnutrition, and secondary infections, which can progress to septic shock. Once treatment is initiated, patients require lifelong monitoring for both disease relapse and detrimental sequelae of prolonged immunosuppressive therapy.

B. Intertriginous involvement — Incorrect. Pemphigus vegetans is distinguished from pemphigus vulgaris by the presence of papillomatous, vegetating plaques in intertriginous surfaces.

C. Isolated throat involvement — Correct. While oral mucosa involvement occurs in 88% of pemphigus vegetans cases, patients seldom present with lesions to one site, and laryngeal involvement is rarely described.⁴ Our patient's isolated laryngeal involvement in biopsy-proven pemphigus vegetans demonstrates a unique presentation that requires immediate medical intervention.

D. Presentation in a 40-year-old woman — Incorrect. Pemphigus vegetans predominantly affects middle-aged women.

E. Mucosal involvement — Incorrect. Oral mucosa is almost invariably involved in pemphigus vegetans, hindering oral intake and often compromising patients' nutritional status. Mucosal lesions most commonly involve the buccal mucosa, lips, and gingivae, but can extend to the conjunctiva, pharynx, larynx, esophagus, vagina, penis, and anus.⁵

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Abbreviations used:

VPG: vegetative pyoderma gangrenosum
HS: hidradenitis Suppurativa
IP: inverse psoriasis
DIF: direct immunofluorescence

Conflicts of interest

None disclosed.

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