

# Solitary Exophytic Plaque on the Left Groin

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A 40-year-old otherwise healthy man presented with an exophytic plaque on the left groin of 1 month's duration. The lesion reportedly emerged as pustules that slowly expanded and coalesced. At an outside institution, cryotherapy was planned for a presumed diagnosis of condyloma acuminatum; however, the patient decided to get a second opinion. He denied recent intake of new drugs. Six months prior he had traveled to China and engaged in unprotected sexual intercourse. Physical examination revealed an approximately 4×2-cm exophytic plaque with a partially eroded and exudative surface on the left inguinal fold. Dermatologic examination, including the oral mucosa, was otherwise normal. Complete blood cell count and sexually transmitted disease panel were unremarkable.

## WHAT'S THE DIAGNOSIS?

- a. granuloma inguinale
- b. Kaposi sarcoma
- c. pemphigus vegetans
- d. pyodermatitis-pyostomatitis vegetans
- e. syphilitic chancre

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The authors report no conflict of interest.

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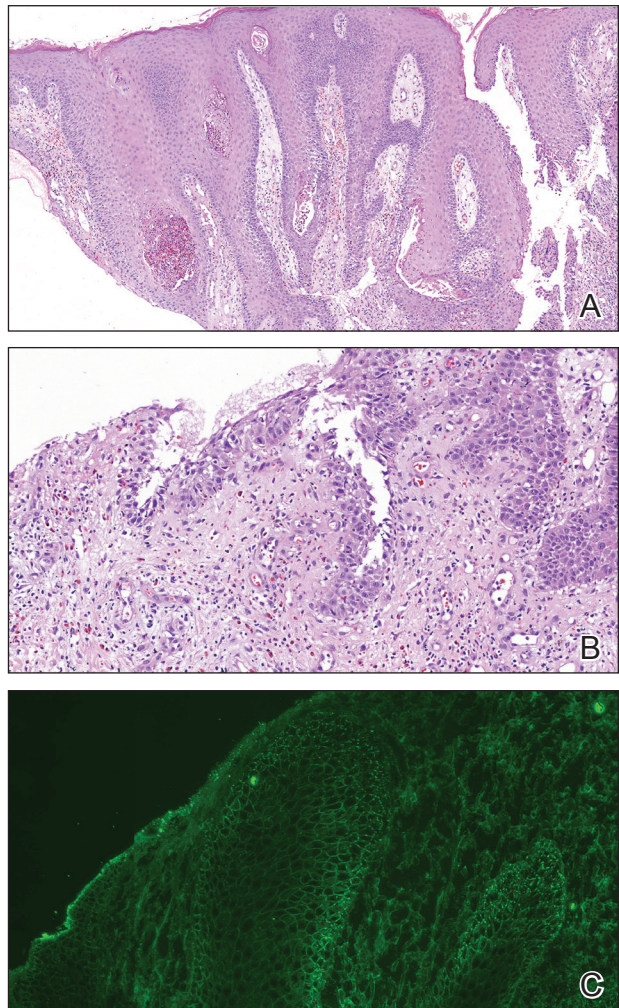
## THE DIAGNOSIS: Pemphigus Vegetans

A punch biopsy was taken from the verrucous plaque, and microscopic examination demonstrated prominent epidermal hyperplasia with intraepidermal eosinophilic microabscesses and a superficial dermatitis with abundant eosinophils (Figure 1A). Suprabasal acantholytic cleft formation was noted in a focal area (Figure 1B). Another punch biopsy was performed from the perilesional skin for direct immunofluorescence examination, which revealed intercellular deposits of IgG and C3 throughout the lower half of the epidermis (Figure 1C). Indirect immunofluorescence performed on monkey esophagus substrate showed circulating intercellular IgG antibodies in all the titers of up to 1/160 and an elevated level of IgG antidesmoglein 3 (anti-Dsg3) antibody (enzyme-linked immunosorbent assay index value, >200 RU/mL [reference range, <20 RU/mL]).

Because there was a solitary lesion, the decision was made to perform local treatment. One intralesional triamcinolone acetonide injection (20 mg/mL) resulted in remarkable flattening of the lesion (Figure 2). Subsequently, treatment was continued with clobetasol propionate ointment 3 times weekly for 1 month. During a follow-up period of 2 years, no signs of local relapse or new lesions elsewhere were noted, and the patient continued to be on long-term longitudinal evaluation.

Pemphigus vegetans (PV) is an uncommon variant of pemphigus, typically manifesting with vegetating erosions and plaques localized to the intertriginous areas of the body. Local factors such as semioclusion, maceration, and/or bacterial or fungal colonization have been hypothesized to account for the distinctive localization and vegetation of the lesions.<sup>1,2</sup> Traditionally, 2 clinical subtypes of PV have been described: (1) Hallopeau type presenting with pustules that later evolve into vegetating plaques, and (2) Neumann type that initially manifests as vesicles and bullae with a more disseminated distribution, transforming into hypertrophic masses with erosions.<sup>1-5</sup> However, this distinction may not always be clear, and patients with features of both forms have been reported.<sup>2,5</sup>

At present, our case would best be regarded as a localized form of PV presenting with a solitary lesion. It may progress to more disseminated disease or remain localized during its course; the literature contains reports exemplifying both possibilities. In a large retrospective study from Tunisia encompassing almost 3 decades, the majority of the patients initially presented with unifocal involvement; however, the disease eventually became multifocal in almost all patients during the study period, emphasizing the need for long-term follow-up.<sup>2</sup> There also are reports of PV confined to a single anatomic site, such as the scalp, sole, or vulva, that remained localized for years.<sup>2,4,6,7</sup> Involvement of the oral mucosa is an important



**FIGURE 1.** Histopathology revealed prominent epidermal hyperplasia with intraepidermal eosinophilic microabscesses. Note dermal edema, vascular proliferation, and eosinophil-rich infiltration (A)(H&E, original magnification  $\times 40$ ). Suprabasal acantholytic blister and prominent eosinophilic spongiosis was noted (B)(H&E, original magnification  $\times 200$ ). Direct immunofluorescence revealed IgG deposition in the intercellular area of the epidermis (C)(original magnification  $\times 100$ ).

finding of PV and the presenting concern in approximately three-quarters of patients.<sup>2</sup> Interestingly, the oral mucosa was not involved in our patient despite the high titer of anti-Dsg3 antibody, which suggests the need for the presence of other factors for clinical expression of the disease.

Although PV is considered a vegetating clinicomorphologic variant of pemphigus vulgaris, PV is histopathologically distinguished from pemphigus vulgaris by the presence of epidermal hyperplasia and intraepidermal eosinophilic microabscesses. Importantly, the epidermis



**FIGURE 2.** Remarkable flattening of the lesion was noted 2 weeks after one intralesional triamcinolone injection.

displays signs of exuberant proliferation such as pseudoepitheliomatous hyperplasia and/or papillomatosis of a varying degree.<sup>1,2,5</sup> Of note, suprabasal acantholysis is usually overshadowed by the changes in PV and presents only focally, as in our patient. The most common autoantibody profile is IgG targeting Dsg3; however, a spectrum of other autoantibodies has been identified, such as IgG antidesmocollin 3, IgA anti-Dsg3, and IgG anti-Dsg1.<sup>8,9</sup>

The most important differential diagnosis of PV is pyodermitis-pyostomatitis vegetans. These 2 entities share many clinical and histopathological

features; however, direct immunofluorescence is helpful for differentiation because it generally is negative in pyodermitis-pyostomatitis vegetans.<sup>2,10</sup> Furthermore, there is a well-established association between pyodermitis-pyostomatitis vegetans and inflammatory bowel disorders, whereas PV has anecdotally been linked to malignancy, human immunodeficiency virus infection, and heroin abuse.<sup>1,2,10</sup> Our patient was seronegative for human immunodeficiency virus and denied weight loss or loss of appetite. For those cases of PV involving a single anatomic site, the differential diagnosis is broader and encompasses dermatoses such as verrucae, syphilitic chancre, condylomata lata, granuloma inguinale, herpes simplex virus infection, and Kaposi sarcoma.

Treatment of PV is similar to pemphigus vulgaris and consists of a combination of systemic corticosteroids and steroid-sparing agents.<sup>1,5</sup> On the other hand, more limited presentations of PV may be suitable for intralesional treatment with triamcinolone acetonide, thus avoiding potential adverse effects of systemic therapy.<sup>1,2</sup> In our case with localized involvement, a favorable response was obtained with intralesional triamcinolone acetonide, and we plan to utilize systemic corticosteroids if the disease becomes generalized during follow-up.

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