

Multiple vegetating lesions over the genitalia

A 41-year-old widow presented with multiple, asymptomatic, raised lesions over the genitalia for 3 months. The lesions had slowly increased in number and extent. There was no history of any fluid-filled lesion or any genital ulcer in the past. She denied any history of extramarital sexual contact. No other family member had similar lesions. On examination, multiple grayish, moist, vegetating papules and plaques of varying sizes were present on the labia majora, perianal area, and groins [Figure 1]. There were erythematous

eroded areas in the oral cavity with candidiasis. Random blood sugar, venereal disease research laboratory (VDRL) test, *Treponema pallidum* hemagglutination (TPHA), HIV spot test, and serum IgM for herpes simplex virus were done. Dark ground illumination (DGI) was performed; Tzanck smear was also taken from the eroded lesion. A 4-mm punch biopsy was taken from the lesion over the right labia majus [Figure 2].

WHAT IS YOUR DIAGNOSIS?

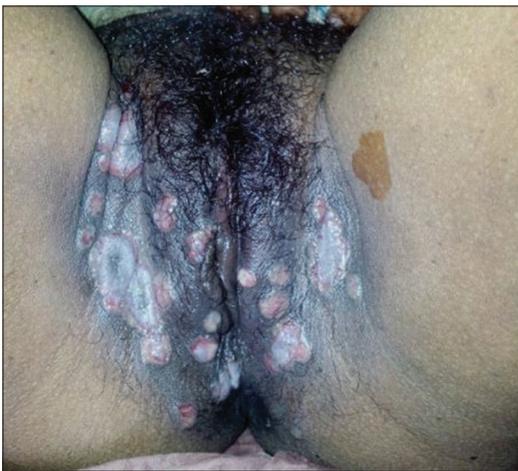


Figure 1: Multiple moist, vegetating papules and plaques on the labia majora, groin and the perianal region

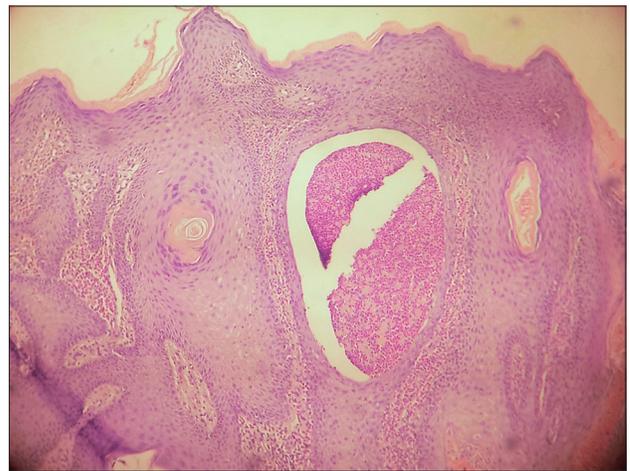


Figure 2: Eosinophils in the epidermis and the dermis

How to cite this article: Saldanha CS, Shenoy MM, Shanthala PR, Amin VB. Multiple vegetating lesions over the genitalia. Indian J Dermatol Venereol Leprol 2015;81:325-6.

Received: March, 2014. **Accepted:** October, 2014. **Source of Support:** Nil. **Conflict of Interest:** None declared.

ANSWER

Pemphigus vegetans

Histopathology

Skin biopsy shows acanthosis and intraepidermal eosinophilic microabscesses. Acantholytic cells were difficult to see. Dermis showed an intense inflammatory infiltrate predominantly comprising eosinophils (H and E, x400).

DISCUSSION

Pemphigus vegetans is a rare clinical variant of pemphigus vulgaris characterized by vegetating plaques in the skin folds and comprises up to 5% of all pemphigus cases.^[1] Two clinical subtypes have been described; the Neumann type usually begins with bullae, has a poor response to therapy and a poor prognosis, similar to pemphigus vulgaris, whereas the Hallopeau type usually begins with grouped pustules and has an excellent response to therapy with many patients going into prolonged remission.^[2,3] Pemphigus vegetans tends to develop in areas of relative occlusion and maceration with subsequent bacterial infection suggesting a response to superinfection.^[4] The histopathology of pemphigus vegetans reveals epidermal hyperplasia, papillomatosis and intraepidermal eosinophilic abscesses as the lesions age, which differs from pemphigus vulgaris.^[5]

In this case, vesiculo-pustular lesions were not apparent clinically and there were no blisters elsewhere on the body. The diagnosis was made based on the clinical morphology of vegetating papules and plaques in the groin and perianal region, erosions in the oral mucosa, and the characteristic histopathological findings. Tzanck smear also showed eosinophils and acantholytic cells. The diagnosis was further confirmed by direct immunofluorescence (DIF) findings of an intercellular pattern in the epidermis [Figure 3]. Condyloma lata, which also presents as flesh-colored, moist, oozing papules in the intertriginous areas was excluded by negative dark field microscopy and non-reactive VDRL and TPHA tests. Genital herpes in immunocompromised cases can also present with vegetative lesions but the characteristic histopathology was lacking.

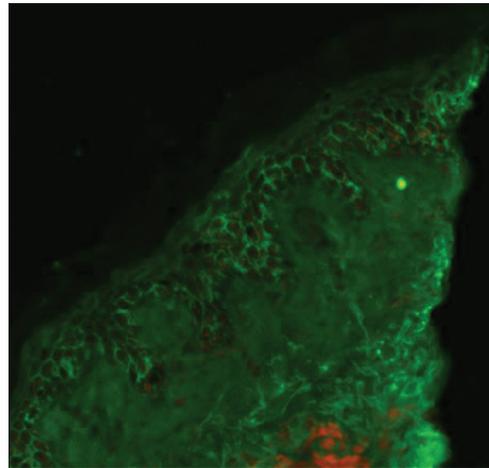


Figure 3: Direct immunofluorescence showing deposits in the intercellular spaces in the epidermis (x400)

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Quick Response Code:	Website: www.ijdv.com
	DOI: 10.4103/0378-6323.155563