Case Report

Verrucous lesions over external genitalia as a presenting feature of pemphigus vegetans

Ankush A. Ajbani, Kajal S. Mehta, Yogesh S. Marfatia
Department of Dermatology, Venereology and Leprosy, SSG Hospital and Government Medical College Baroda, Vadodara, Gujarat, India

Address for correspondence:
Dr. Yogesh S. Marfatia, OPD-1, Department of Dermatology, Venereology and Leprosy, SSG Hospital, Vadodara, Gujarat, India.
E-mail: ym11256@gmail.com

Abstract

Pemhigus vegetans is a rare variant of pemphigus vulgaris which primarily occurs in the flexures of the body. We report a case of pemphigus vegetans in an 85-year-old female presenting with hypertrophic verrucous lesions over external genitalia and perianal region. There was no history of preceding oral lesions. The diagnosis of pemphigus vegetans was considered on the clinical ground and confirmed by histopathological examination.

Key words: Nonvenereal genital dermatoses, pemphigus, pemphigus vegetans

INTRODUCTION

Pemphigus vulgaris is a chronic vesiculobullous disorder with various phases or relapses and remittances.[1] One of the variant of pemphigus vulgaris is pemphigus vegetans, occurring in l%–5% of the patients.[2] It is most commonly caused by the production of autoantibodies against intercellular adhesion proteins desmoglein 1 and desmoglein 3.[3,4] Due to its rarity, it is often misdiagnosed and poses a diagnostic challenge if it occurs on locations seemingly uncommon. We report a case of pemphigus vegetans in an 85-year-old woman presenting to the dermatology outpatient department (OPD) with verrucous hypertrophic growth over the genitalia and perianal region.

CASE REPORT

An 85-year-old female patient presented to the dermatology OPD with the complaints of itchy growth over the external genitalia, perianal region, and left axilla for the past 4 months. It was also associated with painful defecation. She also developed thickening and crusting of the lips and fissuring of the tongue leading to intolerance to spicy food for the past 2 months. On detailed history taking, initially, fluid-filled lesions were present over the same sites which easily ruptured, leaving behind erosions. Subsequently, hypertrophic and verrucous lesions appeared over the external genitalia and perianal region. There was a past history of the appearance of similar lesions 20 years back at similar sites. She was diagnosed as a case of condyloma lata and treated with injection benzathine penicillin 3 months back. As there was no response, she was referred to our OPD. She was a chronic tobacco chewer.

On cutaneous examination, she had hyperpigmented, dry, verrucous, nontender plaques over the genitalia extending to the groin [Figure 1], perianal region [Figure 2], and the left axilla [Figure 3]. There were palpable bilateral discrete inguinal lymph nodes, firm in consistency. The lips were thickened along with fissuring and dry, adherent crustations.

Access this article online

Quick Response Code:

Website: www.ijstd.org

DOI: 10.4103/ijstd.IJSTD_80_19

How to cite this article: Ajbani AA, Mehta KS, Marfatia YS. Verrucous lesions over external genitalia as a presenting feature of pemphigus vegetans. Indian J Sex Transm Dis 2019;40:176-9.

Figure 1: Verrucous vegetating lesions on the vulva and groin at the time of presentation

Figure 2: Lesions around the anus and perineum at the time of presentation

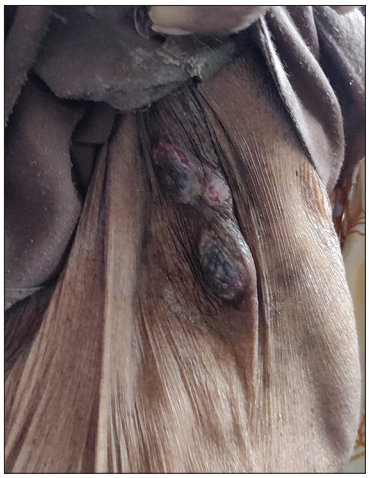

Figure 3: Verrucous hypertrophic lesion in left underarm at the time of presentation

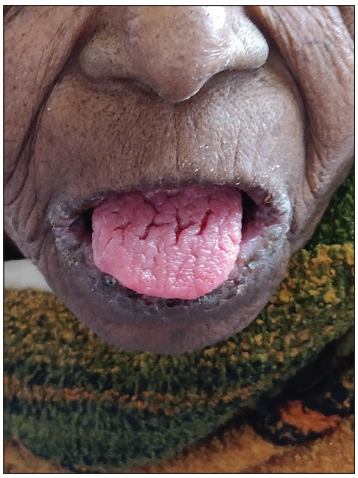

Figure 4: Thickening and fissuring of the lips and tongue at the time of presentation

The tongue was enlarged with deep dorsal fissuring, suggestive of the cerebriform tongue [Figure 4]. The oral mucosa was pale. Systemic examination was within the normal limits. Complete blood count revealed hemoglobin level of 9.8 g/dL. Serum HIV and (RPR) Rapid Plasma Reagin were nonreactive. Local inguinal ultrasonography confirmed the presence of enlarged lymph nodes bilaterally. Punch biopsies were taken from lesions over the right inguinal region and axilla. Differentials considered included pyoderma vegetans, condyloma acuminata, pemphigus vegetans, and malignancy. Pus culture and sensitivity taken from the lesion through a sterile swab stick showed no growth at the end of 7 days.

Histopathological examination showed superficial and mid-perivascular and interstitial infiltrate of lymphocytes, neutrophils, and eosinophils with pseudocarcinomatous hyperplasia of the epidermis. Hyperplastic epidermis showed focal spongiosis with neutrophils and eosinophils, with suprabasal clefts at places with scant acantholysis [Figures 5 and 6]. The diagnosis of pemphigus vegetans was thus confirmed.

She was initially treated with oral prednisolone (30 mg) once a day, injectable cefotaxime (1 g) intravenous (IV) 8 hourly, and injectable metronidazole (400 mg) IV 8 hourly. Subsequently, prednisolone was tapered 5 mg every 15 days. Hematinics were given to treat the anemia. Topically, she was treated with clobetasol propionate (0.05%) and framycetin sulfate creams. On follow-up after 6 weeks, she improved markedly in the form of a reduction in the thickness of the lesions [Figure 7] and decrease in the intensity of pruritus. Significant improvement was present in the lesions over the lips and tongue [Figure 8].

DISCUSSION

Pemphigus vegetans is a relatively rare variant of pemphigus vulgaris, commonly with verrucous,

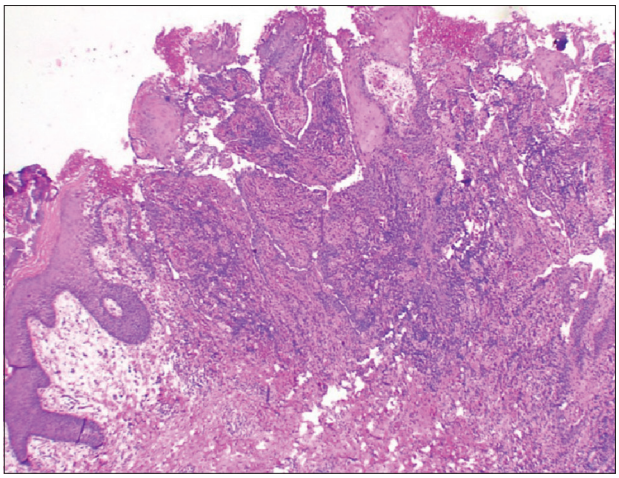

Figure 5: Hematoxylin and eosin stained cut section at ×4 magnification

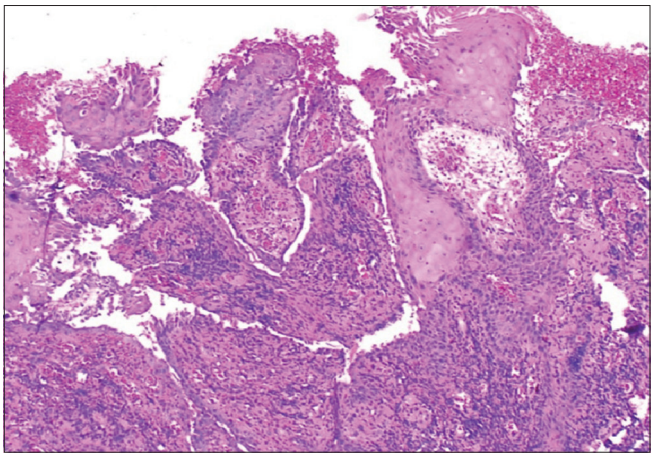

Figure 6: Histopathological examination showing superficial and mid-perivascular and interstitial infiltrate of lymphocytes, neutrophils, and eosinophils with pseudocarcinomatous hyperplasia of the epidermis, along with hyperplastic epidermis showing focal spongiosis with neutrophils and eosinophils, with supra-basal clefts at places with scant acantholysis. (×10 magnification)

Figure 7: View of the groin and vulva at 6-week follow-up, showing significant improvement

Figure 8: View of the lips and tongue at 6-week follow-up, showing significant improvement

cauliflower-like growth in the flexures of the body, and the periorificial regions, although cases with isolated involvement of the arms, feet, fingers, breasts, and oral cavity have been reported.[2,5,6] In this case, verrucous lesion over the external genitalia was presenting a feature of pemphigus vegetans and then involved the perianal region, axilla, and lips. In pemphigus vegetans, there is the development of antibodies against desmoglein 3 and/or desmoglein 1 along with antibodies against desmocollin 1, 2, and 3.[7,8]

Clinically, it is classified into two types – the Neumann type and the Hallopeau type.[9] In the Neumann type, the lesions begin as the classical lesions of pemphigus vulgaris and then form heaped-up vegetative plaques. In the Hallopeau type, the lesions begin as pustular lesions in groups, later leading to the formation of the heaped-up plaques. The Hallopeau type has a relatively benign course and responds to a lower dose of steroids and has prolonged remission, while the Neumann type needs higher doses of systemic corticosteroids and is resistant to treatment.[3,9] In our patient, benign course and good response to steroids were in favor of the diagnosis of Hallopeau type. Differential diagnoses of the lesions include vegetating herpes simplex infection, acrodermatitis continua suppurativa, pyoderma gangrenosum, pyoderma vegetans, Bowen's disease, squamous cell carcinoma, paraneoplastic pemphigus, epidermolysis bullosa acquisita, and pyodermatitis-pyostomatitis vegetans.[5,10] Positive serum anti-desmoglein 3 and immunofluorescence staining of the biopsy should be done to have a gold standard diagnosis, although it was not done in our case due to the nonavailability of antibody detection and staining method in the institute. In the present case, the diagnosis was confirmed by histopathological examination and

response to systemic and topical corticosteroids. This case was unique, as the presenting feature of pemphigus vegetans was verrucous lesions over the external genitalia and then extending to the perianal region, groin, axilla, and lips.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Basu K, Chatterjee M, De A, Sengupta M, Datta C, Mitra P, *et al.* A clinicopathological and immunofluorescence study of intraepidermal immunobullous diseases. Indian J Dermatol 2019;64:101-5.
2. Khullar G, De D, Narang T, Saikia UN, Handa S. Pemphigus vegetans localized to unusual sites. Indian J Dermatol Venereol Leprol 2015;81:509-11.
3. Nanda S, Grover C, Garg VK, Reddy B S. Pemphigus vegetans of hallopeau. Indian J Dermatol 2005;50:166-7.
4. Cozzani E, Christana K, Mastrogiacomo A, Rampini P, Drosera M, Casu M, *et al.* Pemphigus vegetans neumann type with anti-desmoglein and anti-periplakin autoantibodies. Eur J Dermatol 2007;17:530-3.
5. Mergler R, Kerstan A, Schmidt E, Goebeler M, Benoit S. Atypical clinical and serological manifestation of pemphigus vegetans: A case report and review of the literature. Case Rep Dermatol 2017;9:121-30.
6. Jain V, Jindal N, Imchen S. Localized pemphigus vegetans without mucosal involvement. Indian J Dermatol 2014;59:210.
7. Saruta H, Ishii N, Teye K, Ono F, Ohyama B, Koga H, *et al.* Two cases of pemphigus vegetans with IgG anti-desmocollin 3 antibodies. JAMA Dermatol 2013;149:1209-13.
8. Hashimoto K, Hashimoto T, Higashiyama M, Nishikawa T, Garrod DR, Yoshikawa K, *et al.* Detection of anti-desmocollins I and II autoantibodies in two cases of hallopeau type pemphigus vegetans by immunoblot analysis. J Dermatol Sci 1994;7:100-6.
9. Ahmed AR, Blose DA. Pemphigus vegetans. Neumann type and Hallopeau type. Int J Dermatol 1984;23:135-41.
10. Messersmith L, Krauland K. Pemphigus vegetans. In: StatPearls. Treasure Island, FL: StatPearls Publishing; 2019. Available from: https://www.ncbi.nlm.nih.gov/books/NBK545229/. [Last accessed on 2019 Aug 03].