

Crusted scabies in a patient with pemphigus vulgaris after treatment with rituximab and corticosteroids



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INTRODUCTION

Crusted scabies, previously called *Norwegian scabies*, is the rare, severe infestation of the arthropod mite, *Sarcoptes scabiei*.¹ It most often arises among individuals who are immunocompromised or institutionalized or those with cognitive disabilities.^{1,2} Although common scabies is often restricted to specific sites of the skin because of mechanical (eg, scratching) and immune-mediated destruction, crusted scabies is characterized by the epidermal infestation of thousands to millions of mites due to impaired host response.¹ This article presents an unusual case of crusted scabies primarily localized to patches previously affected by pemphigus vulgaris (PV) after treatment with rituximab and prednisolone.

CASE REPORT

A 57-year-old woman with a 3-year history of refractory, biopsy-confirmed mucocutaneous PV presented with 2 weeks of a severe burning sensation on dry patches of her skin. The burning sensation had gradually progressed and was primarily localized to hyperpigmented, scaly patches scattered on her body from prior PV flares. She reported trouble sleeping because of her symptoms, and thought her dry skin may be contributory. The patient denied itch, oral lesions, genital or eye involvement, gluten sensitivity, fever, night sweats, weight loss, herbal supplement intake, or any topical medication use. She had a history of PV refractory to prednisolone and azathioprine, which was replaced with prednisolone and mycophenolate mofetil 6 months prior to her current presentation. Because

Abbreviation used:

PV: pemphigus vulgaris

of a developing a flare 3 months later, her regimen was changed to 500 mg rituximab per week for 4 weeks and 20 mg of daily prednisolone, which led to complete resolution of her PV in 2 weeks. Her medications also included vitamin D supplements, calcium, and alendronate.

Physical examination was notable for diffuse, fine white scale on the trunk and extremities, most prominent on multiple, dusky erythematous plaques with coarse scale secondary to postinflammatory hyperpigmentation from PV (Figs 1 and 2). Scaly, velvety plaques were scattered in her axillae and on her thighs. There were no oral or mucosal findings or cutaneous vesiculobullous eruptions or erosions.

A punch biopsy of a new hyperkeratotic, velvety plaque on her left, lateral chest was performed. Histologic examination found hyperkeratosis and parakeratosis of the epidermis with mild spongiosis, and perivascular lymphocytic infiltrate with eosinophilia in the superficial dermis. Based on these findings, a skin scraping was obtained and numerous eggs and adult mites were seen on microscopic examination, consistent with crusted scabies.

Upon collecting history from family members, the patient's daughter and grandson, both of whom lived with the patient, reported severe itching, and their examinations found excoriated papules, consistent with classic scabies. The patient was started on 200 µg/kg ivermectin and 5% permethrin cream,

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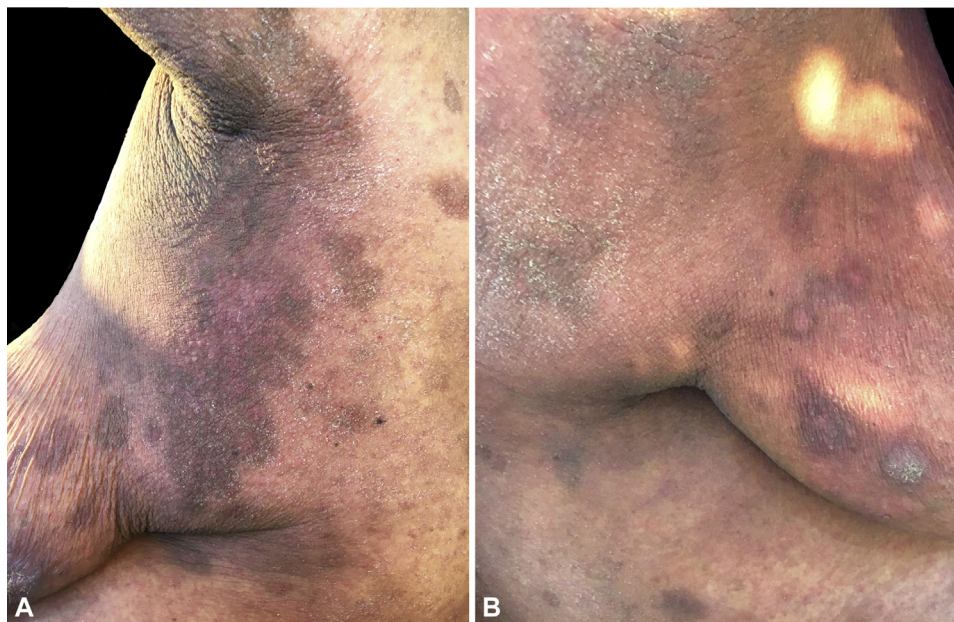


Fig 1. Diffuse fine, white scale on the trunk primarily localized to (A) scaly velvety plaques in the axillae and (B) multiple, dusky erythematous plaques on the breast and lateral chest.

and her family members were started on 5% permethrin cream. Additionally, they were counseled to thoroughly clean and vacuum their home and to collect all items (eg, clothing, linen) used in the prior week and either wash them with hot water and dry them in a hot dryer or store the items in a bag for 10 days. The patient's symptoms fully resolved in 2 weeks.

DISCUSSION

To our knowledge, there are no previous reports of crusted scabies in a patient after treatment with rituximab. Although crusted scabies has been reported among immunosuppressed patients with HIV, systemic lupus erythematosus, leukemias, transplants, and acquired immunodeficiencies, it has traditionally been thought to be caused by an impaired cell-mediated immune response.²⁻⁴ Although this patient likely had impaired cell-mediated immunity because of her chronic corticosteroid use, which by itself may have contributed to crusted scabies, she had crusted scabies only after she also had impaired B-cell response by initiating rituximab. A study examining the immune response in crusted scabies found a predominantly cytotoxic CD8⁺ T-lymphocyte population with minimal helper CD4⁺ T lymphocytes and absent B cells in the dermis, which led the investigators to conclude that the imbalanced inflammatory response results in failure to control the parasite's growth.⁵ This case suggests that B-cell impairment with rituximab

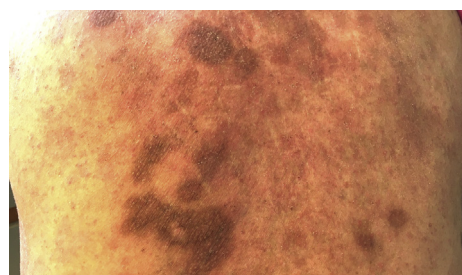


Fig 2. Scaly, hyperkeratotic plaques scattered across the back on sites of postinflammatory hyperpigmentation secondary to PV.

may increase patients' susceptibility to developing crusted scabies, highlighting a need to consider crusted scabies as a potential side effect of dual treatment with corticosteroids and rituximab.

Another unique aspect of this case was the localization of the hyperkeratotic plaques to areas previously affected by PV. This observation is consistent with Wolf isotopic response, an uncommon phenomenon that is characterized by the occurrence of a new disease in the locations of previous healed lesions from an unrelated disease.⁶ Only 1 report described Wolf isotopic response in a patient with crusted scabies, whereby the patient's scabies lesions were localized to the sites of a burn.⁷ Wolf isotopic response has not been previously described after PV.⁸ Given that the mites burrow into the superficial epidermis, we hypothesize scabies may disproportionately affect areas with

damaged skin barriers or thin skin because they are easier entry points for mites than intact skin.

The diagnosis of crusted scabies can be challenging. First, the differential diagnosis for crusted scabies is broad, as it includes disorders associated with hyperkeratotic plaques such as psoriasis, nonbullous impetigo, seborrheic dermatitis, Darier disease, palmoplantar keratoderma, atopic dermatitis, ichthyosis, and dermatitis herpetiformis.⁹ Additionally, its presentation can vary widely from powdery white scales to psoriasis-like thick plaques, and pathognomonic serpiginous threadlike burrows may be absent. Pruritus, which is caused by a delayed type IV hypersensitivity reaction, may also be limited or absent because of immunosuppression.⁴ A burning sensation, as reported by this patient, is an unusual symptom for crusted scabies that has been reported in only 1 other case in conjunction with pruritus.⁷

The diagnosis of crusted scabies requires considering a patient's immune status, soliciting information on the family history of pruritus, and performing a skin biopsy when there is clinical uncertainty.¹⁰ In addition, this case shows that patients with a history of PV treated with a combination of rituximab and corticosteroids may be at higher risk of crusted scabies. Clinical vigilance by keeping a broad differential diagnosis in patients with refractory skin diseases and periodic surveillance of at-risk patients

with PV who are taking rituximab and corticosteroids are critical toward recognizing and treating this highly contagious skin disease.

REFERENCES

1. McCarthy JS, Kemp DJ, Walton SF, Currie BJ. Scabies: more than just an irritation. *Postgrad Med J*. 2004;80(945):382-387.
2. Roberts LJ, Huffam SE, Walton SF, Currie BJ. Crusted scabies: clinical and immunological findings in seventy-eight patients and a review of the literature. *J Infect*. 2005;50(5):375-381.
3. Aiempnakit K. Crusted scabies in a patient with methamphetamine abuse. *JAAD Case Rep*. 2018;4(5):480-481.
4. Bhat SA, Mounsey KE, Liu X, Walton SF. Host immune responses to the itch mite, *Sarcoptes scabiei*, in humans. *Parasit Vectors*. 2017;10:385.
5. Walton SF, Beroukas D, Roberts-Thomson P, Currie BJ. New insights into disease pathogenesis in crusted (Norwegian) scabies: the skin immune response in crusted scabies. *Br J Dermatol*. 2008;158(6):1247-1255.
6. Wolf R, Brenner S, Ruocco V, Filioli FG. Isotopic response. *Int J Dermatol*. 1995;34(5):341-348.
7. Dastgheib L, Boroujeni NH, Aslani FS. Crusted scabies masquerading as psoriasis plaques in a patient suffering from burn scars. *Dermatol Online J*. 2018;24(6).
8. Mahajan R, De D, Saikia UN. Wolf's isotopic response: report of a case and review of literature. *Indian J Dermatol*. 2014;59(3):275-282.
9. Cassell JA, Middleton J, Nalabanda A, et al. Scabies outbreaks in ten care homes for elderly people: a prospective study of clinical features, epidemiology, and treatment outcomes. *Lancet Infect Dis*. 2018;18(8):894-902.
10. Cohen PR. Scabies masquerading as bullous pemphigoid: scabies surrepticius. *Clin Cosmet Investig Dermatol*. 2017;10:317-324.