Pemphigus Herpetiformis Masquerading as Tinea Corporis

Sir,

Pemphigus herpetiformis (PH) is a rare entity and an uncommon subtype of pemphigus. This entity was first introduced in 1975 by Jablonska *et al*.[1] It has atypical clinical presentation with characteristic histopathology of pemphigus. In clinical practice, this entity poses diagnostic challenge. Therefore, a delay in the diagnosis is common.[2] We herein report a case of PH with annular pruritic erythematous plaques and bullous lesions mimicking tinea corporis. A 45-year-old female presented with chief complaints of itchy red annular lesions on the inframammary area, back, abdomen, and lower limbs since 2 months. She had been diagnosed as a case of tinea corporis with tinea cruris and received treatment of oral terbinafine 250 mg twice a day and luliconazole 1% cream once daily. After 5 days of treatment she came to us with appearance of new annular plaques along with blisters, targetoid lesions, and erosions all over the body with blisters mainly over the acral areas. There were no systemic complaints. General physical examination was normal. On cutaneous examination, there were tense, clear fluid-filled vesicles, erosions, and bullae over the normal skin mainly over hands and feet. In addition, she had few targetoid lesion over the forearm and annular plaques with polycyclic margins were present over the chest, abdomen, and back [Figure 1a and b]. Bulla spread sign and Nikolsky's sign were negative. Systemic examination was normal. A differential diagnosis of drug rash, erythema multiforme, pemphigus herpetiformis, bullous pemphigoid, and linear immunoglobulin A (IgA) disease was kept. On investigations, Tzanck smear did not show acantholytic cells and KOH smears for fungal hyphae were negative on multiple occasions. Complete blood count and serum biochemistry were normal except, absolute eosinophil count, which was increased. Skin biopsy from the vesicle revealed focal spongiosis, intraepidermal bulla with acantholytic cells, and neutrophilic exocytosis [Figure 2]. Dermis showed perivascular infiltrate of lymphocytes admixed with neutrophils. Direct immunofluorescence (DIF) showed intercellular deposition of IgG and C3 in the epidermis [Figure 3]. Based upon clinical and histopathological findings, final diagnosis of pemphigus herpetiformis was made. We had started patient on tablet 40 mg prednisolone and dapsone 100 mg daily. Patient showed improvement with clearing of lesions in 2 weeks. Pemphigus herpetiformis is a rare entity and sporadic variant of pemphigus.[2] The incidence of PH is approximately 6% to 7.3%.[1,3-5] It mainly affects adults with no gender predilection.[3,5] Presentation is different from the classical forms of pemphigus.[2] Jablonska *et al*.[1] and Floden and Gentele[6] described this entity in early 1955 and named it as dermatitis herpetiformis with acantholysis. PH has atypical presentation and a benign course. Usual presentation is erythematous annular plaques, vesicles, and bullous lesions.[1,3,5] As a result of centrifugal spread of inflammatory process, the lesions tend to form annular shape.[1,5] Usually, groups of small or abortive vesicles, occasional pustules, often in herpetiform pattern, are present on erythematous base and/or over plaques.[3-5] Sometimes lesions might be just urticarial papules and plaques.[7,8] Common sites of involvement are trunk and proximal extremities.[1,5] In majority of cases mucous membranes are spared.[1-3,4]

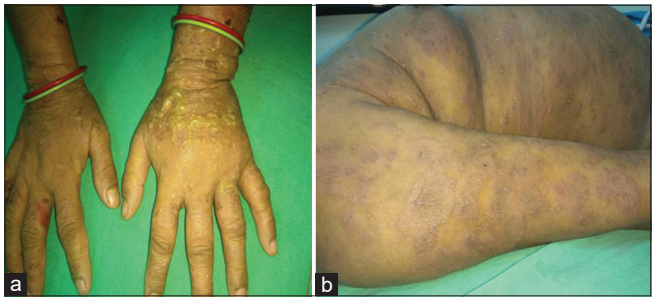

Figure 1: Tense, clear fluid-filled vesicles, erosions, and bullae over the hands and feet (a) and annular erythematous plaques seen over the abdomen, chest, and forearms (b)

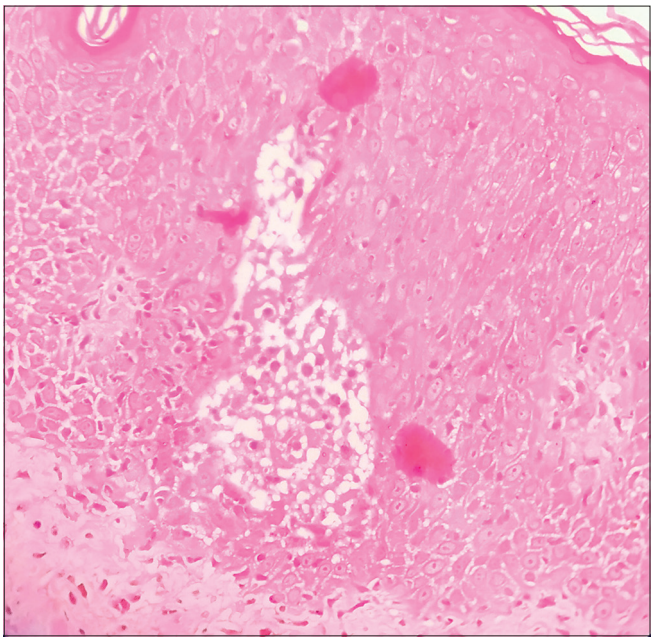

Figure 2: Intraepidermal bulla and neutrophilic exocytosis along with perivascular infiltrate of lymphocytes admixed with neutrophils in the dermis (H and E 40×)

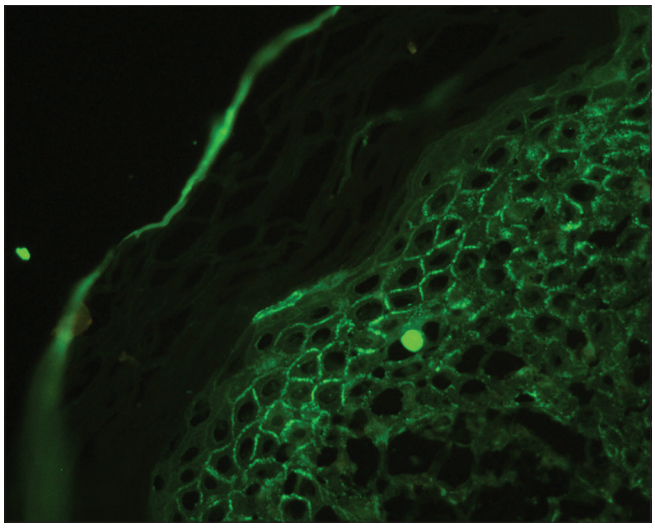

Figure 3: Intercellular deposition of IgG and C3 in the epidermis on DIF

Pruritus often accompanies skin lesions, sometimes it might be severe.[1,3,4] Eosinophilia is found in peripheral blood and had been reported in 37.5% of cases by Laws *et al*.[3] Histopathologically, PH shows the presence of intraepidermal bullae,[3,4] or pustules[3,5] mostly in the subcorneal, and occasionally suprabasal or in the spinous layer.[3,5,8] Dermal papillary neutrophilic microabscesses can also be seen.[8] Spongiosis and epidermal infiltration of eosinophils and/or neutrophils is common.[3] Acantholysis is often mild or absent.[3,5,8] On DIF, intercellular IgG and C3 deposits are most often seen in the superficial layers of the epidermis, and less frequently in the lower layers, mainly when circulating anti-desmoglein-3 antibodies are present.[1] Similar clinical, histopathological, and DIF findings were found in our case. In literature it has been reported that PH can transform into the classical forms of pemphigus (PV and PF). However, the opposite has also been described in the literature.[3] Subacute cutaneous lupus erythematosus (SCLE) is usually manifest as either papulosquamous or annular erythematous scaly lesions. In literature there have been few patients with a recognized and well-documented history of systemic lupus erythematosus (SLE) who developed SCLE after initiation of oral terbinafine for onychomycosis.[9]

Drug of choice is dapsone (100-300 mg daily), as monotherapy or in combination with systemic steroids.[3,4] However, low-dose systemic corticosteroid is enough to achieve complete remission.[3] Other therapeutic options are methylprednisolone as pulse therapy (1 gm/day for 3 days) together with azathioprine 150 mg/day,[3] or azathioprine as monotherapy, cyclophosphamide, sulfapyridine,[1] mycophenolate mofetil, methotrexate,[5] high-dose intravenous immunoglobulin, and plasmapheresis, for more severe cases or cases evolving to classical forms of pemphigus. Recently, therapeutic response to minocycline and nicotinamide has been published.[5] Our patient responded well to 40 mg prednisolone and dapsone 100 mg daily with clearing of lesions in 2 weeks. This emphasizes the importance of keeping pemphigus herpetiformis as a differential diagnosis in this kind of presentation, especially in current scenario of increasing resistance of superficial fungal infection.

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.

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Conflicts of interest

There are no conflicts of interest.

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