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Chronic annular pustular psoriasis resembling subcorneal pustular dermatosis: A case report

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Abstract

Generalized pustular psoriasis and subcorneal pustular dermatosis are generalized pustular dermatoses that are characterized by the subcorneal accumulation of neutrophils. Careful examination is important in distinguishing these diseases for appropriate management. Patients with acute generalized pustular psoriasis are systemically unwell with discrete pustules as opposed to the chronicity and associated hypopyon lesions in subcorneal pustular dermatosis. Generalized pustular psoriasis lesions demonstrate psoriasiform changes on histology and the increased expression of Th17 cytokines. We describe a middle-aged woman presenting with chronic annular generalized pustular psoriasis, initially mistaken for subcorneal pustular dermatosis due to their clinical and histological semblance. The patient had recurrent skin disease for 6 years despite conventional therapy of oral retinoid, immunosuppressant and biologic therapy. Complete and persistent clearance of her skin lesions was achieved with secukinumab, an interleukin 17A inhibitor.

Keywords

Subcorneal pustular dermatosis, Sneddon–Wilkinson disease, annular generalized pustular psoriasis, secukinumab, interleukin-17

Introduction

The differentiation of SPD (subcorneal pustular dermatosis or Sneddon–Wilkinson disease) and generalized pustular psoriasis (GPP) poses a diagnostic challenge as they present similarly both clinically and histologically. They are relapsing neutrophilic dermatoses characterized by widespread symmetrical crops of sterile pustules over an erythematous base that arise predominantly over the trunk and flexural aspects of limbs.[1,2] GPP is further subtyped into acute (von Zumbusch) and subacute annular variants. The presenting morphology of both SPD and annular GPP is similar where the pustules coalesce to form annular or serpiginous patterns.[2] In addition, histological changes in both SPD and GPP include subcorneal pustules filled with neutrophils and occasional eosinophils sitting on top of the epidermis. The upper dermis also shows perivascular and interstitial infiltration of neutrophils and occasional monocytes and eosinophils.[3] Commonly, with appropriate treatment, the pustules of both SPD and GPP resolve completely within several days. However, relapse of skin disease is a characteristic feature of both SPD and GPP.

This case highlights the diagnostic challenges in differentiating between GPP and SPD. We document a unique presentation of annular GPP that was initially mistaken for SPD and treated with a multiple drugs. Recognizing key features and differences between the two skin conditions are important in understanding their long-term disease course and management.

Case report

A woman in her 50s with a 3-year history of pustular dermatosis was referred for assessment of a 1-month history of extensive flare of eruption to the back, arms and legs. The patient complained of low-grade fever, pruritus and skin pain. She had lost 8 lbs unintentionally over the prior month. Three years prior, she was diagnosed with SPD on the basis

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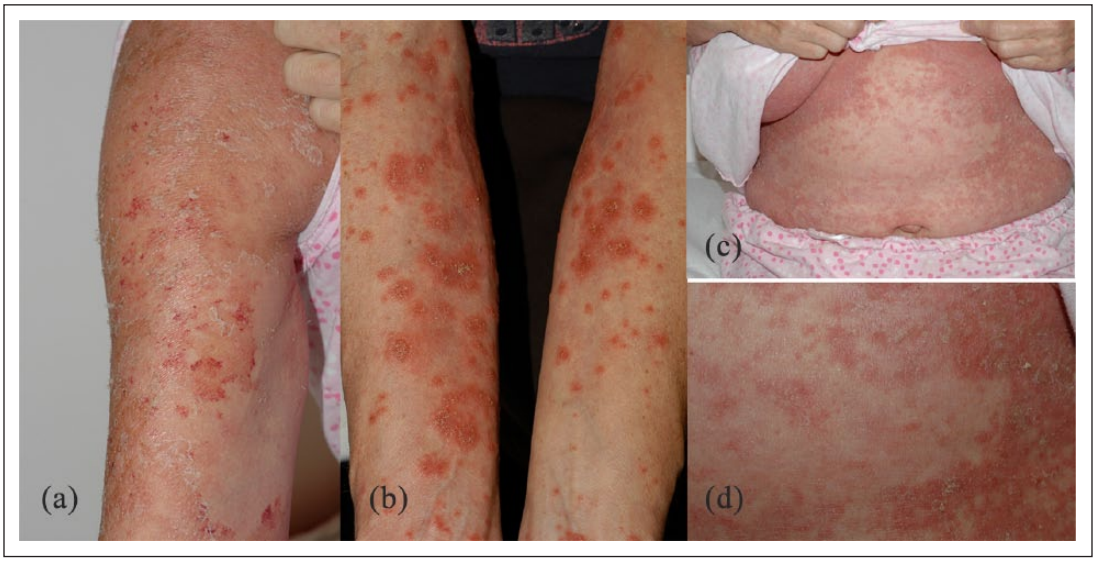

Figure 1. Annular generalized pustular psoriasis: widespread pustular lesions over an erythematous base forming a circinate pattern. (a) Right upper arm, (b) forearms bilaterally, (c) trunk and (d) back.

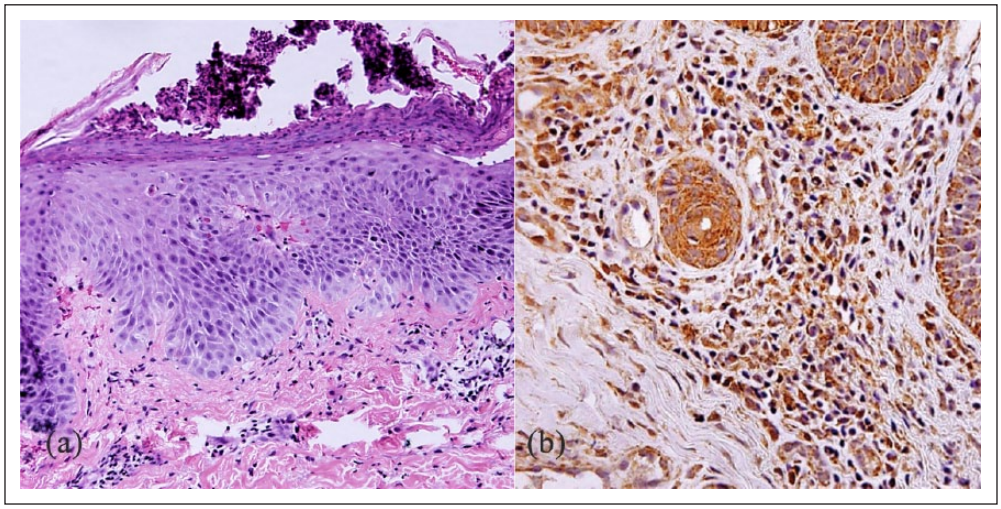

Figure 2. Histological examination of annular generalized pustular psoriasis: (a) H&E staining demonstrates classic psoriasiform changes of acanthosis, parakeratosis, subcorneal pustules, mild spongiosis and (b) histochemical staining demonstrates IL-17A in dermal infiltrates.

of skin biopsy and clinical presentation. She had been started on dapsone, 50 mg daily, and clobetasol propionate ointment, 1 year prior to our assessment with minimal improvement. Additional medication use included betamethasone valerate cream, 0.01% twice daily; citalopram, 50 mg daily; hydroxyzine, 10 mg three times daily; lorazepam, 1 mg as needed; and nasal mometasone furoate, daily.

On examination, she presented with a generalized poly-annular and poly-cyclic pustular and papular eruption with trailing scale and erythema affecting 50% of her body – including the trunk and limbs but sparing the face (Figure 1). A repeat biopsy of the right flank showed spongiosis with parakeratosis, several intracorneal and subcorneal pustules (Figure 2(a)), negative direct immunofluorescence and normal serum protein electrophoresis. These tests ruled out SPD type IgA pemphigus and monoclonal gammopathy-associated disease. The psoriasiform histology and the clinical presentation were consistent with annular GPP.

The patient was started on a tapering regimen of prednisone, 40 mg every morning for 1 week followed by 20 mg every morning for 1 week and then 10 mg every morning for 1 week. The dose of dapsone was increased to 100 mg, daily. Over the course of 6 years, the patient continued to have persistent eruptions consisting of annular pustules with erythematous macules over the trunk and extremities with recurrent flares and systemic illness that required hospitalization on one occasion. Her chronic annular GPP demonstrated minimal improvement with acitretin, cyclosporine, infliximab and ustekinumab. Concurrent hepatitis C was viewed as a

relative contra-indication to therapy with methotrexate. Immunohistochemical analysis of lesional skin demonstrated interleukin (IL)-17A expression by the epidermis and dermal lymphocytes (Figure 2(b)). Treatment with secukinumab was initiated with complete and sustained clearance of skin disease over 2 years of continued therapy (300 mg/month).

Discussion

This case serves to document the nosological confusion surrounding the diagnosis of GPP and SPD, due to their clinical and histological semblance. Demographically, the subacute annular variant of GPP has been described mostly in children, whereas both acute GPP and SPD occur more commonly in adults between the ages of 40 and 60 years.[2,4,5] Clinically, GPP initially develops as painful erythematous papules and pustules, accompanied by fever, rigors and malaise. Patients often appear systemically ill, and life-threatening complications can occur without supportive treatment.[1] In contrast, SPD has a benign and chronic clinical course. In addition, the pustules in SPD arise as half pustular, half vesicular blisters, as opposed to purely pinpoint pustules commonly seen in GPP.[2] Histologically, the hallmark differences between GPP and SPD include the classic psoriasiform changes seen in the epidermis of patients with GPP, parakeratosis, elongation of rete ridges spongiform pustules of Kogoj and acantholysis.[2,6] However, biopsy of older pustules in GPP can be histologically identical to SPD, which lack spongiform changes from epidermal healing.

First-line treatments for SPD and GPP are oral dapsone and acitretin, respectively. However, relapses are common and patients are directed towards second-line therapies, which include anti-neutrophilic, topical or oral steroids, psoralen plus ultraviolet light (PUVA) and newer biologic therapies.[2,7] No randomized trials have addressed the efficacy of therapy for SPD or GPP. Recently, case series have documented the efficacy of anti–tumour necrosis factor α (TNFα) and anti-IL-12/23 agents in the resolution of skin disease in both SPD and GPP.[2,7]

Like TNFα, IL-17A, a pro-inflammatory cytokine, plays an important role in the recruitment of neutrophils to the epidermis of patients with psoriasis. The pathogenesis of GPP is postulated to involve the interaction between the adaptive immune system through the IL-17/23 axis with the keratinocyte-derived IL-36 pathway of the innate immune system. A recent study has demonstrated the potential role of IL-36 in inducing the IL-17A pathway either directly or indirectly via IL-23 expression and that it may act concertedly with IL-17A in the exacerbation of inflammation in pustular psoriasis.[8] Secukinumab, an IL-17A monoclonal antibody, is approved for the treatment of plaque psoriasis. The efficacy of secukinumab for acute GPP has recently been documented in a 52-week open-label study, as well as in case reports.[9,10] The efficacy of secukinumab on SPD or annular GPP has yet to be reported. Although our patient's clinical presentation and histologic findings were similar to that of SPD, the lack of remission to the initial therapeutic regiments, concomitant psoriasiform epidermal hyperplasia on skin biopsy, and the chronic course of pustulation and erythema with recurrent flares of systemic illness suggest an unusual presentation of chronic annular GPP.

This case highlights the importance of recognizing the subtle clinical and histological differences between SPD and annular GPP. The pro-inflammatory cytokine, IL-17A, possibly in synergy with IL-36, plays an important role in the pathogenesis of GPP. Recognition of annular GPP can lead to the initiation of secukinumab, or other IL-17 inhibitors, for complete and sustained clearance of skin disease.

Authors' Note

In the past 2 years, Dr Dutz has been a member of advisory boards and a speaker for Janssen, Abbvie, Amgen, Leo Pharma, Celgene, Lilly, Bausch Health, Sanofi Genzyme and Novartis and has participated in clinical trials supported by Janssen, Abbvie and Novartis.

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Informed consent

A written informed consent was obtained from the patient.

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