Subcorneal pustular dermatosis in a 54-year-old woman

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A 54-year-old woman presented to our dermatology department with an eruption that she had experienced episodically for 20 years. Flares were characterized by pustules that appeared within flexural regions and expanded over several days to form larger arcuate plaques. The lesions persisted for about 4–8 weeks, followed by periods of remission lasting 1–2 years. She had been treated with topical corticosteroids and dapsone, with partial remission of the pustules.

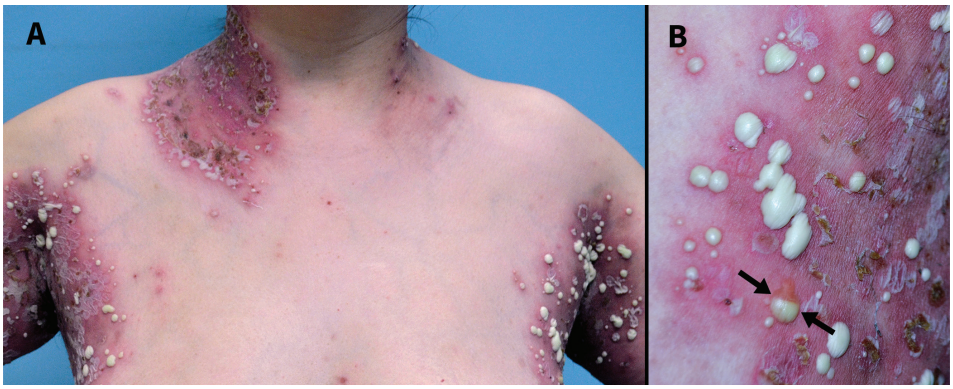

Figure 1: (A) Flexural truncal distribution of scaly pustular eruption on a 54-year-old woman with subcorneal pustular dermatosis. (B) A close-up image shows the hypopyon sign, with pus accumulating in the lower-half of a flaccid blister (black arrows).

When we examined the patient, she was afebrile with normal vital signs. We saw serpiginous pustules in a flexural distribution (Figure 1A). We noted the hypopyon sign with pus in the lower half of flaccid blisters (Figure 1B). Test results of serum protein electrophoresis with immunofixation, monoclonal immunoglobulin levels, antinuclear antibody panel and rheumatoid factor were normal. Cultures of the pus yielded no bacteria or fungi. Skin biopsy showed subcorneal accumulation of neutrophils, which confirmed the diagnosis of subcorneal pustular dermatosis. The pustular rash resolved completely 2 months after starting oral acitretin (25 mg/d) and we tapered the dose to 10 mg daily. The patient had no new lesions when we saw her 5 months after her initial visit.

Subcorneal pustular dermatosis, also known as Sneddon–Wilkinson disease, is an uncommon disease that usually follows a chronic, relapsing and remitting course, and most commonly affects women aged 50–80 years.[1] The disease starts as flaccid, pea-sized pustules that evolve over 24–48 hours to an annular or serpiginous distribution, often with the hypopyon sign. Subcorneal accumulation of neutrophils is the salient pathologic feature.[2] The differential diagnosis includes acute generalized exanthematous pustulosis, annular pustular psoriasis, pemphigus vulgaris, impetigo and candidal intertrigo. Subcorneal pustular dermatosis can be associated with other conditions such as immunoglobulin A paraproteinemia, multiple myeloma, rheumatoid arthritis and Sjogern syndrome; patients with subcorneal pustular dermatosis should be investigated for these conditions as clinically indicated.[1,3] Pustules can rapidly improve with dapsone (50–200 mg daily) or acitretin (0.25–1 mg/kg daily).[2] Maintenance therapy is often required to prevent relapse.

References

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