

RESIDENT’S FORUM

Isolated erythematous plaque on the right cheek

Case report

A 54-year-old man without underlying disease had one mild itchy rash on the right cheek for 6 months. He tried topical fluocinolone acetonide ointment and the lesion became less itchy. However, the rash enlarged gradually, so he came to our clinic. Cutaneous finding was a 4 × 5 cm, erythematous-to-brownish plaque on the right cheek with focal erosions. No lesions were found elsewhere (Figure 1A). A skin biopsy was done, which revealed acantholytic cells mainly in the granular layer and nonspecific perivascular and perifollicular infiltration in the dermis (Figure 1B). Focal hydropic changes and pigment incontinence were also present. Direct immunofluorescence (DIF) showed positive immunoglobulin (Ig) G deposition at the intercellular space and along the basement membrane zone (BMZ; Figure 1C). Indirect immunofluorescence (IIF) showed anti-intercellular space antibody 1:80 (+). Antinuclear antibody (ANA) was 1:40 (−) and antiextractable nuclear antigen (ENA) antibodies were all negative. He was treated with oral prednisolone 50 mg daily by tapering dose during the following 5 months; the lesion improved gradually (Figure 1D).

Figure 1 (A) One 4 × 5 cm, erythematous-to-brownish plaque on the right cheek with focal erosions; (B) acantholytic cells mainly in the granular layer (hematoxylin & eosin, 200×); (C) direct immunofluorescence shows positive immunoglobulin G deposition at the intercellular space and along the basement membrane zone (100×); (D) rash resolved into one brownish patch after 5 months of treatment.
Diagnosis

Pemphigus erythematosus (PE).

Discussion

PE is originally described as a variant of pemphigus with features of lupus erythematosus (LE), such as typical butterfly pattern of facial lesions, deposition of immunoglobulins at the dermoepidermal junction, and LE features in the histopathology or serologic findings of systemic LE. However, a true dual diagnosis of pemphigus and systemic lupus erythematosus (SLE) is less common and the two diseases appear to progress independently. The patients with dual diagnosis are more similar to the demographic profile of SLE than pemphigus vulgaris, and nearly 100% of such patients are positive for ANA (ANA titer \( \geq 1:80 \)). Overall, a positive ANA is detected in 30%–80% of patients with PE, and our patient had a negative result of ANA.

Clinically, PE usually presents as small, flaccid bullae with scaling and crusting on the scalp, face, upper chest, and back. On the face, PE presents on the bridge of the nose and on the malar areas in a butterfly distribution as seen in LE. There is no report of unilesional PE in the literature. However, unilesional lesions in other immunologic diseases, such as LE, are occasionally reported.

On histologic examination, pemphigus foliaceus and PE demonstrate acantholysis just below the stratum corneum or within the granular layer. The blister may contain numerous acute inflammatory cells, particularly neutrophils. Lichenoid dermatitis might be seen, but the lupus feature might not always exist.

DIF and IIF could be a supportive evidence for diagnosis. DIF shows IgG or C3 deposition on epidermal cell surfaces in 90% of patients and most, but not all patients (in 70%–80% patients sampled from sun-exposed areas) have additional finding of granular IgG or C3 deposition at the BMZ. IIF reveals 80%–100% positive of anti-intercellular space antibody (from 1:10 to 1:2560).

Our case has the DIF result of IgG deposition both on epidermal cell surfaces and at the BMZ and the IIF result is anti-intercellular space antibody 1:80 (+).

Conclusion

PE is a superficial form of pemphigus that shows some features of LE. The course of PE is usually milder and it responds to lower doses of systemic corticosteroids.

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