ACANTHOLYTIC PEMPHIGUS VULGARIS, HALLOPEAU VEGETATING FORM

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Abstract. A clinical observation of a patient with pemphigus vegetans, Hallopeau type, is described. A literature review concerning the etiology, pathogenesis, clinical manifestations, diagnosis, and treatment of this disease is also presented.

Key words: acantholytic pemphigus vulgaris, Hallopeau type pemphigus vegetans, clinical presentation, diagnosis, treatment.

Introduction. Acantholytic pemphigus, or pemphigus, belongs to the group of autoimmune blistering diseases of the skin and mucous membranes. Its histological characteristic is epidermal blistering caused by acantholysis, while its immunological feature is IgG directed against the surface of keratinocytes.

In 1886, G. Neumann distinguished pemphigus vegetans from true pemphigus. In 1898, H. Hallopeau described vegetating pyoderma. Lever and Schaumburg-Lever later regarded pemphigus vegetans as a variant of pemphigus vulgaris, distinguishing the classical Neumann type and the benign Hallopeau type.

Pathogenesis. The disease is associated with IgG autoantibodies against intercellular adhesion proteins, principally desmoglein 1 and desmoglein 3. Disruption of desmosomal adhesion produces acantholysis and intraepidermal blister formation.

Clinical presentation. The vegetating form may present with blisters, erosions, pustules and vegetations, particularly around natural orifices and in intertriginous areas. The Hallopeau type is generally localized, especially in skin folds.

Laboratory diagnosis. Direct immunofluorescence demonstrates IgG on keratinocytes. Indirect immunofluorescence detects circulating antibodies. ELISA can identify antibodies against specific intercellular adhesion proteins. Tzanck cells are supportive but are not specific.

Treatment. The article describes systemic glucocorticosteroids, principally prednisolone, together with immunosuppressive therapy. Additional approaches discussed include methylprednisolone pulse therapy, high-dose intravenous gamma globulin, plasmapheresis, cyclosporine, gold preparations, antimalarial drugs and extracorporeal photochemotherapy.

CLINICAL CASE

A male patient born in 1951 presented with facial lesions. He associated their onset with trauma from a wild-plum thorn in July 2012. He had previously been treated for pyoderma and microbial (paratraumatic) eczema of the face with systemic, topical and physiotherapeutic treatments, without improvement.

At admission in late October, the process was limited, asymmetric and regional. On the scalp, predominantly in the frontoparietal region, there were pustules and coin-sized erosions with an infiltrated peripheral ridge. On the right half of the face, from the nasolabial fold to the temporal region, there was an irregular plaque with sharply defined borders, erythema, slight infiltration, crusts, isolated erosions and microfissures.



Figure 1. Lesion on the skin of the right half of the face.

Microscopic examination of material from the lesion revealed no fungal mycelial hyphae. Because sporotrichosis was suspected, itraconazole 0.1 g, one capsule twice daily, was prescribed. No positive effect was observed after 10 days.

On days 10–14 of hospitalization, the scalp lesions increased and new lesions appeared on the oral mucosa, right inguinal fold and perianal region. The oral mucosa showed whitish, painful aphthae. Scalp erosions had a pronounced peripheral ridge and vegetating bases covered with purulent discharge. The right inguinal lesion extended onto the anterior thigh and showed vegetations, microfissures, erosions and pustules; vegetations were also present perianally.

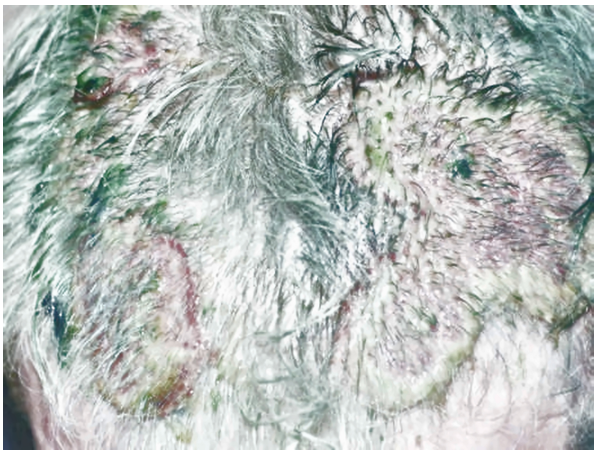


Figure 2. Erosions with a purulent peripheral ridge on the scalp.

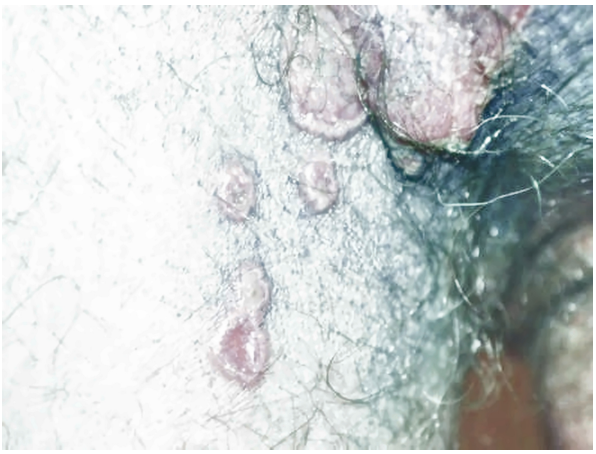


Figure 3. Lesions of the inguinal region extending onto the anterior surface of the thigh.

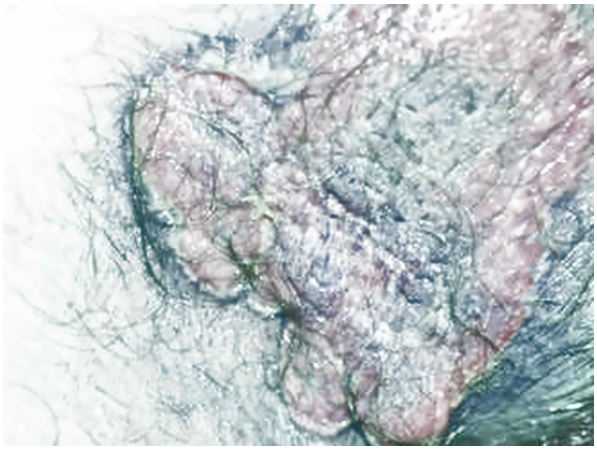


Figure 4. Vegetating lesions of the inguinal region.



Figure 5. Vegetations in the perianal region.

DIAGNOSIS AND HISTOPATHOLOGY

Based on the history, complaints and clinical manifestations, a preliminary diagnosis of acantholytic pemphigus vulgaris, vegetating form, was made. Acantholytic cells were detected in smear preparations from lesions of the oral mucosa, scalp, inguinal and perianal regions, surrounded by eosinophils.

A skin biopsy was performed under local anesthesia. Histopathology showed pronounced epidermal acanthosis, moderate papillomatosis, an epidermal cleft containing necrotic cellular material predominantly composed of eosinophils, suprabasal acantholytic clefts and isolated Tzanck cells. Eosinophilic spongiosis was present, with moderate polymorphous dermal infiltration predominantly consisting of eosinophils and dilated vessels.

Histopathological conclusion: acantholytic pemphigus vegetans, Hallopeau type.

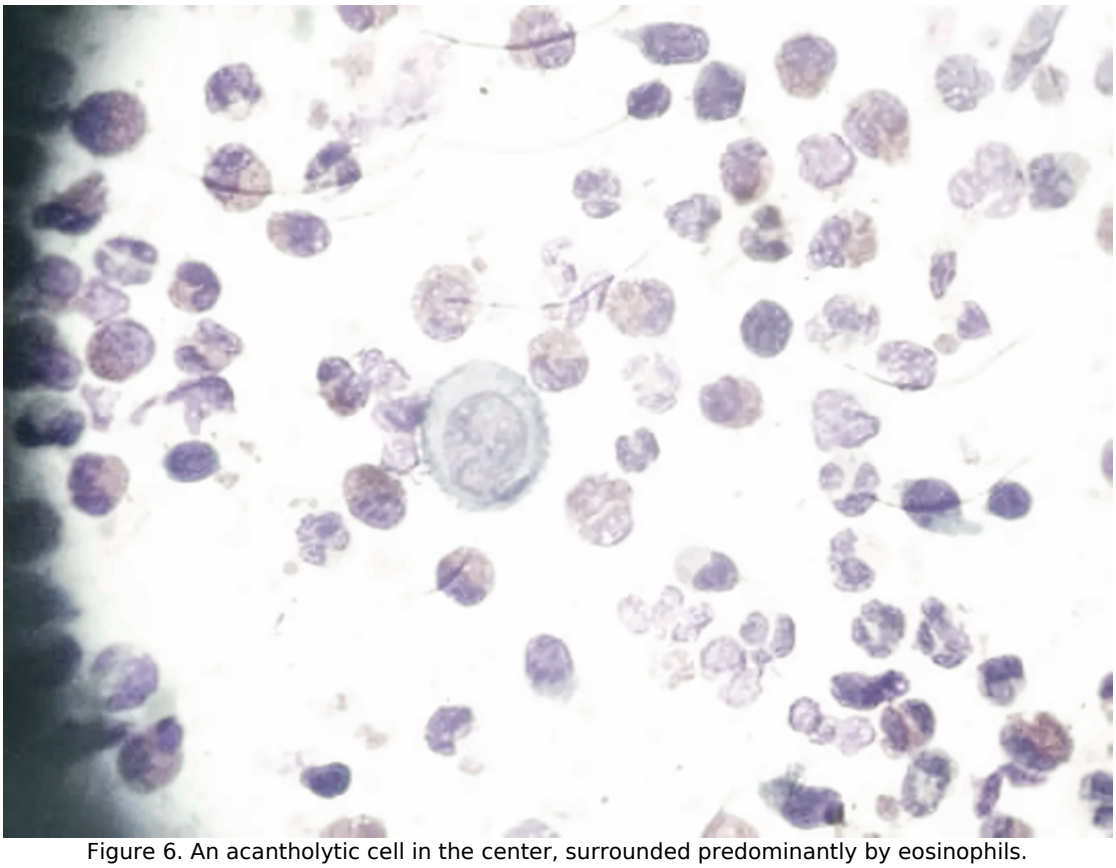


Figure 6. An acantholytic cell in the center, surrounded predominantly by eosinophils.



Figure 7. Histopathological section showing isolated Tzanck cells in a suprabasal acantholytic cleft and eosinophilic spongiosis.

LABORATORY FINDINGS AND TREATMENT

The complete blood count, urinalysis and biochemical parameters were without pathological abnormalities. MPR and HIV tests were negative. Microscopy for fungi showed no mycelial hyphae and culture showed no growth. Tzanck cells were detected cytologically.

Chest X-ray and ultrasound of the internal organs revealed no visible pathology. Colonoscopy found three polyps in the ascending colon, with follow-up recommended after one year. Prednisolone 90 mg/day was prescribed. On the tenth day, a pronounced positive therapeutic effect was observed.

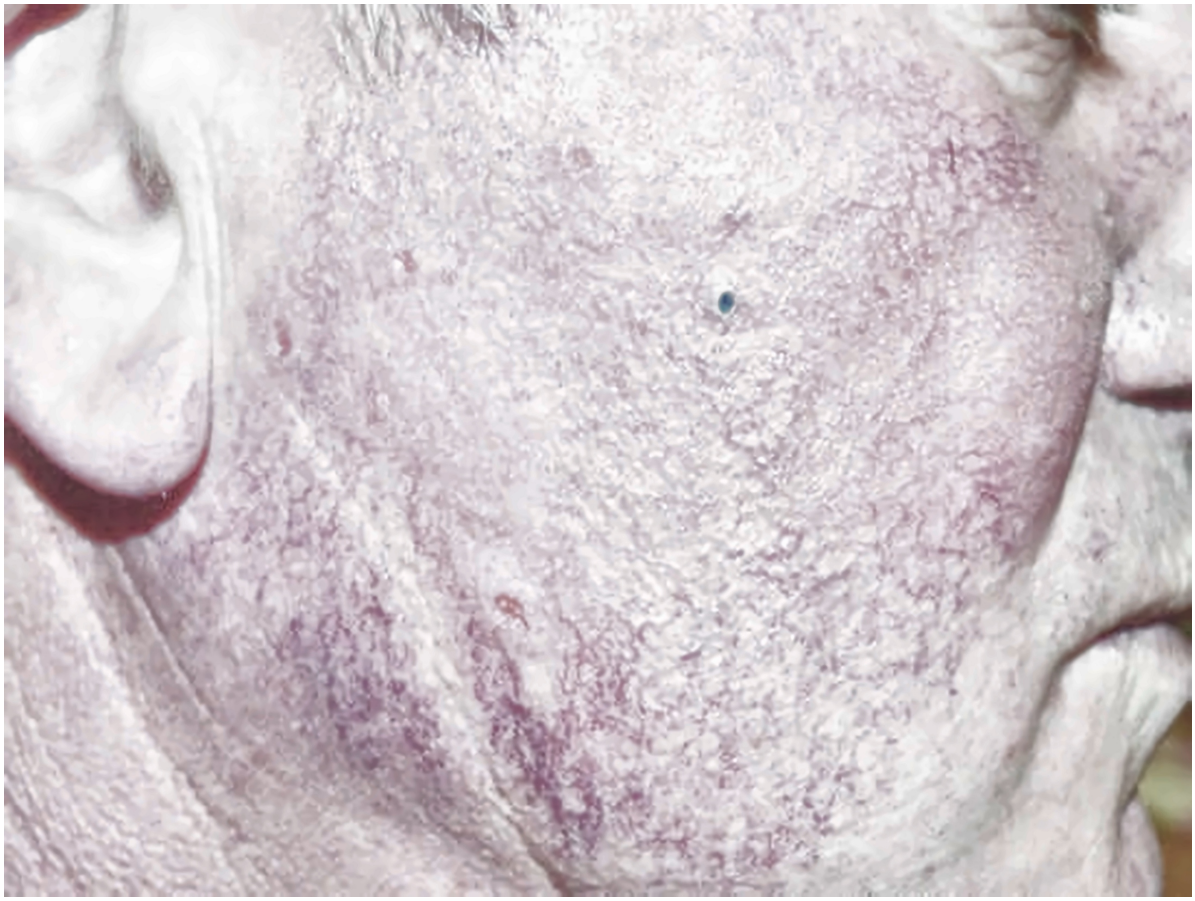


Figure 8. Appearance during treatment on the 10th day.

DISCUSSION

The authors note that Hallopeau-type pemphigus vegetans is frequently not diagnosed, and patients may be observed under diagnoses such as microbial eczema, sporotrichosis, chromomycosis, vegetating Darier disease and chronic familial benign pemphigus (Hailey–Hailey disease).

The clinical features emphasized are pustules merging into infiltrated plaques with rapidly developing vegetations; localization in intertriginous areas; histological acantholysis with eosinophils and eosinophilic spongiosis; IgG deposition on keratinocytes by direct immunofluorescence; and the reported response to glucocorticosteroid therapy.

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