CHICAGO DERMATOLOGIC SOCIETY

Key location(s):
Scalp, tongue, inguinal folds, oral commisures

CASE 5

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History of Present Illness

43 year-old Hispanic male presented with mildly pruritic inguinal lesions over a one-week period. Initially, the lesions had a clear to yellow discharge; subsequently, they thickened, and he developed associated burning pain. Prior to presentation, his primary care physician had started him on mometasone 0.1% ointment daily, with mild improvement. Of note, he had been on prednisone 20 mg daily and dapsone 25 mg daily for over ten years to control a skin condition, which manifested as painful "bumps" on the scalp, with occasional drainage. Other than his skin lesions, he reported no symptoms. He is married, lives in Chicago, and had no recent travel.

Past Medical History

Diabetes Mellitus Type II

Medications/Allergies

Insulin, prednisone, dapsone, mometasone ointment/NKDA

Social History

Social drinker
No tobacco/illicit drugs
Mechanic
Married

Review of Systems

Negative for fever, chills, weight loss, fatigue, night sweats, dysphagia, oral ulcers, arthralgia, nausea, vomiting, diarrhea

Physical Exam

Skin:	Bilateral inguinal creases: well-defined, vegetative, violaceous plaques with peripheral pustules
	Scalp: scattered, flesh colored, papillomatous plaques
	Face: diffuse, open comedones
	Chest: numerous folliculocentric, hyperpigmented macules, pink papules, and scattered pustules
Mucosa:	Dorsal tongue: deep fissures forming gyri
	Oral commissures: fissures and pink vegetative plaques

Laboratory Data

The following labs were remarkable/abnormal:

Platelets	121-160 k/μL	[161 – 369 k/μL]
Eosinophil %	1.6-10.1%	[0.4 – 5.8%]
HIV	Non-reactive	

Histopathology

INGUINAL CREASE, PUNCH BIOPSY:
Epidermis: Acanthosis with focal eosinophilic pustule formation
Dermis: Fibrosis with predominantly eosinophilic infiltrate

INGUINAL CREASE, DIRECT IMMUNOFLUORESENCE:
No IgG, IgA, IgM, C3, C5b-9 or fibrinogen deposition

SCALP, PUNCH BIOPSY:
Pseudoepitheliomatous hyperplasia and mixed inflammatory cell infiltrate composed of predominantly eosinophils, with focal eosinophilic pustule formation.
PAS stain for fungus negative.

SCALP, DIRECT IMMUNOFLUORESENCE:
Linear/granular IgG deposition throughout the epithelial cell surfaces.
Linear/granular C3 deposits on the lower two-thirds of the epithelial strata.
No immunoreactants at the basement membrane zone
No IgA, IgM, C5b-9 or fibrinogen deposition

Diagnosis

Pemphigus Vegetans, possibly Hallopeau type

Treatment and Course

The previous regimen of prednisone 20 mg daily and dapsone 25 mg daily was continued, with the new additions of clobetasol 0.05% ointment and mycophenolate mofetil, 1000 mg BID. Mycophenolate mofetil was titrated to the current dose of 1500 mg BID, and dapsone was titrated to the current dose of 100 mg daily. Prednisone has been tapered to 10 mg every other day. He continues to have intermittent scalp lesions. The inguinal lesions have been clear for many months.

Discussion

Pemphigus vegetans is a rare variety of pemphigus, encompassing only 1-2% of all pemphigus cases. Clinically, two variants exist: Neumann, first reported in 1876, and Hallopeau. Initially described in 1889. Both subtypes manifest as vegetative and papillomatous plaques that are preceded by either polycyclic pustules in the Hallopeau type, or flaccid bullae in the Neumann type. The Hallopeau type is believed to have a better prognosis, even self-resolving in some cases.

Although pemphigus vegetans can occur at any age, it has a predilection for middle-aged women. Lesions typically arise in intertriginous zones and oral mucosal sites, including the lips, hard palate, and buccal mucosa. Oral involvement, which is present in 60-80% of cases, is often the initial manifestation of the disease. On rare occasion, lesions may be limited to one site, such as the oral cavity, tongue, vulva, groin, and very rarely, the soles and scalp. In a retrospective study, eight of seventeen patients with pemphigus vegetans had involvement of the scalp, with one patient having disease confined to the scalp. In addition, there can be considerable morphological variation and associated clinical findings. Unusual reported presentations, for example, include an acrodermatitis continua suppurativa-like picture with nail involvement (loss of nail and paronychia. Similarly, disease may present as cerebriform tongue, which can precede the development of more typical lesions. Indeed, in one retrospective study, six out of ten patients with pemphigus vegetans had this finding. Given these variations, diagnosis can be challenging. Other diseases such as pyodermatitis pyostomatitis vegetans, vegetating

variants of other autoimmune bullous diseases (IgA pemphigus, bullous pemphigoid), halogenoderma, and pyoderma vegetans should be considered in the differential diagnosis.

Diagnosis is confirmed by characteristic histopathologic and immunopathollogic findings. Early lesions present with surpabasilar acantholysis. Classic findings are epidermal hyperplasia, papillomatosis, and eosinophilic pustules. Immunopathollogic findings, indistinguishable from pemphigus vulgaris, include deposition of IgG and C3 around keratinocytes. Desmoglein 1 and desmoglein 3 are the most commonly reported antigens. Although not a sensitive finding, eosinophilia has been reported in several cases.

Treatment consists of systemic corticosteroid and steroid-sparing agents such as azathioprine, mycophenolate mofetil, methotrexate, dapsone, and cyclosporine. To date, there are no large-scale studies reviewing treatment or prognosis.

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