BRIEF ARTICLE

Cutaneous Type Pemphigus Vulgaris of the Scalp: A Rare Unilesional Presentation

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ABSTRACT

Pemphigus Vulgaris (PV) is the most common subtype of pemphigus, a rare group of autoimmune bullous diseases affecting the skin and mucous membranes. PV can be further subdivided into mucocutaneous and mucosal dominant types, depending on the extent of cutaneous involvement. Almost all cases of PV have mucosal involvement; however, a rare variant of cutaneous-only PV has been reported in the literature. To our knowledge, only two previous accounts of unilesional scalp PV have been reported. We present an unusual case of cutaneous-only PV involving the scalp.

INTRODUCTION

Pemphigus represents a rare group of autoimmune bullous diseases affecting the skin and mucous membranes with an incidence of 1 to 16 per million annually.[1,2] Pemphigus Vulgaris (PV) is the most common subtype, comprising 70% of all cases of pemphigus and can be further subdivided into mucocutaneous and mucosal dominant types, depending on the extent of cutaneous involvement.[1] Almost all cases of PV have mucosal involvement; however, a rare variant of cutaneous-only PV has been reported in the literature.[3] We present a case of cutaneous-only PV involving the scalp.

CASE PRESENTATION

A 36-year-old otherwise healthy Caucasian woman presented to the dermatology office with a painful, pruritic, erythematous plaque isolated to the vertex scalp that had been present for one year. The patient had previously seen an outside provider and was prescribed a four-week oral prednisone taper, oral minocycline, ketoconazole shampoo, and gentamicin ointment. Minimal improvement was noted on this regimen, and the patient was still experiencing pain and irritation which led her to seek secondary evaluation by a dermatology physician. Physical examination at this time revealed a beefy red, macerated plaque on her vertex scalp with overlying scale (Figure 1). No other cutaneous or mucosal lesions were noted. Two 4-mm punch biopsies were performed and sent for histopathological and immunohistochemical analysis. Laboratory studies included CBC, CMP, QuantiFERON-TB Gold, a hepatitis panel, and an HIV test.

Histopathology revealed suprabasilar acantholytic dermatitis at all levels of the epidermis (Figure 2). Direct

immunofluorescence staining was positive for linear/granular IgG deposition throughout the epithelial cell surfaces. Linear/granular deposits of C3 were also noted in the lower two-thirds of the epithelial strata (Figures 3, Figure 4). No immunoreactants were noted at the basement membrane zone and no IgA, IgM, C5b-9 or fibrinogen deposits were seen. These immunofindings, coupled with the histological analysis, strongly supported the diagnosis of pemphigus vulgaris.

Figure 1. Initial presentation—beefy, red, macerated plaque on the vertex scalp.

The patient was treated with mycophenolate mofetil 500mg twice daily, prednisone 20mg twice daily, doxycycline 100mg twice daily, and fluocinolone oil. One-month follow-up revealed significant improvement of the scalp lesion. The patient was maintained on mycophenolate mofetil 500mg twice daily with topical fluocinonide oil and prednisone was tapered off. At six-month follow-up she had nearly complete resolution. Serum indirect immunofluorescence was assessed after nine months of treatment and was nonreactive on monkey esophagus, salt-split skin, and murine bladder correlating with successful disease remission and clinical clearance.

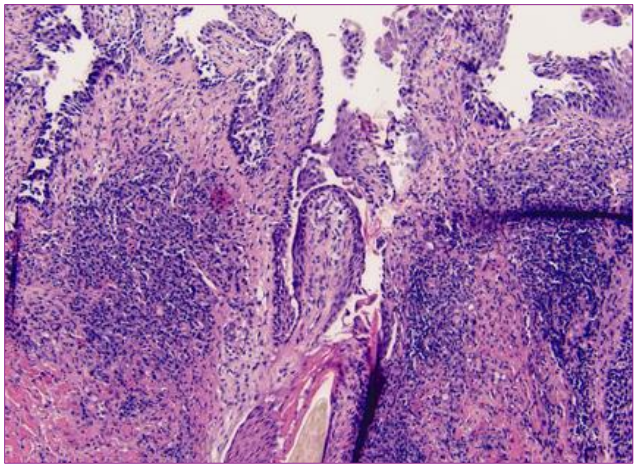

Figure 2. Histopathology showing suprabasilar acantholysis and characteristic “tombstoning” of basilar keratinocytes (H&E, 10x).

DISCUSSION

PV is an autoimmune bullous disease that can affect both the skin and mucous membranes. Autoantibodies to desmoglein (Dsg) antigens represent the major pathogenic factor in pemphigus. Mucosal dominant PV has predominantly anti-Dsg3 autoantibodies, while mucocutaneous PV has both anti-Dsg3 and anti-Dsg1 autoantibodies. Cutaneous-type PV (cPV) is an extremely rare variant of PV that presents with cutaneous findings in the absence of any mucosal involvement. Currently, there is limited literature on this mucosal-sparing phenotype with only a handful of documented cases.[3,4,5] Rangel reported one case of cutaneous-type PV associated with pregnancy.[4] Gheisari et al. recently reported that 30/560 (5.3%) of their studied patients with PV had only cutaneous involvement, suggesting that cPV is much more prevalent than previously thought.[5] Furthermore, only two previous accounts of unilesional scalp PV have been reported.[6,7] Our case is a unique presentation of unilesional cutaneous PV in a young woman.

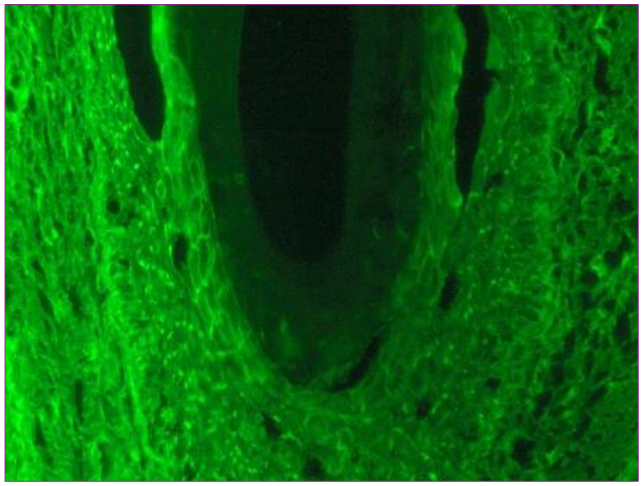

Figure 3. Direct immunofluorescence showing IgG deposition involving follicular epithelium of the scalp. (IgG, 400x)

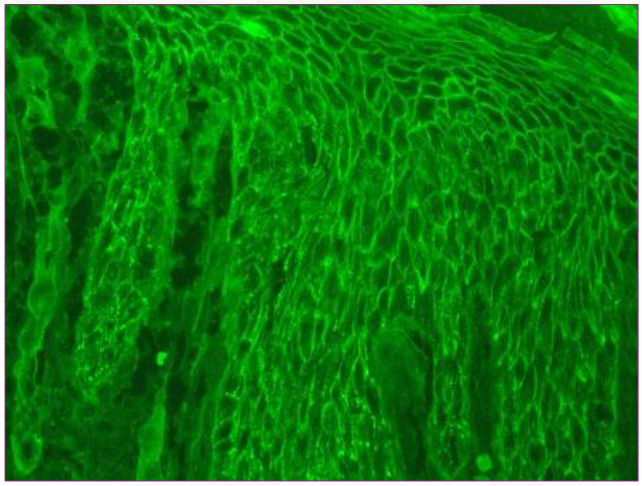

Figure 4. Direct immunofluorescence showing linear/granular deposits of IgG throughout the epidermis of the scalp. (IgG, 400x)

Scalp involvement is seen in 16-60% of patients with PV and is the primary location in 9-15% of patients.[8] The high frequency of scalp involvement in PV is likely due to the presence of high levels of desmogleins in hair follicles.[8] In cases of suspected pemphigus limited to the scalp, the additional use of trichoscopy can aid in diagnosis.[7]

Several hypotheses for the pathogenesis of cPV have been proposed in the literature. Yoshida et al. hypothesized cPV to be caused by a combination of pathogenically weak anti-Dsg3 with potent anti-Dsg1.[3] Sar-Pomian et al. suggested that other antigens may be involved as targets in the pathogenesis of PV and that the desmoglein autoantibodies detected may be the result, rather than the cause, of the observed acantholysis.[8] Carew et al. reported concomitant pathogenic and non-pathogenic epitopes of Dsg3 in mice.[2] Further studies are needed to fully understand this complex pathogenesis. In our case, ELISA was not performed in addition to IIF, but could be a consideration in future cases to help potentially understand the pathogenesis of cPV.

CONCLUSION

The clinical significance of this case highlights a rare form of PV without mucosal involvement, a hallmark of PV. It is important to consider pemphigus vulgaris, even in the absence of mucosal involvement when evaluating isolated scalp lesions.

Abbreviations:
PV – Pemphigus vulgaris
PF – Pemphigus foliaceous
GERD – Gastroesophageal reflux disease
Dsg – Desmoglein
DIF – Direct immunofluorescence

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