Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

Pemphigus induced alopecia : a rare presentation

DOI: 10.70780/medpeer.000QGNU

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ABSTRACT

Pemphigus vulgaris (PV) is a rare autoimmune blistering disease characterized by mucocutaneous erosions due to autoantibodies against desmogleins. While scalp involvement is relatively common, progressive alopecia as a presenting symptom remains rare and may delay diagnosis. We report the case of an 80-year-old woman who initially presented with an erosive alopecic plaque of the scalp, evolving over three months. The diagnosis of PV was established following the appearance of bullous lesions on the trunk and limbs, supported by histology and direct immunofluorescence findings. Despite the diagnosis, the patient rapidly deteriorated due to a massive pulmonary embolism and died within 24 hours of admission. This case highlights the importance of recognizing scalp erosions with alopecia as a possible initial manifestation of PV. Awareness of this atypical presentation is essential for early diagnosis and management, potentially improving patient outcomes.

KEYWORDS

Pemphigus vulgaris, Scalp alopecia, Autoimmune blistering disease

MAIN ARTICLE

Introduction

Pemphigus vulgaris (PV) is a rare autoimmune blistering disorder caused by autoantibodies directed against desmogleins 1 and 3 (Dsg), leading to intraepidermal acantholysis. It is characterized by the presence of flaccid bullae and erosions on the skin and mucous membranes [1]. Erosions affecting the scalp is common [2], but progressive hair loss remains rare, which can delay diagnosis.

Observation

We report the case of an 80-year-old woman with a three-month history of an erosive alopecic plaque affecting the right frontal, vertex, and parietal scalp (Fig 1). Before admission, she had consulted multiple physicians and received oral and topical antibiotics without improvement. Two weeks prior to hospitalization, she developed flaccid bullae and erosions larger than 10 cm on her upper trunk, back, and limbs, along with painful oral erosions. The biopsy of a bullae showed suprabasal acantholysis, and inter keratinocyte IgG and C3 were found on the direct immunofluorescence technique. She was then diagnosed with severe pemphigus vulgaris with a severity score of 55. Unfortunately, within 24 hours of admission, she developed a massive pulmonary embolism, leading to rapid deterioration and death.

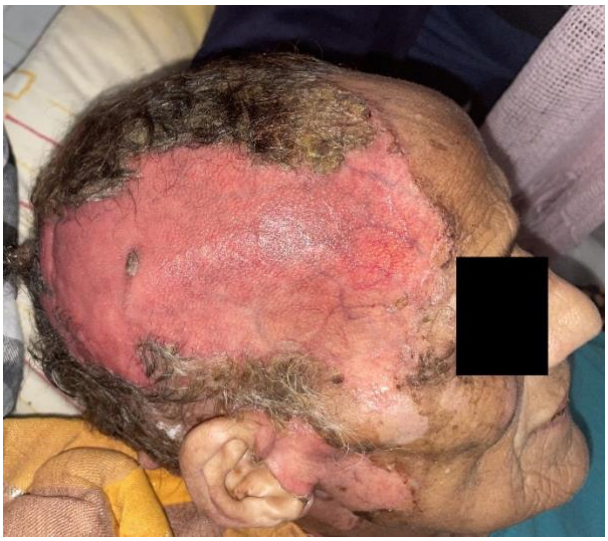

Figure 1: erosive alopecic plaque affecting the right frontal, vertex, and parietal scalp

Discussion

PV accounts for 70% of cases of autoimmune blistering disorders [3]. The classic presentation involves oral mucosal blistering in 80% of patients, followed by skin involvement [2]. However, PV can have different presentations, which can lead to a diagnosis delay. Our case demonstrates an uncommon presentation based on the anatomical site of the initial lesion. Although scalp erosions are frequent, progressive alopecia remains rare, with only 5.4% of PV cases reported with alopecic lesions in a study [4]. Additionally, scalp involvement is less frequent in PV compared to other autoimmune blistering diseases, making diagnosis more challenging [2].

The mechanism of alopecia in PV involves acantholysis extending into the outer root sheath (ORS) of anagen hair follicles, causing hair shaft detachment and loss. Immunopathological analysis has demonstrated intercellular IgG deposits in ORS keratinocytes, confirming the role of autoantibodies in follicular involvement [5]. Notably, scalp alopecia in PV is non-scarring, with hair regrowth occurring after immunosuppressive therapy. However, secondary factors like infections may worsen hair loss. A study reported bacterial colonization in alopecic lesions, suggesting infections may weaken follicular anchorage, leading to alopecia [4].

Scalp involvement in PV correlates with greater disease severity and longer remission time. A prospective study found that patients with scalp involvement had higher PDAI scores, prolonged remission time, and elevated anti-Dsg1 autoantibody levels, suggesting a more severe course requiring aggressive treatment [6].

The differential diagnosis of alopecia with scalp erosions includes lichen planopilaris, discoid lupus erythematosus, and bullous impetigo. The presence of flaccid bullae, histopathological confirmation of suprabasal acantholysis, and direct immunofluorescence positivity for intercellular IgG and C3 deposits help distinguish PV from other inflammatory scalp disorders [5]. Diagnostic tools like trichoscopy and the hair pull test are also useful in assessing scalp involvement in PV. The hair pull test, showing painless extraction of anagen hairs with intact outer root sheath, correlates with disease activity and serves as a non-invasive diagnostic indicator. This phenomenon, called "anagen effluvium," is considered a hair equivalent of the Nikolsky sign [5].

Conclusion

Our patient presented with erosive scalp alopecia as the first manifestation of PV, a rare presentation. Unfortunately, she developed a massive pulmonary embolism within 24 hours of admission, leading to sudden death, preventing further evaluation of her scalp lesions under treatment. This case highlights the need for awareness of atypical PV presentations, especially scalp involvement. Physicians should consider PV in patients with progressive scalp erosions and hair loss, even without classical lesions, to avoid delays and improve outcomes.

ACKNOWLEDGEMENTS

The authors have no acknowledgements to declare and report no conflicts of interest.

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