



SCARRING HAIR LOSS IN A CASE OF PEMPHIGUS VULGARIS

Meenakshi Patial*

Medical Officer [Specialist], Civil Hospital, Nadaun, Himachal Pradesh, India.

***Corresponding Author: Meenakshi Patial**

Medical Officer [Specialist], Civil Hospital, Nadaun, Himachal Pradesh, India.

Article Received on 04/11/2022

Article Revised on 25/11/2022

Article Accepted on 15/12/2022

ABSTRACT

Pemphigus is an autoimmune skin disease characterized by blistering of skin and mucous membranes. There are several types of pemphigus, but the most common one is pemphigus vulgaris, accounting for 70% of all the cases. It most commonly appears between the age group of 30 to 60 years and is more common found in Jews and Indians than in other races, because for genetic reasons or it can be drug induced. Hair loss is an uncommon clinical finding in pemphigus patients. In this case report, a young patient in her twenties presented in the OPD with multiple skin erosions and oral erosions and with multiple patches of hair loss over the scalp.

KEYWORDS: Acantholysis, Alopecia, Desmogleins.

INTRODUCTION

Pemphigus vulgaris is an autoimmune bullous disease that results from the action of autoantibodies against Desmogleins 1 and 3. It is most severe form of pemphigus.^[1] They present chronic evolution, with impairment in quality of life.^[2] Clinically, it may present with mucosal or mucocutaneous involvement. First manifestation is painful oral erosions in 50%-70% of cases and occur in 90% of patients during the course of the disease.^[3] Most patients develop flaccid blisters, resulting in painful erosions that bleed easily, become covered by crusts, with no tendency to heal. Healing is usually without a scar, with pigmentary changes. Scalp is also affected due to the presence of Desmogleins in the hair follicles with erosions and crusted scaly plaques which may progress to alopecia.^[5] For the laboratory diagnosis of PV, Tzanck smear, histopathological examination, direct immunofluorescence or immunohistochemical examination, ELISA can be used. Treatment should be initiated as early as possible with the use of systemic medications such as corticosteroids or when the condition cannot be controlled with corticosteroids alone, or when the patient has clinical contraindications for corticosteroids, adjuvant drugs can be used such as Azathioprine, Methotrexate, Mycophenolate mofetil, Cyclophosphamide, IVIG, Rituximab.

CASE

The patient was a 21-year-old female presented with painful oral erosions associated with difficulty in eating hot and spicy food for 6 months. After 2 weeks, she developed multiple fluid filled lesions over trunk, groin, thighs and scalp. The lesions used to burst after minor trauma within 1-2 days, forming painful raw erosions

which showed little tendency to heal and developed overlying crusting. There was no history of drug ingestion prior to the onset of symptoms, no photosensitivity and no weight loss or loss of appetite. On examination, multiple well-defined erosions of size ranging from 0.5 x 0.5 cm to 5 x 5 cm with overlying brownish adherent crusting were seen over face, neck, chest, axilla, back, abdomen, pubic area and thighs [Figure 1-2]. Direct and indirect Nikolsky sign was negative. Over scalp, erosions of size 0.5 x 0.5 cm with brownish crusting were present with 4 patches of hair loss with scarring and loss of follicular opening [Figure 3]. In oral mucosa, erosions were present over hard palate and lower gingiva. Conjunctival and genital mucosa was normal. Cutaneous ABSIS was 24.5 and oral ABSIS was 2/11 with normal systemic examination. On histopathological examination, superficial perivascular mixed infiltrate of lymphocytes and neutrophils with mild focal spongiosis. In a single focus incipient, suprabasal clefting is seen with occasional partially acantholytic cells. On direct immunofluorescence, IgG 3 + positivity at intercellular space of epidermis was there, confirming the diagnosis of Pemphigus vulgaris.

Figure legends



Figure 1 and 2- Multiple well-defined erosions over chest and trunk, with overlying brownish adherent crusting.



Figure 3: Multiple patches of hair loss with scarring over scalp with crusting.

DISCUSSION

Pemphigus vulgaris patients usually present with fluid filled lesions and erosions of mucous membranes and the skin caused by autoantibodies against Desmoglein 1 and 3.^[6] Scalp erosions are seen in some patients with localized and generalized forms of Pemphigus vulgaris, however, alopecic patches in Pemphigus vulgaris have been rarely reported in the literature.^[7] In a study^[8], it was suggested that the combined effect of anti-desmoglein autoantibody-mediated acantholysis and concomitant infection may cause the weakening of the hair follicle anchorage and result in alopecia. It was

found that acantholysis between keratinocytes of the outer root sheath extended from the infundibulum to the suprabulbar level of the hair follicle in pemphigus vulgaris, whereas in pemphigus foliaceus acantholysis was restricted to the infundibulum.^[9]

REFERENCES

1. Porro AM, Seque CA, Ferreira MCC, Enokihara MMSS. *An Bras Dermatol*, 2019; 94(3): 264-78.
2. Hsu DY, Brieva J, Sinha AA, Langan SM, Silverberg J. Comorbidities and inpatient mortality for pemphigus in the USA. *Br J Dermatol*, 2016; 174: 1290-8.
3. Wojnarowska F, Venning VA. Immunobullous Diseases. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rook's Textbook of Dermatology*. 8th ed. New York: Ed Blackwell Science, 2010; 2: 40.1.
4. Venugopal SS, Murrell DF. Diagnosis and clinical features of pemphigus vulgaris. *Dermatol Clin*, 2011; 29: 373-80.
5. Sar-Pomian M, Rudnicka L, Olszewska M. The Significance of Scalp Involvement in Pemphigus: A Literature Review. *Biomed Res Int*, 2018; 2018: 6154397.
6. Amagai M. Autoimmune and infectious skin diseases that target desmogleins. *Proc Jpn Acad*, 2010; 86: 524-537.
7. Wilson CL, Dean D, Wojnorowska F. Pemphigus and the terminal hair follicle. *J Cutan Pathol*, 1991; 18: 428-431.
8. Veraitch O, Ohshima M, Yamagami J, Amagai M. Alopecia as a rare but distinct manifestation of pemphigus vulgaris. *J Eur Acad Dermatol Venereol*, 2013 Jan; 27(1): 86-91.
9. M. Sar-Pomian, J. Czuwara, L. Rudnicka, and M. Olszewska, "Miniaturization of sebaceous glands: A novel histopathological finding in pemphigus vulgaris and pemphigus foliaceus of the scalp," *Journal of Cutaneous Pathology*, 2017; 44(10): 835-842.