Case Report

A Case Report of *Pemphigus foliaceus*

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ABSTRACT

We report a case of the rarely seen disease pemphigus foliaceus in a 32-year-old female patient with a past medical history of superficial red raised lesion over the body face associated with itching and burning sensation for the treatment which has taken previously and healed pigmentation. She did not respond to topical steroids, and the condition flared up when oral steroids were tapered. Immunofluorescence biopsy and antibody titers can help establish the diagnosis. We discuss the clinical features, pathophysiology, histology, disease course, and treatment of *Pemphigus foliaceus*.

Keywords: *Pemphigus foliaceus*, topical steroids, immunofluorescence.

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INTRODUCTION

Pemphigus encompasses a group of organ-specific, antibody-mediated autoimmune diseases of the skin characterized by keratinocyte detachment that leads to the development of blisters and erosions [1]. The term pemphigus stems from the greek pemphix meaning blister or bubble. Pemphigus is usually divided into 2 major forms depending on blister location: Pemphigus Vulgaris (PV) and pemphigus foliaceus (PF) [2]. In most cases, the disease develops independently, but certain medications can also cause pemphigus to develop. The lesions are usually accompanied by a burning sensation. Differently of pemphigus Vulgaris (PV), in Pemphigus foliaceus (PF) patients do not have mucosal involvement. In histology, there is acantholysis in the upper epidermis [1]. This disease has IGG pathogenic autoantibodies against the cell on the surface of keratinocytes. Desmoglein 3 is focused on in pemphigus Vulgaris while desmoglein 1 is designated in Pemphigus foliaceus and its endemic structure Fogo selvegem [3].

Case report:

A 32 years old woman presented with 3 months back history of superficial red raised lesion over the face associated with itching and burning sensation for the treatment which has taken previously and healed pigmentation. After 2 months started developing fluid-filled lesions over the scalp, upper arm, the chest. On examination of the patient was peeling of skin with erosion and oozing of fluid since 1 month, ulcer over the right elbow joint since 5 days. The patient history was hypothyroidism for 3 years.

The patient was initially started on medication drug intake and topical application for lesions over the face for 3 months, oral and topical corticosteroids –T. deflazacort 12mg, oral and topical antifungals Luliconazole, Candiforce and lumivoid for pigmentation.

Cutaneous examination

The generalized scaly erythematous exfoliation of skin over the bilateral upper limbs and lower limbs is shown in (fig 1).

Diffuse raw erosion and crusting disturbances over the scalp, back, upper chest, upper limbs.

Erythematous exfoliation and peeling of skin over the palms.

Oral mucosa – Diffuse patches of pigmentation.

Laboratory values of patient

Hemoglobin - 7.9 (11.5-16.0 gm%),

RBC count – 3.1 (4.0-5.2 millions/cumm),

total WBC count -5000 (4000-11000 cells/cumm),

platelet count-1.4(1.5-4.5 lakhs/cumm),

mean corpuscular heamoglobin-25.1(27-32 pg.),

RDWcv-14.9(11.6-14.0%),

kidney basic screen -calcium- 7.9(5.0-20.0%),

prothrombin time -15.4(10-15 seconds),

INR-1.14(2.5-3.5)

Ds DNA: positive

SmD1-positive (3+)

Ro-52 recombinant (52): positive (3+)

Ss-B/La: positive (1+)

RNP/SM: positive (3+)

Clinical examination:

Due to seizures episodes, the patient underwent to CT scan of the brain based on the report doctor confirmed that status epilepticus.

Figure 1: Superficial red raised lesion

DISCUSSION

Pemphigus foliaceus is a rare immunobullous illness that can have diffuse or restricted signs it commonly occurs in patients aged 30 to 60 years and affects equally in males and females [4]. It includes six major types of disorder in the pemphigus family. They are pemphigus Vulgaris, pemphigus herpetiformis, IgA pemphigus, paraneoplastic pemphigus, drug-induced pemphigus, and pemphigus foliaceus. Pemphigus foliaceus has three subtypes: 1) classic or sporadic pemphigus foliaceous, 2) pemphigus erythematosus, 3) endemic pemphigus foliaceus [5]. The three subtypes are present with identical histological findings. The visualization of pemphigus foliaceus is better compared to pemphigus Vulgaris most likely given the more superficial nature of the blistering process [6]. It occurs due to the loss of Malpighian cells which binds to the IgG+/- C3 antibody against a polypeptide antigen complex on the keratinocyte surface [7]. The most common sites are the scalp, face, trunk, upper and lower limbs, the Generally mucous membrane is not involved [8]. Triggering factors include UV exposure, drugs, and various infections [9].

CONCLUSION

We report a case of a pemphigus foliaceus 32-years-old woman. Based on the clinical presentation of this condition is characterized with other forms of pemphigus. It is a harmless illness of the long term that reacts to treatment with corticosteroids which the patient may ultimately recover. Steroids suppress cutaneous lesions and improve well-being, but a long period of steroid therapy is not indicated.

REFERENCES

1. A case report of pemphigus foliaceus exuberant in a 30-year-old man, Journal of the American Academy of Dermatology, 2018;66(4):AB102.

2. Ashley Walker, DO; Tracy Favreau, DO, Localized Pemphigus Foliaceus, 2017;99:31-39.

3. James KA, Culton DA, Diaz LA. Diagnosis and clinical features of pemphigus foliaceus. *Dermatol Clin*. 2011;29(3):18-24.

4. Tyler Vukmer, DO, John Hassani, DO, Adriana Ros, DO Pemphigus Foliaceus in an Otherwise 35 years old man, dermatology residency, Hackensack university medical centre, 2021;39(3):9-13.

5. Claire Galambrun, Frédéric Cambazard, Catherine Clavel, Pascale Versini, Jean Louis Stéphan, Pemphigus foliaceus, Archives of Disease in Childhood 1997;77:255–257.

6. Melchionda V, Harman KE. Pemphigus Vulgaris and pemphigus foliaceus: an overview of the clinical presentation, investigations, and management. Clin Exp Dermatol. 2019 Oct;44(7):740-746.

7. Kridin K. Pemphigus group: overview, epidemiology, mortality, and comorbidities. Immunol Res. 2018 Apr;66(2):255-270.

8. Orne CM, Hale CS, Meehan SA, Leger M. Pemphigus foliaceus. Dermatol Online J. 2014 Dec 16;20(12):41-50.

9. Galambrun C, Cambazard F, Clavel C, Versini P, Stéphan JL. Pemphigus foliaceus. Arch Dis Child. 1997 Sep;77(3):255-7.

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