Localized pemphigus foliaceus with unilateral facial involvement: A case report

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Pemphigus foliaceus is an autoimmune blistering condition more often than not associated with generalized lesions. Localized Pemphigus foliaceus is a rare entity. Herein, we report a case of pemphigus foliaceus localized on the face.

A 64-year-old woman was referred to our clinic with a 2-year history of erythematous plaque on the right side of her face. The histology report was compatible with pemphigus foliaceus. The lesion improved completely in response to oral prednisolone at 40 mg daily.

Pemphigus foliaceus has to be suspected in presumed eczema that is refractory to appropriate topical corticosteroid treatment. A histopathologic study is required to establish localized pemphigus foliaceus.

Keywords: Localized pemphigus foliaceus, face, corticosteroid, case report.

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INTRODUCTION

Pemphigus foliaceus is an autoimmune condition ultimately entailing acantholysis and blister formation within the epidermis [1]. Pemphigus foliaceus has low incidence and prevalence; hence a rare disease [2]. Pemphigus foliaceus lesions does not frequently spread over the whole integument of body [3]. Moreover, only a few cases of pemphigus foliaceus localized on the face have been reported to date. In the present research, a case of pemphigus foliaceus localized on the face is reported.

CASE REPORT

The patient was a 64-year-old woman referred to our clinic with a 2-year history of erythematous plaque on the right side of her face. The patient did not make mention of any previous surgery, irradiation, trauma prior to lesion appearance and pruritus. Diagnosed as eczema, the lesion had been treated unsuccessfully with topical corticosteroid ointment.

The physical examination revealed an erythematous plaque with scales and crusting on the right cheek (Figure 1), on which a skin biopsy was performed.

The histology report revealed that vacuoles led to the formation of cleft-shaped space in the granular layer that contained acantholytic cells within the cleft. Further observed were hydropic degeneration within the basal layer and lymphocyte infiltration between dermis and epidermis. The histopathology report was compatible with pemphigus foliaceus (Figure 2). The diagnosis was corroborated by direct and indirect immunofluorescence, while antinuclear antibodies (ANA) tests were negative.

The patient underwent oral corticosteroid therapy, and as there was no prior drug application, according to the patient, the lesion had relapsed

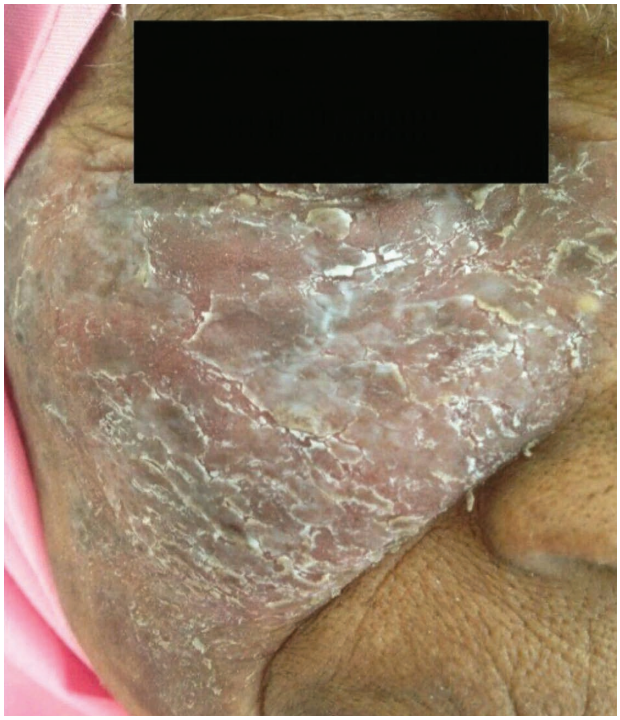

Figure 1. A plaque with scales and crusting on the right cheek.

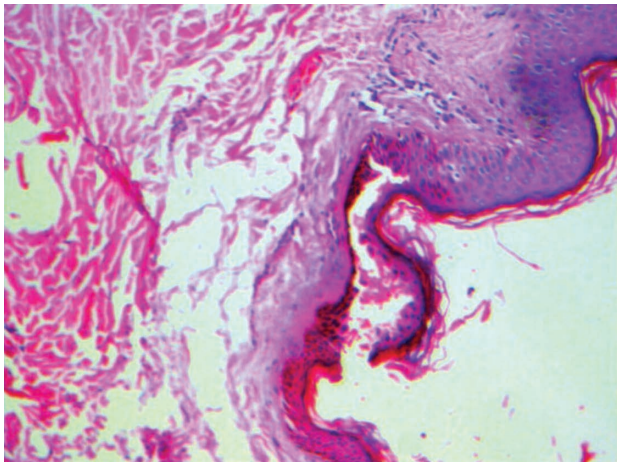

Figure 2. Histopathology of lesion shows that epidermis is mildly hyperplastic, with overlying focal parakeratosis and certain orthokeratosis. There is a superficial bulla with the split high in the epidermis. Dyskeratotic cells with hyperchromatic nuclei are observed in the granular layer. (H&E ×100)

after a year. Afterwards, the lesion improved completely in response to oral prednisolone at 40 mg daily. Thereafter, prednisolone was tapered and the lesion was resolved in the following month.

DISCUSSION

Pemphigus is known as a group of autoimmune diseases causing oral cavity and blisters on the skin [1]. Such condition is characterized by the existence of autoantibodies directed against Dsg3 and/or Dsg1 that ultimately leads to acantholysis [4]. Pemphigus vulgaris, pemphigus foliaceus, and paraneoplastic pemphigus are the three major forms of pemphigus [5]. Pemphigus foliaceus is typically presented as a generalized disease, whereas localized manifestations of pemphigus are primarily less common [6].

Only a few cases of localized pemphigus foliaceus (Table 1) have been reported in the literature [7]. In our patient, the lesion did not spread and was localized on the right side of the face.

Although several hypotheses have been put forth to date, the pathophysiology of pemphigus is yet to be fully understood [3]. Among the most common drugs that can trigger pemphigus foliaceous, mention can be made of penicillamine, and angiotensin-converting enzyme inhibitors (ACEIs) such as captopril [1]. The patient in the present study did not report any prior use of the foregoing drugs.

The differential diagnosis of pemphigus foliaceus includes lupus erythematosus, drug eruption, bullous impetigo, IgA pemphigus and seborrheic dermatitis [2].

In contrast to this study, pemphigus foliaceus more frequently than not occurs in middle-aged people. The fact is that the presumed eczema did not respond to appropriate topical corticosteroid treatment and the persistence of the lesion prompted

Table 1. Previous cases of pemphigus foliaceus localized to the face.

Author	Age	Sex	Location	Treatment	Follow up
Maderal et al [7]	19 y/o	Female	Right cheek and temple	Remission with Prednisone	18 months
Lin et al [8]	53 y/o	Female	Left side of face	Prednisone	N/A
Yamamoto et al [9]	81 y/o	Female	Right cheek	Minocycline, Nicotinamide Betamethasone Valerate	N/A
Zaraa I et al [2]	34 y/o	Female	Right cheek	Prednisone + Cyclophosphamide	16 months*
Our case	64 y/o	Female	Left side of face	Remission with Prednisolone	12 months

N/A: none Y/o:Years old

*They reported three cases of localized pemphigus foliaceus and the patients presented with no extension after an average period of 16 months (6 months to 3 years). Only one of the cases was localized to the face.

us to suspect an immunobullous disorder. In pemphigus foliaceus, the sensitivity of ELISA test of anti-desmoglein-1 antibodies is as high as 97.9% [10].

Our patient had a negative ELISA, yet the indirect immunofluorescence results were positive.

Prior to the introduction of systemic corticosteroids, currently the mainstay of treatment for pemphigus [7], the fatality rate of pemphigus was 60% [5]. Other medications have been used to treat localized pemphigus foliaceus. For instance, G. Tyros et al., used topical Pimecrolimus to treat pemphigus foliaceus lesions localized on the face and scalp area [5]. In addition, C. C. Termeer et al., made use of topical tacrolimus for the treatment of pemphigus foliaceus localized on the scalp area [11]. The patient in the present investigation responded to the oral corticosteroid and the lesion was completely ameliorated after a month.

CONCLUSION

This case indicated that, although rare, pemphigus foliaceus can occur locally, and is to be suspected in presumed eczema, which is refractory to appropriate topical corticosteroid treatment. Histopathologic studies are further required to establish localized pemphigus foliaceus.

Conflict of Interest: None declared.

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