Case Report

Concurrent of Pemphigus Vulgaris and Pemphigus Foliaceus in the Same Patient : Rare Case

Cut Putri Hazlianda[1], Angela Fovina[2]

Abstract

Easily ruptured superficial vesicles or bullae are a hallmark of the chronic autoimmune illness pemphigus. The two primary subtypes of pemphigus are pemphigus vulgaris (PV) and pemphigus foliaceus (PF), which can infrequently coexist or even change from one to the other. Changes in the autoantibody profile for desmoglein that are both qualitative and quantitative may help to explain this process. We describe a case of PF and PV present at the same time on a 73-year-old woman from clinical findings revealed multiple flaccid bullae on the face, neck, left and right arm and thigh, stomach, and back. Erosions, excoriations, and crusts were present. Nikolsky's sign was positive. Her histopathological examination showed subcorneal and suprabasal acantholysis. Basal cells were formed in a "*row of tombstones*" appearance.

Keywords: Pemphigus vulgaris; pemphigus foliaceus.

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Introduction

Pemphigus vulgaris (PV) and pemphigus foliaceus (PF) both have autoimmune etiology that targets desmoglein (Dsg).[1,2] The two primary kinds of pemphigus are PV and PF, which only rarely cooccur or even switch from one to the other. Changes in the desmoglein autoantibody profile on both a qualitative and quantitative level may help to understand this process.[3] Clinical examination triggers the diagnosis of pemphigus; histological investigation of tissue collected from a lesional area and demonstrating acantholysis is needed to confirm the diagnosis. The "gold standard" for confirming the diagnosis is direct immunofluorescence investigation of biopsy samples. indirect immunofluorescence (IIF) and enzyme-linked immunosorbent tests (ELISA) are used to identify autoantibodies that attack epithelial cells. Treatment of PV and PF is systemic corticosteroids.[4-6]

Case Report

Since two months, a 73-year-old woman has had blisters that easily rupture and agony all over her body. She had a history of oral mucosal erosions 3 months ago. Otherwise, there is no history of food or drug allergy, and no previous history of skin diseases in this patient. Based on the physical examination, she was moderately ill, vital signs were within normal limits. Dermatological examination revealed multiple flaccid bullae on the face, neck, left and right arm and thigh, stomach, and back. Erosions, excoriations, and crusts were present (Fig. 1). Nikolsky's sign was positive. Histopathological examination showed subcorneal and suprabasal acantholysis (Fig 2). Basal cells were formed in a "*row of tombstones*" appearance. She was diagnosed as PV and PF. The patient was admitted to the hospital and was given systemic Methylprednisolone 62.5 mg/24 hours, Ranitidine 50 mg/12 hours, NaCl compress 0.9% for 15 minutes every 6 hours, topical Fucidic Acid and topical Desoxi-

methasone 0.25% twice a day. After 2 weeks of therapy, the skin condition improved, no new vesicles or bullaes. Dermatological examination revealed hyperpigmented macules on the face, left-right arm, and thigh, and back. Erosions, excoriations, and crusts were present (Fig. 3). The patient was then treated with oral Methylpredni-solone 8 mg-8, oral Ranitidine 150 mg twice a day, NaCl compresses 0.9% for 15 minutes every 6 hours, topical Fusidic Acid, and topical Desoximethasone 0.25% twice a day. The prognosis is quo ad vitam dubia ad malam, quo ad functionam dubia ad bonam, and quo ad sanactionam dubia ad bonam for this patient.

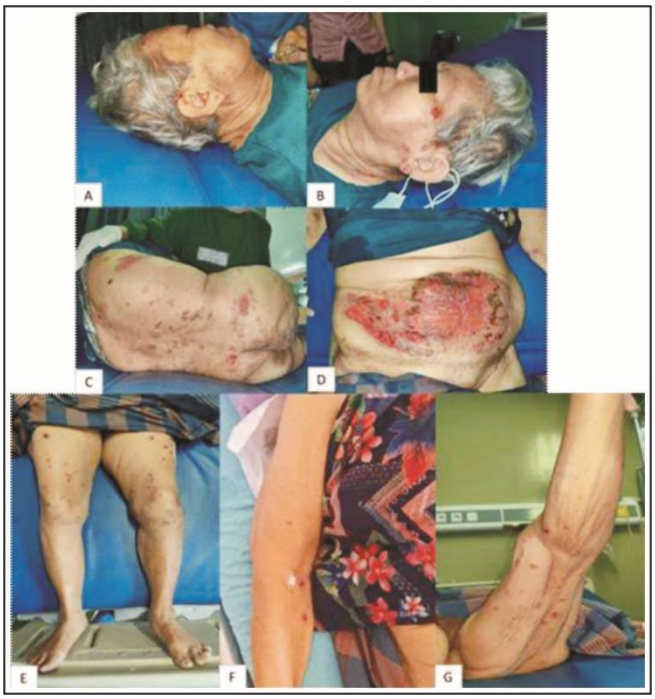

Figure. 1 (A-G): Multiple flaccid bullae on the face, neck, left and right arm and thigh, stomach, and back. Erosions, excoriations, and crusts were present.

Discussion

There are not many examples of concurrent PV and PF, according to a literature search. PV produces blisters in the suprabasal layers of the epidermis. The target antigen for PV is Dsg3. Because Dsg1 is the target antigen in PF, acantholysis only occurs in the higher layers of the epidermis.[7]

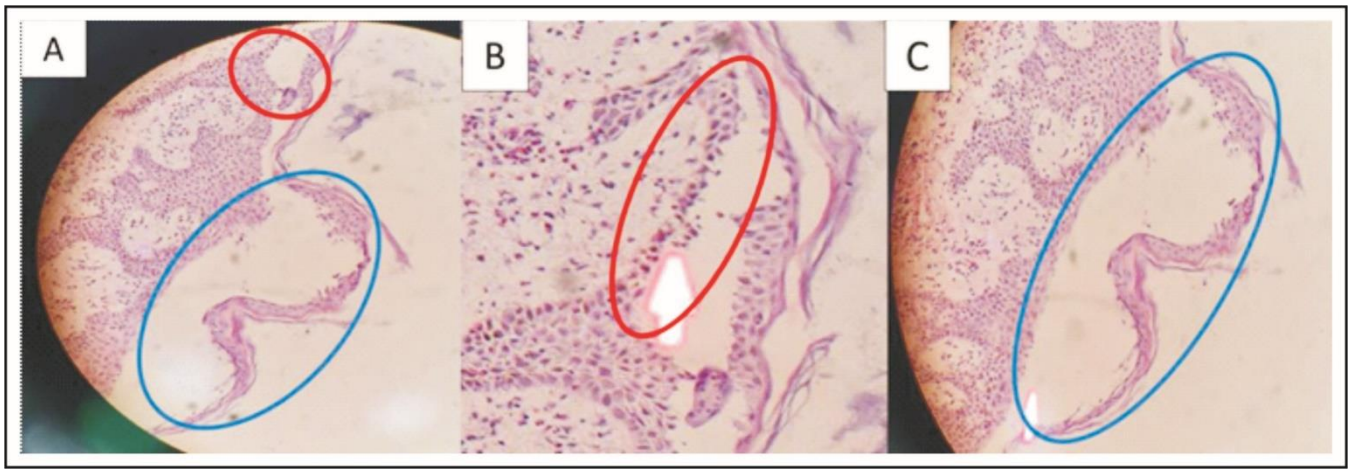

Figure. 2 (A): Histopathological examination showed subcorneal and suprabasal acantholysis. **(B):** Basal cells were formed in a "*row of tombstones*" appearance in PV. **(C):** Subcorneal acantholysis in PF (**B & C):** Hematoxylin-eosin stain; original magnification: x200).

Almost 60% of PV patients also exhibit circulating Dsg1 autoantibodies without any PF-like lesions or clinical signs.[3] It is unusual for PV and PF to be contemporaneous. This contemporaneous scenario is a reflection of qualitative and quantitative alterations in the profile of Dsg1 and Dsg3 autoantibody-developed autoantibodies.[1] Though the exact method is unknown, epitope spreading has been suggested as one possible explanation.[3]

Our patient was diagnosed with PV from history of oral mucosal erosions, dermatological examination revealed multiple flaccid bullae causing erosions, and histopathological examination of suprabasal acantholysis with "row of tombstones"

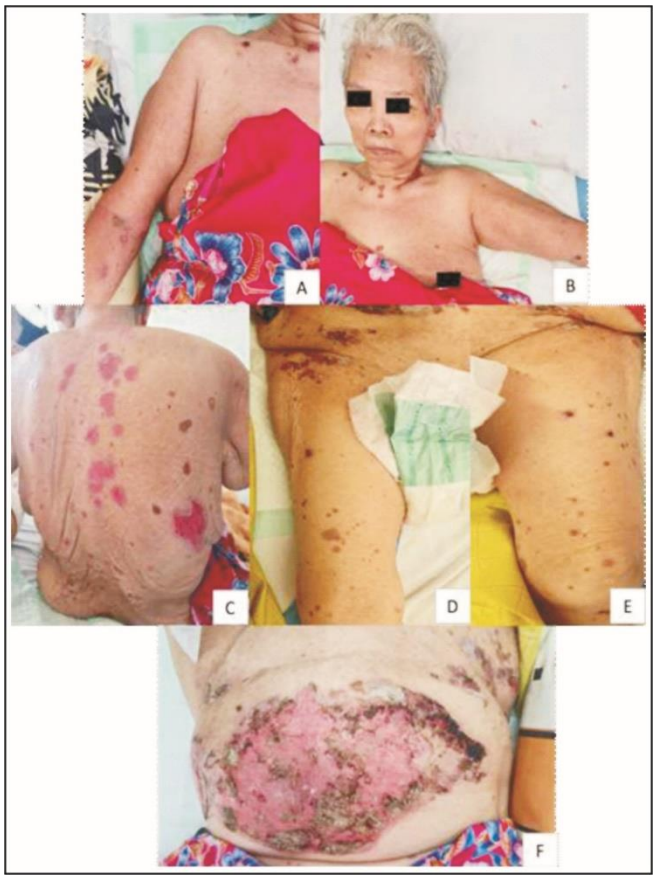

Figure 3 (A-F): Hyperpigmented macules on the face, left-right arm, and thigh, and back. Erosions, excoriations, and crusts were present.

appearance. This case also matched with PF by histopathological examination showed subcorneal acantholysis. First line therapy of pemphigus is systemic corticosteroids.[8,9] The patient has a quo ad vitam dubia ad malam prognosis because age of onset 65 years in PV and PF is associated with mortality.[10]

Conclusion

We described a rare instance of PF and PV co-existing at the same time that was determined by clinical and histological findings, including subcorneal and suprabasal bullae associated with the etiology of both PF and PV illness. Although the exact mechanism is not entirely understood, epitope spreading has been suggested.

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Author's Contribution:

CPH: Conceived, designed, edited the manuscript, given final approval of the version to be published, critical revisions

AF: Manuscript writing, final approval of the version to be published, agree to be accountable for all aspect of the work.

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