Localized Pemphigus Foliaceous: A case report

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ABSTRACT:

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Pemphigus Foliaceous (PF) is a superficial pemphigus causing by a pathologic autoantibody to desmoglein 1 protein which functions in intercellular adhesion, mainly in the superficial part of epidermis. Lesions usually distributed in the seborrheic area. Only few cases of localized PF have been reported in English literatures. We reported here a case of 17-year-old Thai girl who presented with multiple grayish-brown hyperpigmented discoloration on the left side of face. Careful physical examination revealed a primary lesion of scaly purplish-brown plaque on the left paranasal area, of which histopathology showed intragranular acantholysis. Direct immunofluorescence (DIF) study showed intercellular IgG deposition, confirmed a diagnosis of localized pemphigus foliaceous. The patient was treated with topical steroids for the active PF lesion and bleaching agents for hyperpigmented areas. After treatment, active lesion slowly resolved but hyperpigment lesions improved only minimally and new lesions still infrequently developed.

Key words: Pemphigus foliaceous, superficial pemphigus, localized

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บทคัดย่อ :

กชมล วนดุรงค์วรรณ, เบ็ญจ์สชีว์ ปัทมดิลก รายงานผู้ป่วยโรค LOCALIZED PEMPHIGUS FOLIACEUS วารสารโรคผิวหนัง 2558; 31: 127-131.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

Pemphigus Foliaceus (PF) เป็นโรคตุ่มน้ำผิวหนังในชั้นตื้น เกิดจากการมีแอนติบอดีต่อโปรตีน desmoglein 1 ซึ่งมีหน้าที่สำคัญในการยึดเกาะระหว่างเซลล์ผิวหนังในชั้นหนังกำพร้า โดยเฉพาะส่วนบน รอยโรคมักกระจายทั่วบริเวณผิวหนังที่มีต่อมไขมัน ผู้ป่วย PF ที่มีรอยโรคเฉพาะที่ (localized) พบได้น้อย รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงอายุ 17 ปี มาตรวจรักษาด้วยอาการสำคัญ คือ รอยคล้ำสีเทาอมน้ำตาล หลายตำแหน่ง ที่บริเวณใบหน้าด้านซ้าย จากการตรวจร่างการโดยละเอียด พบรอยโรคปฐมภูมิที่มีลักษณะเป็นปื้นนูนสีม่วงอมน้ำตาลร่วมกับมีขุยเล็กน้อย ที่บริเวณข้างจมูกด้านขวา ซึ่งผลตรวจทางพยาธิวิทยาพบ acantholysis ในชั้น granular layer ร่วมกับผลการตรวจ Direct immunofluorescence (DIF) พบการสะสมของ IgG ที่บริเวณ intercellular ทำให้สามารถยืนยันผลการวินิจฉัยว่าเป็น localized PF ผู้ป่วยรายนี้ได้รับการรักษาด้วยยาทาสเตียรอยด์สำหรับรอยโรคบริเวณข้างจมูก ร่วมกับ ยาฟอกผิวขาวทาบริเวณรอยคล้ำ ภายหลังรักษา รอยโรคดีขึ้นเล็กน้อย แต่รอยคล้ำดีขึ้นน้อยมาก และยังมีรอยโรคใหม่เกิดขึ้นเป็นครั้งคราว

คำสำคัญ: โรคตุ่มน้ำพองใส โรคตุ่มน้ำพองใสชนิดตื้น เฉพาะที่

Introduction

Pemphigus is a group of autoimmune vesiculobullous skin diseases, comprising pemphigus vulgaris (PV), pemphigus foliaceous (PF), pemphigus erythematosus(PE), IgA pemphigus, and paraneoplastic pemphigus. These diseases are mediated by autoantibodies targeting specific intercellular adhesion proteins. The different targeted autoantigens among each particular types contribute to the different planes of epidermal separation. Pemphigus foliaceous is a superficial type of pemphigus characterized by a pathogenic antibody targeting desmoglein 1 (Dsg1). Lesions are typically widespread in the seborrheic areas of the face, scalp and upper trunk.[1, 2] However, PF can rarely manifest as localized lesions.[3] We reported here a case of localized PF that skin lesions were repeatedly developed, resolved and recurred in the same area of the face and scalp for ten years

Case report

A 17-year-old Thai girl who visited Institute of Dermatology with a chief complaint of multiple purplish-brown hyperpigmented patches on the left side of face and scalp. Her problem was spontaneously developed ten years ago. She denied history of local tissue injury; such as burn or local trauma, and did not use any topical medication before skin lesions developed. Each lesions were primarily occurred as scaly plaques that slowly resolved with residual

hyperpigmentation. She has never been observed any vesicle or bulla. She was treated by several doctors but the lesions improved only slightly. A thorough physical examination revealed multiple ill-defined greyish brown hyperpigmented patches on the left side of face and scalp (Figure 1) with a localized area of purplish-brown scaly plaque on the left paranasal area (Figure 2). Provisional differential diagnoses included localized morphea, psoriasis and seborrheic dermatitis. A skin biopsy was done. Histopathological study revealed intragranular acantholysis (Figure 3). Direct immunofluorescence study showed IgG depositions in intercellular pattern. Stainings for IgM, IgA, C3 and fibrinogen were negative, same as the result of indirect immunofluorescent study (IIF). She was diagnosed as localized pemphigus foliaceous with post-inflammatory hyperpigmentation. She was treated with topical corticosteroids for erythematous lesions and bleaching agent with sunscreen for hyperpigmented areas. Lesions were slightly improved. Unfortunately, new lesions were continuingly developed.



Figure 1 Multiple ill-defined grayish-brown hyperpigmented patches on the left side of face

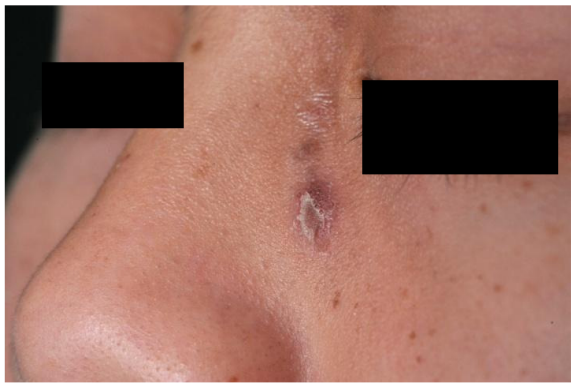


Figure 2 A solitary well-defined purplish-brown scaly plaques on left paranasal area.

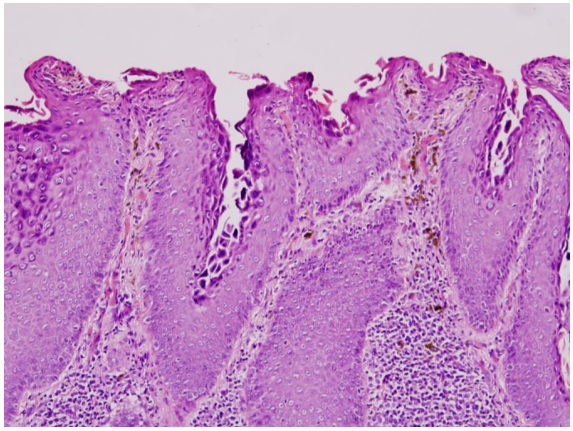


Figure 3: Intragranular acantholysis of epidermis with dense superficial perivascular lymphohisteocytic infiltrations. (H&E, original magnificationx400)

Discussion

Pemphigus foliaceous is an autoimmune blistering skin disease characterized by a pathogenic antibody targeting desmoglein 1 (Dsg1), a desmosomal cadherin that functions in cell-cell attachment mainly in the superficial part of epidermis. Disrupted Dsg1 adhesion resulting in a separation in granular layer, which can be seen as fragile blisters that oftenly ruptured before being recognized. Consequently, patients more commonly present with erythematous scaly plaques rather than vesicles or bullae.[2] PF lesions are typically widespread in the seborrheic areas of the face, scalp and upper trunk, correlated with areas of Dsg1 distribution.[4] However, localized PF has been reported in 13 cases. These patients commonly acquired lesions in limited areas of the face and/or scalp and were unilateral.[3] The pathophysiology of the localized distributions are not clearly established.[3] Some explained by lower level of circulating autoantibody[5] correlated with the result of IIF and ELISA that were not infrequently negative in these patients.[3,6] In two cases, lesions appeared after the use of topical medications i.e. Topical NSAIDs and topical imiquimod.[7] Other possible triggering factors include local injury and sun exposure.[8, 9] However, in the remaining cases, lesions occurred spontaneously. Histopathological findings were identical to those of the generalized PF, composing an intraepidermal separation within subcorneal and intragranular layers. Direct immunofluorescence study showed positive intercellular antibody depositions in all cases, however indirect immunofluorescent tests and ELISAs can detected only 62.5% and 50% of these cases respectively.[3] The treatment of PF is mainly based on topical corticosteroids and other symptomatic treatments such as antihistamines. Systemic treatment is not required unless the disease is widespread.[1] In most cases, high potency topical steroids were suffice in controlling localized PF lesions. However, in some cases, treatment with oral prednisolone and other immunosuppressive drugs were required to control the disease progression or prevent the relapse. Prognosis of localized PF was generally favorable. Although in three cases, the disease progressed to generalized PF. [3] For the disease associations, generalized PF carried a clear association with myasthenia gravis and thymoma[10], but neither disease has been observed in patients with localized variant.

In our patient, the peculiar disease distribution gave

rise to the differential diagnoses of other diseases included localized morphea and seborrheic dermatitis. After tissue biopsy was done, microscopic results were clearly compatible with localized pemphigus foliaceous. The diagnosis was confirmed by positive result of direct immunofluorescence. Since IIF and ELISA can be negative in half of localized PF patients, the negative IIF titer in our patient did not rule out the diagnosis and ELISA study was not done. For the management, as the severity is mild and new lesions occur infrequently, her disease was controlled by only moderate potent topical corticosteroids and oral antihistamines. However, hyperpigmented lesions were improved only slightly by topical bleaching agents and sunscreen.

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