Seborrheic pemphigoid

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INTRODUCTION

Bullous pemphigoid (BP) is the most common autoimmune subepidermal vesiculobullous disease usually affecting older patients.[1] In BP, there is no ethnic or racial predilection and it affects both men and women equally.[2-5] Based on the reported data in the Philippine Dermatological Society Health Information System, the incidence of BP for 2014 is 0.0009 per 100,000,[6] which is low compared with the 0.2 to 3 per 100,000 in developed countries such as the United Kingdom.[7] Precipitating factors include intake of drugs such as furosemide, phenacetin, enalapril, and ibuprofen; radiation therapy; ultraviolet A and B exposure; burns; surgery; and organ transplantation.[8] BP is typically characterized by mild to intractable pruritic macules, patches, and plaques (urticarial stage) eventually evolving into tense serous or hemorrhagic vesicles and bullae with excoriations and erosions (bullous stage) typically seen on flexural areas, lower abdomen, and lower extremities.[1] Nikolsky and Asboe-Hansen signs are usually negative.[9] In some cases, BP presents clinically as atypical variants including dyshidrosiform and intertrigolike, prurigo nodularis–like, eczematous, localized, and erythrodermic forms.[10-18] In addition to these atypical presentations, BP may be seen on seborrheic areas (scalp, pre- and postauricular, auricular, chest) and is referred to as *seborrheic pemphigoid*, which has very few reported cases.[19] Here, we report a case of seborrheic pemphigoid in a 67-year-old Filipino man.

Abbreviations used:

BP: bullous pemphigoid
SP: seborrheic pemphigoid

CASE REPORT

We present a case of a 67-year-old Filipino man who is hypertensive, diabetic, and dyslipidemic who is on the following maintenance medications: olmesartan plus hydrochlorothiazide, metformin, aspirin, and simvastatin for more than 5 years. He had no history of seborrheic dermatitis. He presented with a 4-week history of erythematous, severely pruritic, recurrent macules and patches on the cheeks and chest topped with clear fluid-filled vesicles and bullae spreading to the pre- and postauricular areas, mandibular areas, and scalp. A few scattered lesions also appeared on the arms and thighs. Persistence of the lesions prompted a visit to the outpatient department of a tertiary hospital in Makati City, Philippines. Physical examination found a few tense, clear fluid-filled vesicles and bullae on the right preauricular area, chest, and sole of the right foot (Fig 1). Several erythematous erosions were also noted on the scalp, pre- and post-auricular areas, malar areas, mandible, and chest, with a few on the arms and thighs (Fig 2). Nikolsky and Asboe-Hansen signs were negative. No other significant skin or mucosal lesions were appreciated.

A 4-mm skin punch biopsy specimen from the edge of the blister on the chest was submitted for histologic examination (hematoxylin-eosin stain), which found a subepidermal blister with a superficial perivascular lymphohistiocytic and eosinophilic infiltrate (Fig 3). Direct immunofluorescence of perilesional skin from the chest showed linear deposits of IgG (+1) and C3 (+2) in the basement membrane zone (Fig 4). Serum enzyme-linked immunosorbent assay to BP180 was positive at 73.45 (cutoff value for positivity, 9 U/mL). Based on the clinical distribution and histologic and

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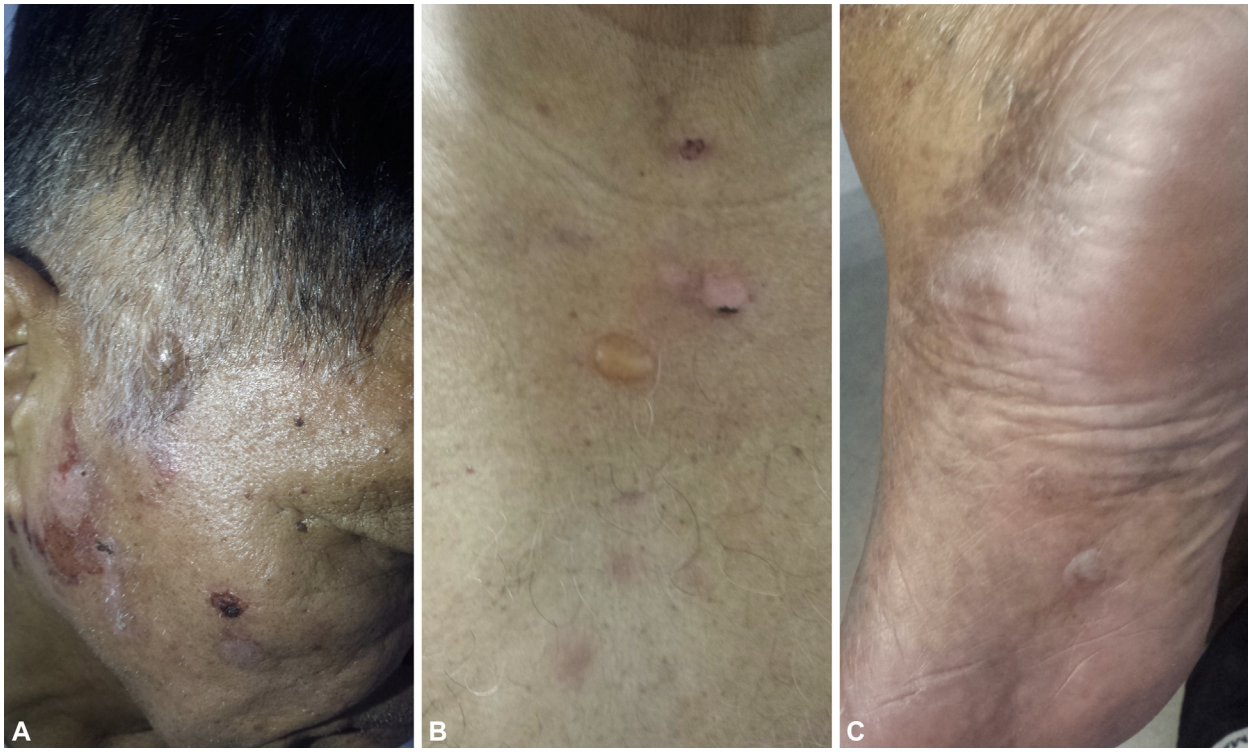

Fig 1. Few tense, clear, fluid-filled vesicles and bullae on the (**A**) right preauricular area, (**B**) chest, and (**C**) sole of right foot.

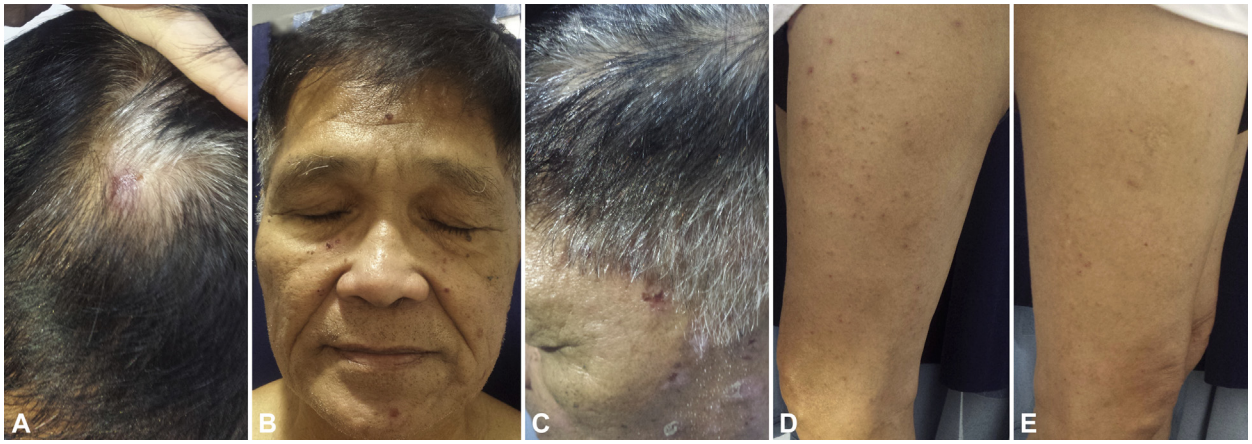

Fig 2. Erythematous erosions on the (**A**) scalp, (**B**) malar areas and mandible, (**C**) pre- and postauricular areas, and (**D** and **E**) a few on the thighs.

immunologic findings, seborrheic pemphigoid was diagnosed. Treatment with prednisone at a low dose of 0.2 mg/kg/d (gradually tapered), together with niacin, 1500 mg/d, and doxycycline, 200 mg/d, produced significant improvement with clearing of lesions and symptoms. After 7 weeks, the patient achieved complete remission and has stayed clear for 2 months with no maintenance medications.

DISCUSSION

Seborrheic pemphigoid (SP) is a very rare variant of BP with only 4 cases reported in the literature. The first reported case was an elderly woman described by Schynder in 1969. SP is characterized by vesicles, bullae, and erosions involving the seborrheic areas of the body.[19,20] Goldberg et al[21] found that in BP, expression of the BP antigens was variable, with greatest levels on sites of predilection and lowest

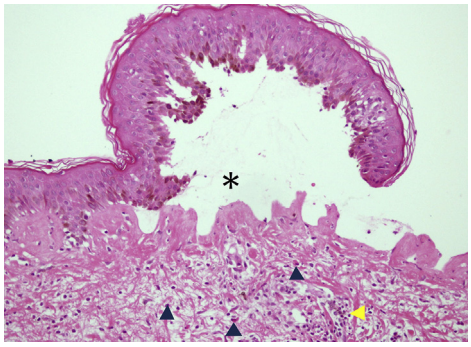

Fig 3. Subepidermal blister (asterisk) with superficial perivascular lymphohistiocytic (yellow triangle) and eosinophilic infiltrate (blue triangle). (Hematoxylin-eosin stain; original magnification: ×20.)

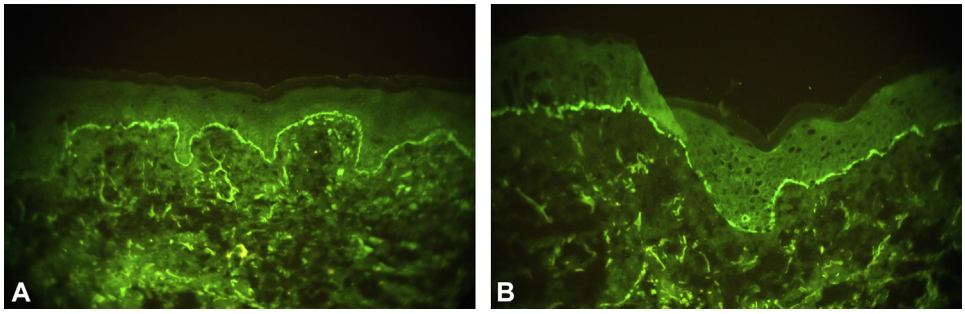

Fig 4. IgG direct immunofluorescence of perilesional skin shows linear deposits of (**A**) IgG (+1) and (**B**) C3 (+2) in the basement membrane zone.

levels on the scalp, face, and extensor arms. In SP, it is possible that expression of BP antigens is high at the seborrheic areas producing the typical vesicles and bullae. Other findings suggest that the presence of *Malassezia* and the excess sebum production on seborrheic areas may contribute to the development of SP via koebnerization, acting as a trigger for the disease. In addition, *Malassezia* is known to activate the classical or alternative complement pathways, possibly augmenting the complement-mediated inflammation seen in typical BP.[19,20] Because of the rarity of this disease, there are no specific established treatment regimens. However, according to Errichetti et al,[19] treatment with low-dose systemic corticosteroids is effective in SP. Our patient started oral prednisone at 0.2 mg/kg/d with a subsequent taper.

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