A case of pemphigus vegetans occurring at an unusual site and followed by gastric cancer

Dear Editor,

Pemphigus is a rare autoimmune bullous disorder involving skin and mucous membrane. Pemphigus vegetans is a rare variant of pemphigus and accounts for 5% of all pemphigus cases.1 It is typically characterized by vegetating plaques on skin folds and mucous membranes. The folds involved mainly include the axillary, inguinal, perianal, submammary, and umbilical areas. The diagnosis is based on clinical, histopathological, and immunofluorescence studies. We present a case of pemphigus vegetans occurring on the left upper arm in a patient who also developed gastric cancer.

A 68-year-old woman with a history of old stroke was seen at the dermatology clinic complaining of a 3-month history of a gradually enlarging skin lesion on her left upper arm. Neither itching nor pain was noted. The lesion was an oval scaly vegetating erythematous plaque of 3 cm × 4 cm (Figure 1). No other cutaneous or mucosal lesions were found. The initial clinical differential diagnosis included deep fungal infection, nontuberculous infection, and Bowen's disease. A skin biopsy specimen was taken for pathologic study and the specimen was also sent for fungal and mycobacterial cultures. The histopathological examination showed mild parakeratosis, focal erosion, marked acanthosis with intraepidermal abscesses containing acantholytic cells, eosinophils and neutrophils, heavy eosinophilic and neutrophilic exocytosis, marked papillomatosis with congested vessels, and heavy perivascular infiltrates of lymphocytes, plasma cells, and numerous scattered eosinophils, and neutrophils in dermis (Figure 2A and B). No fungi were found in the horny layer by the Periodic acid-Schiff (PAS) stain. Tissue cultures for fungi and mycobacteria were negative. The diagnosis of pemphigus vegetans was confirmed by direct immunofluorescence study, which showed positive stainings of immunoglobulin G (IgG) and 3rd component of complement (C3; Figure 2C and D). She was treated with intramuscular dexamethasone phosphate 5 mg weekly and with topical steroids twice a day. Her skin lesion gradually subsided. However, 1 month after the diagnosis of pemphigus vegetans, she started to suffer from poor appetite and vomiting. She was found to have gastric adenocarcinoma 5 weeks after the diagnosis of pemphigus vegetans and received partial gastrectomy. Her clinical condition deteriorated after the operation and she died of pneumonia 1 month later.

Pemphigus vegetans can occur in people of all age groups, and the median age of onset is 40–50 years. Two clinical subtypes have been described based on clinical features and courses. Patients with the Neumann type often begin with flaccid bullae that erode to form secondary hypertrophic papillated plaques and require higher doses of systemic corticosteroids with protracted courses and frequent relapses. The relatively benign Hallopeau type starts as pustules as the primary lesions and followed by vegetations. It requires lower doses of systemic corticosteroids and usually has a prolonged remission.2 However, in a recent series of pemphigus vegetans,3 no significant difference was noted between these two clinical subtypes regarding relapses, wound healing period, survival, and death. Because we did not know the patient’s initial presentation of the skin lesion, we could not subclassify the present case.

Intertriginous areas and mucous membranes are the most frequently involved sites for pemphigus vegetans. The involvement of nonintertriginous areas is extremely rare. Only a few cases with solitary lesions limited to the scalp, face, and soles have been reported.1,2,4,5 A case with involvement of nonintertriginous areas including the trunk and right upper arm has also been reported.6 Our case occurred on the upper arm exclusively.

Figure 1 One oval-shaped, scaly vegetating erythematous plaque on the left upper arm.
The pathogenesis of pemphigus vegetans remains unknown. The frequent involvement of intertriginous areas may be attributed to relative occlusion and maceration with subsequent bacterial infection, suggesting a response to superinfection. However, this could not explain the skin lesions of nonintertriginous areas.

It is clear that patients with pemphigus vulgaris have an increased risk of incidence of internal malignancy. Pemphigus may precede or follow malignant tumor. Coexistence of pemphigus vegetans and neoplasms are rarely reported. Pemphigus vegetans associated with lymphoproliferative disorders (non-Hodgkin lymphoma and chronic lymphocytic leukemia) and internal malignancy (lung and gastric cancer) have been described. Koga et al reported a case of pemphigus vegetans with coexisting gastric cancer. In contrast to our case, their patient typically presented with vegetating and exudative lesions on the scrotum and bilateral inguinal areas. The patient was diagnosed with gastric cancer 4 months after starting the corticosteroid treatment and died of multiple organ failure 4 months after operation. Our case and that of Koga et al seem to suggest that gastric cancer was related to pemphigus vegetans.

Systemic corticosteroid is the mainstay of therapy. The addition of immunosuppressive agents (azathioprine, cyclosporine, cyclophosphamide, mycophenolate mofetil, and methotrexate) is often considered whereas systemic corticosteroid alone does not induce disease remission. Immunosuppressive agents may improve remission rates and allow the steroid-sparing effect to occur.

In conclusion, this is a rare case of pemphigus vegetans occurring at an unusual site and the patient developed gastric cancer later. Patients with pemphigus vegetans are warranted to work up for possible associated malignancy.

Chi-Shou Lo
Department of Dermatology, Chang Gung Memorial Hospital, Keelung, Taiwan
Chang Gung University, College of Medicine, Taoyuan, Taiwan

Tseng-tong Kuo
Department of Pathology, Chang Gung Memorial Hospital, Taipei, Taiwan
Chang Gung University, College of Medicine, Taoyuan, Taiwan

Jheng-Wei Lin
Department of Dermatology, Chang Gung Memorial Hospital, Keelung, Taiwan
Chang Gung University, College of Medicine, Taoyuan, Taiwan

Corresponding author. Department of Dermatology, Chang Gung Memorial Hospital, No. 199, Dunita Rd., Songshan Dist., Taipei City 105, Taiwan. Tel.: +886 27135211x3397; fax: +886 27191623. E-mail address: s81095@gmail.com

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