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LETTER TO THE EDITOR

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A case of pemphigus herpetiformis associated with a progressive gastric cancer and negative envoplakin and periplakin autoantibodies

Dear Editor,

Pemphigus herpetiformis (PH) is a rare variant of pemphigus that shows similar clinical manifestations with dermatitis herpetiformis and immunological features of pemphigus.[1] We report here a patient with PH who presented a progressive gastric cancer.

A 74-year-old man presented a 3-week history of itchy annular erythema on his trunk. The lesions had an indurated border, extended centrifugally with central clearing (Figure 1A). His past medical history was significant for an early stage of esophagus cancer, thyroid cancer, and upper pharyngeal cancer. He had no signs of tumor recurrence for more than 5 years.

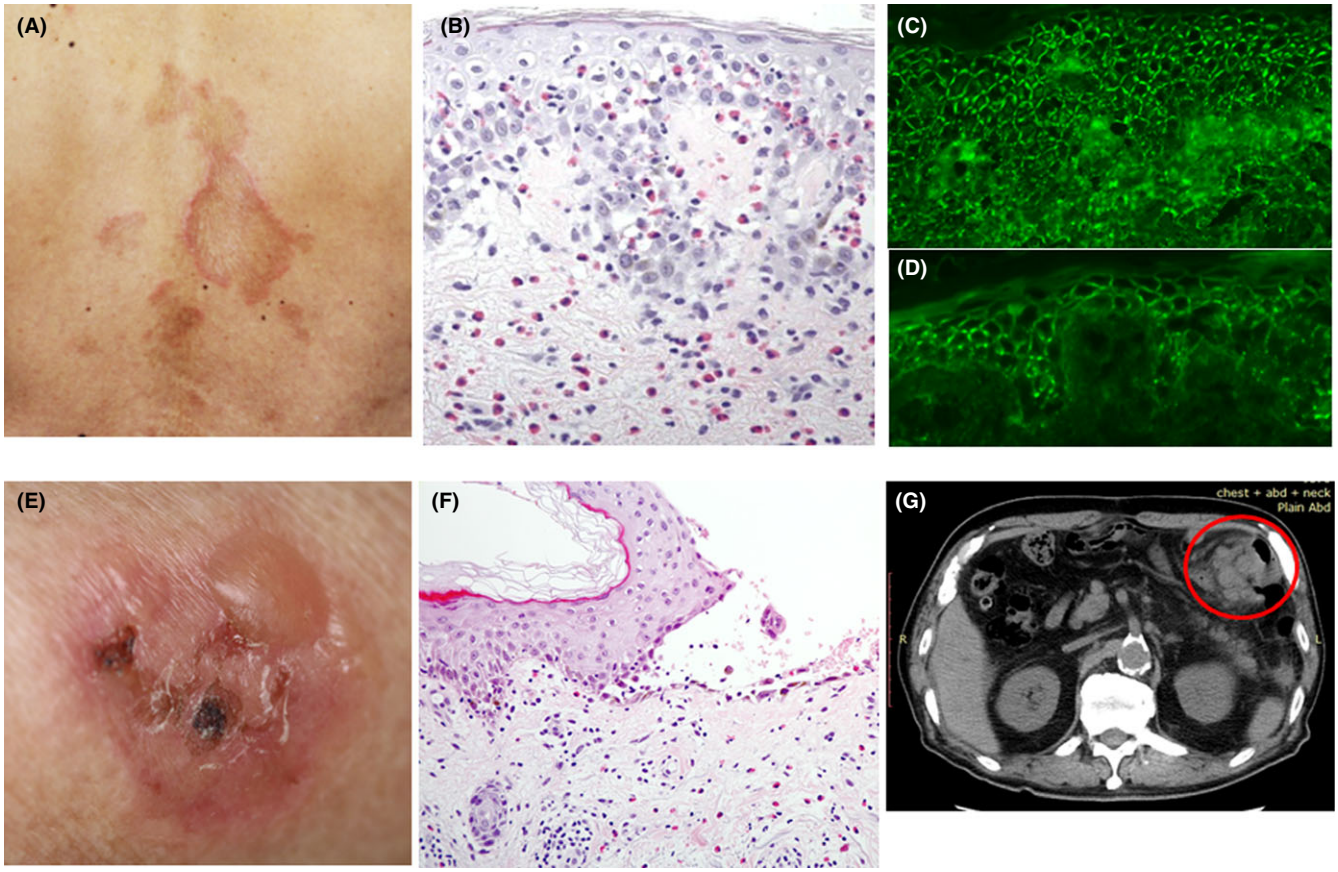

FIGURE 1 A, Annular erythema with indurated border on his trunk; B, Hematoxylin-eosin (HE) staining of the annular erythema on the trunk showing eosinophilic spongiosis (original magnification × 200); C-D, Intercellular deposition of IgG on (C) direct immunofluorescence and (D) indirect immunofluorescence using normal human skin substrate; E, Blisters on the border of annular erythema on his right forearm; F, HE staining of (E) showing suprabasal clefts with acantholytic cells (original magnification × 100); G, A computed tomography (CT) scan of the abdomen revealed gastric cancer (red circle)

Histological examination of a skin biopsy from the erythematous border revealed eosinophilic spongiosis (Figure 1B). Serum analysis was positive for antidesmoglein 3 (Dsg3) antibodies (782.0 U/mL, normal level < 20 U/ml) and negative for anti-Dsg1 and anti-BP180 antibodies by ELISA testing. Direct immunofluorescence (IF) showed the deposition of both IgG and C3, but not IgA in the intercellular spaces of the epidermis (Figure 1C). Indirect IF using normal human skin substrate was also positive (Figure 1D).

Because he has suffered from multiple cancers, extensive investigation was performed. Examinations with computed tomography (CT) scans of the chest and abdomen, gastroscopy, and colonoscopy were normal. To exclude paraneoplastic pemphigus (PNP), autoantibodies against envoplakin and periplakin were examined by Western blotting, but the results were negative. Based on the clinical, immunological, and histological features, a diagnosis of PH was made. Although a few new skin lesions continued to arise, treatment with topical steroid was effective for skin lesions.

Three months later, he developed deep vein thrombosis (DVT) in his left leg. There was annular erythema on his forearms and trunk and blisters were found on the border of the lesions (Figure 1E). No mucosal involvement of oral cavity was present. A histological examination from his forearm revealed eosinophilic spongiosis and suprabasal clefts with acantholytic cells (Figure 1F), which was consistent with PH. An abdomen CT scan showed gastric cancer and peritoneal seeding (Figure 1G). He died 3 months later.

Pemphigus herpetiformis is diagnosed by pruritic annular or gyrate erythematous skin lesions with or without vesicles and immunological findings with deposits of IgG and C3 in the epidermis.[1,2] Autoantibodies against Dsg1 or less frequently, against Dsg3 or desmocollins, are detected in PH.[3,4] PH tends to be less life-threatening than other types of pemphigus and generally to have a good prognosis.[1] In our case, the skin lesions were well tolerated by only topical steroid treatment.

Our patient presented DVT and blisters on the border of annular erythema 3 months after the first admission and an advanced gastric cancer was found. Recently, there were several reports of PH associated with internal malignancies, such as lung cancer,[5] esophageal carcinoma,[6] prostate cancer,[7] and angiosarcoma.[8] To our knowledge, this is the first report of PH associated with gastric cancer, which progressed rapidly. Although the relationship between PH and internal malignancies has not been known yet, we consider that screening for the internal malignancies is important in PH especially when clinical presentation has changed.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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