

Pemphigus vegetans in a patient with colonic cancer

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ABSTRACT

Pemphigus vegetans is a rare variant of pemphigus vulgaris characterized by vegetating plaques in the flexural regions. The coexistence of pemphigus vegetans and internal neoplasm is rare, being described only in four cases in the literature. We report the case of a patient with a typical skin eruption of pemphigus vegetans, who was detected with colonic cancer.

Key words: Colonic cancer, Malignancy, Pemphigus vegetans

INTRODUCTION

Pemphigus vegetans is a rare variant of pemphigus vulgaris and is characterized by vegetating plaques in the flexural regions.^[1] The coexistence of this immunobullous dermatosis and internal malignancy has been rarely reported in the literature, being described only in four cases.^[2–5]

CASE REPORT

An 83-year-old woman presented to our Dermatology Department with a one week history of vegetating lesions in the right temporal area, both groin region and perineal region. There was no history of other skin or systemic diseases; however, a recent history of hematochezia was referred. On physical examination, there were numerous ulcerations and exudative, verrucous, vegetating plaques on the right temporal area [Figure 1], inguinal and perineal area [Figure 2]. Histological findings included acanthosis, papillomatosis, suprabasal acantholysis, intraepidermal microabscesses filled with eosinophils, and an inflammatory infiltrate composed of lymphocytes and numerous eosinophils in the upper dermis [Figure 3]. Direct immunofluorescence of perilesional skin showed intercellular deposits of IgG and C3 [Figure 4].

Antibodies to desmoglein 1 and 3 were analyzed using specific enzyme-linked immunosorbent assay (ELISA). Index values to desmoglein 3 were 231 (normal < 20), whereas those to desmoglein 1 were negative. Indirect immunofluorescence of patient serum on murine bladder epithelium was negative. The total blood cell count was normal except for a hypochromic microcytic anemia and the results of routine serum chemistry were within the normal ranges. Since the patient had anemia and a recent history of hematochezia, a colonoscopy was performed showing a colonic tumor. The patient was diagnosed as having pemphigus vegetans, Hallopeau type, and prednisolone 25 mg daily was started; surgical resection and complete staging was performed, revealing a localized colonic adenocarcinoma with no evidence of metastatic disease (T1N0M0). Within a few weeks the vegetating lesions began to regress, leaving pigmented maculae. Prednisolone was tapered during the first month and a decrease of the desmoglein 3 titer was observed. During the 10 months follow-up, without steroids therapy, no relapse of the skin disease or colonic cancer has been noted and the desmoglein 3 titer continued to be negative.

DISCUSSION

Pemphigus vegetans is a rare clinical variant of

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Figure 1: Vegetating plaque on the right temporal area



Figure 2: Verrucous, vegetating and exudative plaques on inguinal and perineal area

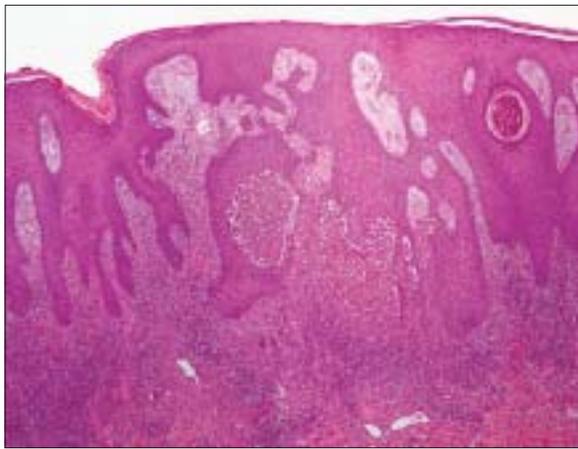


Figure 3: Histopathological examination showing acanthosis, papillomatosis, suprabasal acantholysis, intraepidermal microabscesses filled with eosinophils, and an inflammatory infiltrate composed of lymphocytes and numerous eosinophils in the upper dermis (H and E, x10).

pemphigus vulgaris, which has two clinical subtypes: the Neumann type and the Hallopeau type.^[1,6] Although both types are characterized by vegetating plaques in the skin folds, the primary lesion, response to therapy and prognosis differ. The Neumann type usually begins with bullae, has a worst response to therapy and a poor prognosis, similar to pemphigus vulgaris; the Hallopeau type usually begins with grouped pustules and has an excellent response to therapy with long remission being observed.^[6] A statistically increased incidence of malignancy is observed in patients with pemphigus, mainly pemphigus vulgaris and pemphigus erythematosus.^[7,8] However, the coexistence of pemphigus vegetans and internal neoplasm has been rarely reported in the literature, being described in four cases, two cases of coexistence with lung cancer,^[2,3] one with a lymphosarcoma,^[4]

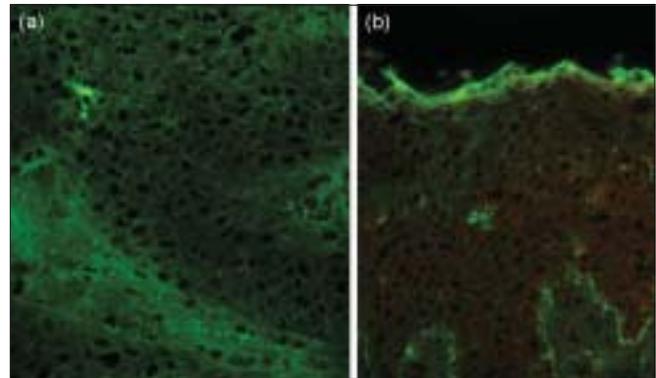


Figure 4: Intercellular deposits of IgG (a) and C3 (b) on direct immunofluorescence

and another associated with a gastric cancer.^[5] In the case described by Bastiaens *et al.*,^[3] the removal of the tumor, a lung cancer, caused the disappearance of the lesions and circulating antibodies of pemphigus, proving the inducing role of the tumor. However, follow-up was not possible because the patient died suddenly after pneumonectomy. The simultaneous occurrence of pemphigus vegetans and gastric cancer, reported by Koga *et al.*^[5] and the recurrence of the skin eruption after surgical removal, when metastatic lesions were found in the liver, also suggests that they were closely associated. In our case, both diseases appeared to occur simultaneously and, since it was a localized neoplasm, the surgical treatment was probably curative, hence the remission of the skin condition and the normalization of the desmoglein 3 titer. For this reason it is considered that pemphigus vegetans and colonic cancer were causally related and that internal malignancy should be considered in patients with this immunobullous disease.

REFERENCES

1. Rosenberg FR, Sanders S, Nelson CT. Pemphigus: a 20-year review of 107 patients treated with corticosteroids. *Arch Dermatol* 1976;112:962-70.
2. Serwin AB, Bokinić E, Chodynicka B. Pemphigus vegetans in a patient with lung cancer. *Dermatology Online J*. Available at: <http://dermatology.cdlib.org>. Accessed March 1, 2005.
3. Bastiaens MT, Zwan NV, Verschueren GL, Stoof TJ, Nieboer C. Three cases of pemphigus vegetans: induction by enalapril-association with internal malignancy. *Int J Dermatol* 1994;33:168-71.
4. Krain LS, Bierman SM. Pemphigus vulgaris and internal malignancy. *Cancer* 1974;33:1091-9.
5. Koga C, Izu K, Kabashima K, Tokura Y. Pemphigus vegetans associated with gastric cancer. *J Eur Acad Dermatol Venereol* 2007;21:1288-9.
6. Ahmed AR, Blose DA. Pemphigus vegetans. Neumann type and Hallopeau type. *Int J Dermatol* 1984;23:135-41.
7. Younus J, Ahmed AR. The relationship of pemphigus to neoplasias. *J Am Acad Dermatol* 1990;23:498-502.
8. Ogawa H, Sakuma M, Morioka S, Kitamura K, Sasai Y, Imamura S, *et al.* The incidence of internal malignancies in pemphigus and bullous pemphigoid in Japan. *J Dermatol Sci* 1995;9:136-41.

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