



CASE REPORT

Paraneoplastic Pyoderma Gangrenosum Associated with Rectal Adenocarcinoma

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Pyoderma gangrenosum (PG) is a rare chronic neutrophilic dermatosis characterized by painful necrotic ulceration. The most common diseases associated with PG are inflammatory bowel disease, certain rheumatologic and hematologic diseases, and malignancy. Here, we describe the case of a 60-year-old man who presented with pruritic and painful erythematous ulcerative macules and patches on both lower extremities, and was diagnosed with PG based on his clinical and histologic features. His PG became exacerbated despite standard therapy with a high-dose systemic steroid in combination with dapsons and cyclosporine. Systemic evaluation of underlying conditions revealed rectal adenocarcinoma at the rectosigmoid junction (T3N0M0), which was completely removed via Hartmann's procedure followed by adjuvant chemotherapy. Two months after anticancer therapy, his PG was completely healed with hypertrophic scarring. Herein, we present the first case of paraneoplastic PG caused by rectal adenocarcinoma in Korea. (*Ann Dermatol* 30(1) 79~82, 2018)

-Keywords-

Colonic neoplasms, Paraneoplastic, Pyoderma gangrenosum

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INTRODUCTION

Pyoderma gangrenosum (PG) is a rare chronic neutrophilic dermatosis characterized by painful necrotic ulceration. It has been reported to be associated with various disorders¹⁻³, including inflammatory bowel disease, certain rheumatologic and hematologic diseases, and malignancy. Paraneoplastic PG was first described in 1993 by Duguid et al.⁴ in 4 patients in association with myeloproliferative malignancy. However, there have been few reports of paraneoplastic PG caused by solid malignant tumors. Here, we present the first Korean case of paraneoplastic PG caused by rectal adenocarcinoma.



Fig. 1. Clinical photograph, showing ulcerative patches on bilateral lower extremities with edema.

CASE REPORT

A 60-year-old man visited our dermatologic clinic with a 5-month history of skin eruptions. The skin lesions were localized on the lower extremities as pruritic and painful ulcers accompanied by leg edema (Fig. 1). He had a 3-year history of hypertension. Laboratory examinations revealed negative results for antinuclear antibody and anti-neutrophil cytoplasmic antibody. To exclude vascular disease, Doppler ultrasonography was performed, which showed no specific findings. Skin biopsy of the right lower leg showed central necrotizing suppurative inflammation with ulceration, demonstrating neutrophilic infiltration with leukocytoclasia and dermolysis in the central part of the lesions without vasculitis. Multiple infections, including mycobacteria, deep fungi, or other bacterial diseases, were excluded based on negative tissue culture results. After ruling out other tentative diagnoses, PG was diagnosed based on his clinical and histopathologic features. The patient received intensive treatment with a high-dose systemic steroid (0.5~1 mg/kg/d) in combination with dapsons and cyclosporine; however, the ulcerative lesions became aggravated. Systemic evaluation of underlying diseases, including routine laboratory tests, peripheral blood smear, abdominal computed tomography (CT), chest CT, and colonoscopy, was performed. On colonoscopy, a huge ulcerofungating mass at the rectosigmoid junction was detected, which was confirmed as rectal adenocarcinoma by histopathologic examination (Fig. 2A).

Two months after complete eradication of the tumor by Hartmann's procedure (Fig. 2B) and adjuvant chemotherapy, the patient's lesions were completely healed with hypertrophic scarring (Fig. 2C). There was no recurrence

of PG after complete resection of adenocarcinoma during our 3-year follow-up.

DISCUSSION

PG is characterized by rapidly progressive ulcers with irregular and undermined borders. PG is a rare noninfectious dermal neutrophilia on the spectrum of neutrophilic dermatoses⁵, which share similar clinical features, such as underlying disorders, a tendency for pathergy, and therapeutic response. Lesions can be classified morphologically as ulcerative, bullous, pustular, or vegetative⁶. To establish diagnosis of PG, clinicians should exclude all other possible associated diseases⁶. Approximately half of PG cases are known to be associated with underlying systemic diseases, such as rheumatoid arthritis, inflammatory bowel disease, or myeloproliferative disorders⁷. However, there have been several reports of PG associated with solid malignancies⁸⁻¹⁵. Although the exact mechanisms by which solid tumors induce PG are unknown, abnormal immune surveillance, such as neutrophilic dysfunction, defects in chemotaxis, or hyperreactivity, may play a role as in PG associated with other conditions¹⁶. Several cases of PG in relation to colorectal carcinoma have been reported (Table 1)^{11,13-15}. In most of these reports, patients presented with a solitary ulcer in various locations^{11,13,14}. On the other hand, Shahi and Wetter¹⁵ reported PG associated with solid organ malignancies, including a case of recurrent PG presenting with numerous ulcers on the legs. Owing to the patient's history of ulcerative colitis, it is uncertain whether the lesions were related to ulcerative colitis or rectal cancer.

Our patient presented with multiple ulcers on the lower

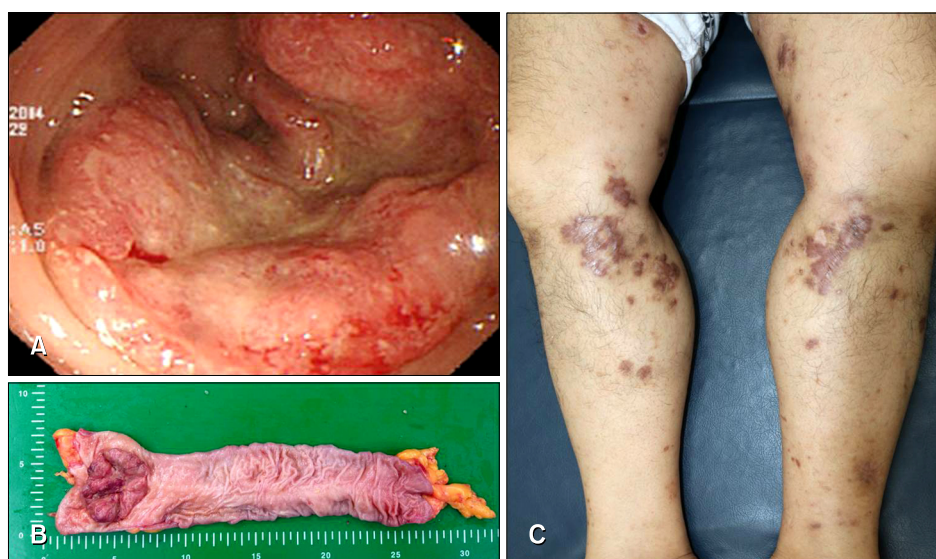


Fig. 2. (A) Colonoscopic image, showing an ulcerofungating mass at the rectosigmoid junction. (B) Hartmann's procedure was performed for resection of colon cancer. A 5.5×4-cm rectal adenocarcinoma (T3N0M0) was found. (C) Clinical photograph, showing completely healed pyoderma gangrenosum lesions with hypertrophic scarring after the fourth rounds of adjuvant chemotherapy.

Table 1. Previously reported cases of PG associated with colorectal malignancies

Study	Sex/age (yr)	PG subtype	PG location	The number of lesions	Solid organ cancer	Timing of PG	Other associated systemic disease
Bunte et al. ¹¹ (2008)	M/53	Ulcerative (classic)	Right scapula (injury while gardening)	Solitary	Adenocarcinoma of the sigmoid colon	4 months before diagnosis of malignancy	None
Foley and Laing ¹³ (2015)	F/70	Ulcerative (classic)	Upper back	Solitary	Colon cancer	NA	None
Sakai et al. ¹⁴ (2006)	F/70	Peristomal	Peristoma	Solitary	Rectal cancer	40 days after ileostomy for the rectal cancer	None
Shahi and Wetter ¹⁵ (2015)	M/36	Ulcerative (classic)	Lower extremities	Multiple	Metastatic rectosigmoid carcinoma to lungs	Synchronous	Ulcerative colitis
Present study	M/60	Ulcerative (classic)	Lower extremities	Multiple	Rectal adenocarcinoma	6 months before diagnosis of malignancy	None

M: male, F: female, PG: pyoderma gangrenosum.

extremities, and systemic evaluation of underlying diseases revealed rectal adenocarcinoma. To our knowledge, there has been no report of PG presenting as multiple ulcers in relation to primary colorectal carcinoma in a Korean patient. This case was confirmed as paraneoplastic PG caused by progressive colorectal cancer based on the following findings: (1) the lesions did not respond to standard immunosuppressive agents used to treat PG; (2) the refractory lesions healed dramatically after complete removal of the tumor; and (3) there was no recurrence of PG after the solid malignancy was removed during our 3-year follow-up.

In conclusion, we reported the first Korean patient with paraneoplastic PG manifesting as multiple leg ulcers caused by rectal adenocarcinoma. From our experience, and because colorectal cancer has become a common malignancy worldwide, intensive evaluation to detect underlying solid malignancies is required, especially in cases of refractory PG that do not respond to conventional therapies.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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