

# A case of generalized bullous pyoderma gangrenosum with myelodysplastic syndrome

## Abstract

Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis which may be associated with systemic disorders. Approximately 50% of patients with PG have an associated systemic disease. It's most associated with inflammatory bowel disease, inflammatory arthritis, and hematological disorders. Bullous PG is rare type of PG and is mostly reported in patients with comorbid myeloproliferative diseases. We present a case of bullous PG who was diagnosed with myelodysplastic syndrome.

**Keywords:** bullous pyoderma gangrenosum, myelodysplastic syndrome, neutrophilic dermatosis, peristomal types, sweet syndrome

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## Introduction

Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis that is usually idiopathic, although it may be associated with various systemic disorders. Clinically, it is classified into ulcerative, pustular, bullous, vegetative, and peristomal types. A few atypical and rare variants have also been described.<sup>1,2</sup>

## Case report

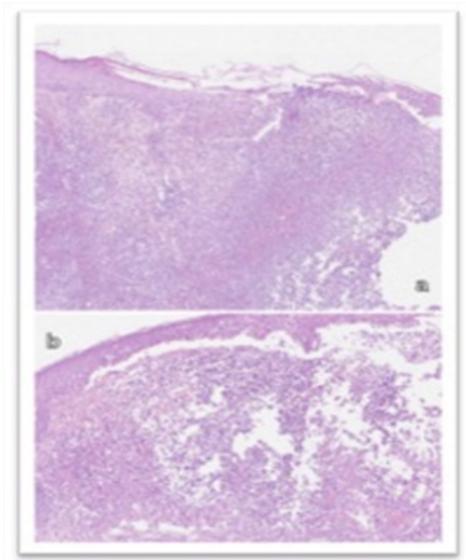
We describe a case of a 70-year-old white female who had been diagnosed with myelodysplastic syndrome 4 months earlier and who was admitted with an acute, painful vesiculobullous eruption that started on the face and neck. The lesions had spread to the patient's trunk, arms, legs, genitals, and oral mucosa, and the size of the ulcers had nearly doubled a few days earlier.

A dermatological examination revealed multiple, variable-sized vegetative, hemorrhagic bullae with central necrosis and erosion on the scalp, face, neck, trunk, and extremities (Figure 1). There were also ulcerations on the patient's oral mucosa. Her laboratory values showed anemia, thrombocytopenia, and leukocytosis due to myelodysplastic syndrome. Wound cultures (including anaerobes and fungi) were negative. The patient underwent a series of serologic tests (including tests for anti-neutrophil cytoplasmic antibodies [ANCA], perinuclear ANCA, rheumatoid factor, cryoglobulins, a hepatitis panel, and anti-nuclear antibodies), which yielded negative results. A skin biopsy showed epidermal necrosis, diffuse dermal neutrophilic infiltration, and vascular stasis (Figure 2). The pathology confirmed the diagnosis of PG. Prednisone (1mg/kg/day) and cyclosporine (5 mg/kg/day) were started. The ulcers diminished in size significantly following the first week of treatment (Figure 3). After 3 weeks of treatment, the prednisone was tapered. The lesions recurred when the prednisone

dose decreased to 0.5mg/kg/day; thus, the dose of prednisone was increased to 1mg/kg/day. Meanwhile, other immunosuppressive or TNF-blocker treatments were discussed. The patient was noted to be febrile on follow-up. Her C-reactive protein and sedimentation rate levels also increased while all cultures were negative. Broad-spectrum antibiotic treatment was started while the prednisone taper was restarted. The patient was brought to a follow-up exam with a loss of consciousness and lesion recurrence. Her hematologic and systemic conditions progressed, and the patient was lost after 1 week.



**Figure 1** Multiple vegetative and hemorrhagic bullae of various sizes with central necrosis.



**Figure 2** (A) Ulceration, a subepidermal blister, and dermal neutrophilic infiltration with abscess formation (hematoxylin and eosin [H&E], X40). (B) Edema, endothelial swelling, erythrocyte extravasation, and vasculitis (H&E, X40).



**Figure 3** Prominent regression of the lesions.

## Discussion

PG is a rare skin disorder with several clinical variants. It affects individuals of all ages, with a peak incidence between 20 and 50 years of age. It affects men and women almost equally.<sup>1</sup> Its pathophysiology is complex and still not fully understood.<sup>3</sup> PG is

often associated with inflammatory or neoplastic disease. It is most associated with inflammatory bowel disease, inflammatory arthritis, and hematological disorders.<sup>4</sup> PG may precede, follow, or occur simultaneously. Clinically, PG presents as several clinical variants; thus, diagnosing the condition can be challenging.<sup>1</sup> Clinically, it is classified into ulcerative, pustular, bullous, vegetative, and peristomal types. Bullous PG often begins at atypical sites such as the face and dorsum of the hands, as seen in our patient. Bullous PG is most associated with myeloproliferative diseases (mainly acute myelogenous leukemia).<sup>1-3</sup> Unfortunately, there are no specific serologic markers for PG, and the histopathology is non-specific. The histopathologic findings depend on the PG subtype and stage of the disease.<sup>5</sup> Therefore, there are many diseases to be considered in the differential diagnosis of PG such as infections, vasculitis and autoimmune diseases, Sweet syndrome, vascular diseases, and insect bites.<sup>1</sup> The treatment depends on the location, number, and size of the lesions, extracutaneous involvement, the presence of associated diseases, comorbidities, and patient preference.<sup>6</sup>

## Conclusion

PG is a complex disease that presents a challenge from both diagnostic and therapeutic points of view. Approximately 50% of patients with PG have an associated systemic disease. Bullous PG is a rare type of PG and is mostly reported in patients with comorbid myeloproliferative diseases. The prognosis of PG is usually variable while bullous PG usually has a poor prognosis, as in our patient.

## Acknowledgements

None.

## Conflicting interests

The author declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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