

## CASE IMAGE

# Classic pyoderma gangrenosum

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**Key Clinical Message**

Pyoderma gangrenosum is a rare inflammatory ulcerative skin disease of unknown etiology. We report an image of a patient with pyoderma gangrenosum who presented right leg ulcers with violaceous margins, histologically characterized by mono- and polynuclear cell infiltrates. The patient was successfully treated with cyclosporin A.

**KEYWORDS**

chronic diseases, dermatology, immunology, rheumatology, surgery

## 1 | CASE PRESENTATION

A 51-year-old woman complained of multiple right leg lesions, apparently due to trauma to the knee as a single lesion that progressively worsened as similar lesions appeared all over the leg in the course of about 1 year. She referred having taken prednisone 1 month earlier (prednisone 0.5 mg/kg/day, then tapering) without benefit. The patient's history revealed no signs or symptoms suggestive of inflammatory bowel disease, arthritis, hematologic or myeloproliferative disorders. On physical examination, her right leg was covered with violaceous lesions including two 8-cm purulent ulcers in the popliteal and lateral malleolar regions (Figure 1A,B). The violaceous color of the ulcer margins suggested pyoderma gangrenosum (PG). Laboratory tests were made to exclude any possible associated systemic diseases, and revealed a modest increase of inflammatory parameters. Serological testing

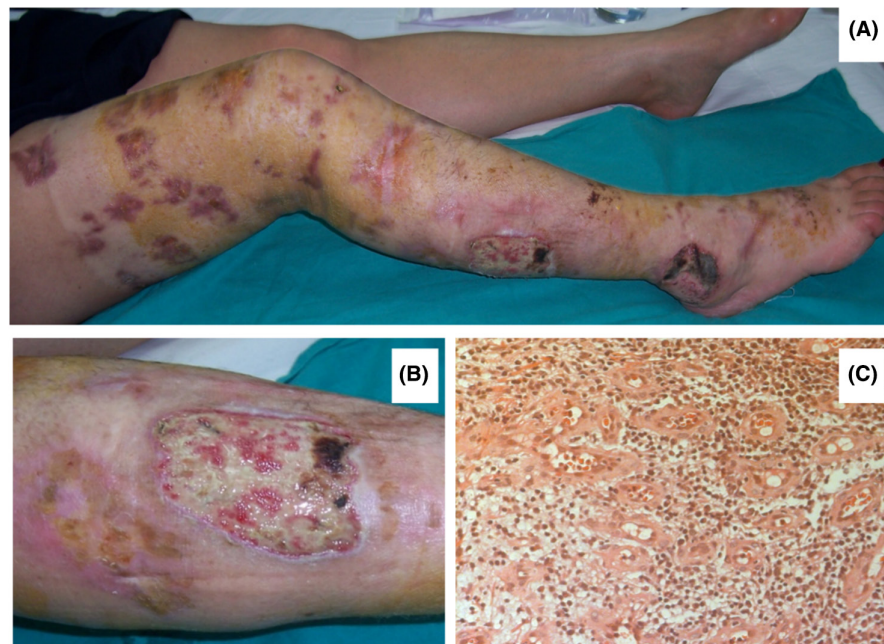
for hepatitis B and C, HIV, tuberculosis, and syphilis were negative, as also autoimmunity screening. Multiple ulcer cultures for pathogens remained negative. A skin biopsy at the margins of the ulcer revealed a diffuse infiltrate of inflammatory perivascular mono- and polynuclear cells in the absence of any infection suggestive of PG<sup>1</sup> (Figure 1C). Cyclosporin A (2.5 mg/kg/day) and prednisone (50 mg/day, then gradually tapered) were started, which yielded a marked improvement in the ulcer healing without any significant side effects. Over a 2-year follow-up period, no evidence of recurrence was observed.

## 2 | DISCUSSION

PG is a chronic neutrophilic skin disease characterized by skin papules or pustules which rapidly progress to painful ulcers.<sup>2</sup> To date, no criteria for PG diagnosis have been

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**FIGURE 1** Macroscopic and histological lesions in pyoderma gangrenosum. (A) Asymmetric distribution of multiple right leg ulcers; (B) deep ulceration with rapidly evolving purulent wound, showing undermined violaceous margins in the popliteal and lateral malleolar regions; (C) Diffuse infiltrate of perivascular mono- and polynuclear cells within the dermis, H&E staining, 20 $\times$ .

validated in the clinical setting, and misdiagnosis often occurs.<sup>1</sup> The differential diagnosis of PG must include infection, malignancy and vasculitis.<sup>3</sup> Recently, *Maverakis et al.*<sup>1</sup> defined a set of nine criteria for PG diagnosis, among which biopsy of ulcer edges demonstrating a neutrophilic infiltrate was the only major criterion. Minor criteria included: (a) exclusion of infection; (b) pathergy; (c) a history of inflammatory bowel disease or inflammatory arthritis; (d) papules, pustules, or vesicles that rapidly ulcerate; (e) peripheral erythema, undermining the border, and tenderness at the ulceration site; (f) multiple ulcerations, of at least one leg on the anterior lower surface; (g) cribriform or “wrinkled paper” scars at healed ulcer sites; and (h) a decreased ulcer size after immunosuppressive treatment.<sup>1</sup> This image report suggests that clinicians should keep this condition in mind and inspect for PG when patients present ulcers with violaceous margins, to facilitate the choice of the right treatment and avoid complications.

#### AUTHOR CONTRIBUTIONS

**Marcella Prete:** Conceptualization; data curation; investigation; writing – original draft. **Vito Racanelli:** Investigation; supervision; writing – review and editing. **Patrizia Leone:** Investigation; visualization; writing – review and editing. **Elvira Favoino:** Data curation; supervision; visualization; writing – original draft. **Federico Perosa:** Conceptualization; project administration; resources; writing – review and editing.

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None.

#### CONFLICT OF INTEREST STATEMENT

The authors declare that they have no conflict of interest.

#### DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

#### CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

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