

## Lesions of Pyoderma Gangrenosum Mimicking Sweet's Syndrome; A Diagnostic Dilemma

Pyoderma gangrenosum (PG), an uncommon, distinctive cutaneous ulcerative condition manifests with a polymorphic clinical picture ranging from a pustule to large rapidly progressive ulcers.<sup>[1]</sup> It may be associated with various systemic disorders of diverse etiology.<sup>[1,2]</sup> We report a case of a middle-aged woman with rheumatoid arthritis with an atypical plaque type of PG mimicking lesions of Sweet's syndrome with coexistent ulcerative PG.

A 44-year-old woman presented with fever, painful lesions, and large ulcers over upper and lower extremities for 4 months. The lesions over lower limbs were initially nodular that progressed to ulcers with foul-smelling discharge. She had similar lesions over face and legs 18 months ago which healed with scar formation. She was diagnosed to have rheumatoid arthritis 8 years ago for which she was on treatment with methotrexate; however, she was irregular.

On examination, there was an annular erythematous indurated tender plaque with central clear zone studded with pustules at the periphery measuring about 10 cm × 12 cm over the extensor aspect of the right forearm [Figure 1]. There were two tender ulcers with violaceous borders, interspersed with pustules, unhealthy granulation tissue and slough, purulent exudate, and hemorrhagic crusts over left thigh and lower 1/3<sup>rd</sup> of left leg [Figure 2a and b]. Multiple erythematous indurated tender plaques were distributed over bilateral forearms and legs. There were cribriform scars over the posterior aspect of right leg suggestive of previously healed lesions [Figure 2c]. Nails, hair, mucosal and systemic examination were unremarkable. We considered a differential diagnosis of PG and Sweet's syndrome.

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Her hemoglobin was 10.5 gm/dL and total leukocyte count was 16,300/μL. She had an elevated erythrocyte sedimentation rate (84 mm/h) and C-reactive protein (45 mg/L). Peripheral smear showed normocytic normochromic anemia with neutrophilic leukocytosis. Pathergy test showed positivity after 48 hours with the appearance of a pustular lesion. Histopathological examination of the skin biopsy specimen from the plaque showed acanthotic epidermis and dense dermal neutrophilic infiltrate along with lymphocytes and eosinophils [Figure 3a and b]. Ulcerative lesion showed a similar feature with karyorrhectic debris [Figure 4a and b]. On the basis of the clinico-histopathological features, we arrived at a diagnosis of PG associated with rheumatoid arthritis. She had ulcerative type of PG in the lower limb and a unique plaque studded with pustules on the right forearm. She was treated with systemic corticosteroids (prednisolone 60 mg gradually tapered to 20 mg over a span of 2 months) along with anti-rheumatoid medications (Methotrexate 15 mg/week). A follow-up visit at 1 and 2 months showed complete resolution of all the lesions with milia formation, scarring, and pigmentary changes [Figure 5a and b].

PG is clinically classified into ulcerative, pustular, bullous, and vegetative types.<sup>[2]</sup> Ulcerative (classic form) PG is the most frequently observed type of PG. It is usually associated with inflammatory bowel disease and arthritis.<sup>[1,2]</sup> Our patient, a known case of rheumatoid arthritis, had ulcerative type of PG over the left thigh and extremities with typical morphology.

Some rare variants of PG have been described including peristomal PG, genital PG, PG in infants and young

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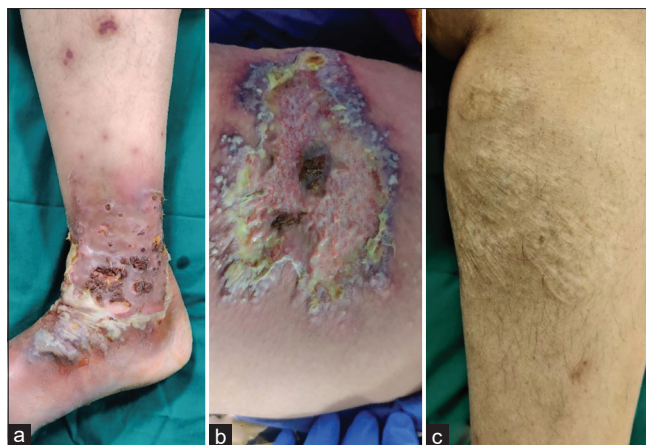
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**Figure 1:** Erythematous and edematous plaque with central depression studded with pustules over the extensor aspect of the right forearm mimicking Sweet's syndrome

children, malignant pyoderma, pyostomatitis vegetans, and extracutaneous neutrophilic disease.<sup>[1,3-6]</sup> Our patient had an unusual presentation of PG in the form of edematous plaques over the upper limbs mimicking Sweet's syndrome in addition to classic lesions. Some authors believe that PG and Sweet's syndrome represent a continuum of spectrum of disease.<sup>[7,8]</sup> Our finding of Sweet's-like lesion also supports this hypothesis. PG and Sweet's syndrome share many features including response to immunosuppressive therapy. The typical forms of the two conditions are clinically fairly versatile: PG showing cutaneous ulceration with a purple undermined border and heal with cribriform scars, and Sweet's syndrome



**Figure 2:** Tender ulcers with violaceous borders, interspersed by pustules, unhealthy granulation tissue and slough over the (a) left thigh; (b) Lower 1/3<sup>rd</sup> of left leg; (c) Cribriform scars over the posterior aspect of the right leg suggestive of previous healed lesions at presentation

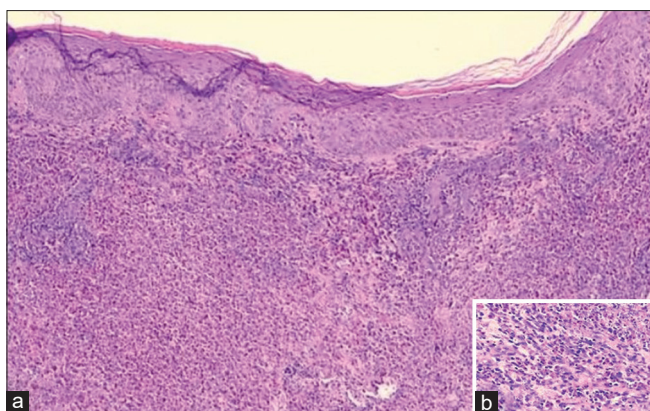
having tender, erythematous, non-ulcerating plaques and nodules which heal without scar formation.<sup>[7]</sup> Very few cases presenting with more than one morphological type of PG have been reported. Patients with typical PG on the legs and atypical PG on the arms/hands have been described.<sup>[2,7]</sup>

Our diagnosis of PG was based on the following evidences:

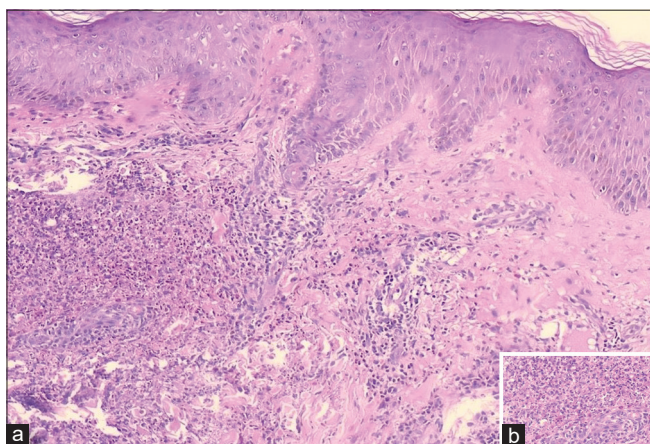
1. Co-existence of classic ulcerative lesions over the lower extremities
2. Histopathology showing massive neutrophil-dominant dermal infiltration
3. Healing of lesions with scarring and in most occasions with classically described-cribriform scarring
4. Association with rheumatoid arthritis.

The histopathological features of PG vary depending on the type, the stage of evolution of the lesion, and the site from which lesion has been biopsied. Enormous infiltrates of neutrophils in the absence of granuloma formation, is characteristic of PG.<sup>[1]</sup> The infiltrate in PG is predominantly peri-vascular with endothelial swelling in some vessels with the presence of a varying amount of fragmentation.<sup>[7]</sup> PG is folliculocentric and histology shows sterile follicular and perifollicular inflammation.<sup>[9]</sup> Vasculitis may be seen in about 40% of cases of PG whereas it is not usually a prominent feature of Sweet's syndrome.<sup>[10]</sup>

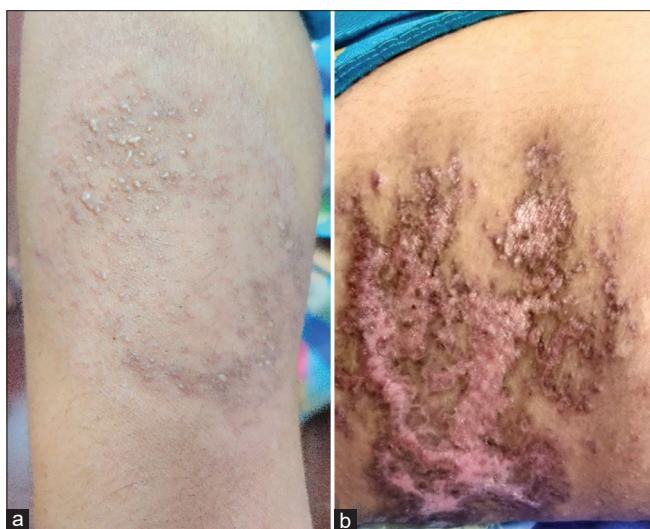
Oral corticosteroids (0.5–1 mg/kg/day) are considered as the cornerstone of treatment of severe PG as well as Sweet's syndrome. Other systemic agents employed for PG with favorable outcomes include cyclosporine, colchicine, sulphasalazine, dapsone, minocycline, apremilast, thalidomide, pulsed intravenous methylprednisolone, methotrexate, mycophenolate mofetil, cyclophosphamide, azathioprine, and high-dose intravenous immunoglobulin. Various biologics like infliximab, adalimumab, etanercept, ustekinumab, anakinra, and canakinumab have shown encouraging results in the treatment of PG.<sup>[3]</sup> Our patient



**Figure 3:** (a) Biopsy from forearm showing acanthotic epidermis and dermis showing a dense inflammatory cell infiltrate composed of neutrophils, lymphocytes and many eosinophils (H and E, 4X). (b) Higher magnification of the same showing inflammatory cell infiltrate (H and E, 40X)



**Figure 4:** (a) Biopsy from the leg showing dense inflammatory cell infiltrate of numerous neutrophils admixed with lymphocytes and eosinophils and karyorrhectic debris (H and E, 4X). (b) Higher magnification of the same showing the inflammatory cell infiltrate (H and E, 40X)



**Figure 5:** (a) Healing of the right forearm plaque with milia formation and pigmentary changes after 2 months. (b) Resolution of lesion over the left thigh with scarring and pigmentary changes after 2 months

was treated with systemic corticosteroids, methotrexate and appropriate wound care, following which there was resolution of the lesions within 2 months.

PG may have Sweet's syndrome-like presentation in the form of erythematous edematous plaques. Histopathological differentiation of PG from Sweet's syndrome is an arduous task. Astute clinical diagnosis supported by the histopathology is the yardstick in the diagnosis of majority of neutrophilic dermatoses including PG. A mere presence of atypical lesion should not deviate from the primary diagnosis.

### *Declaration of patient consent*

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### *Conflicts of interest*

There are no conflicts of interest.

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