Multiple Painful Ulcers of a 42-year-old Man

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CASE REPORT

A 42-year-old male suffered from multiple painful ulcers surrounding by raised, undermined borders over his face, trunk, and legs for 2 months (Fig. 1A, 1B). The skin lesions responded poorly to antibiotics. Laboratory examinations showed leukocytosis with monocytosis, anemia, and thrombocytopenia. A biopsy from the edge of one ulcer showed hemorrhagic necrosis and a dense infiltration of neutrophils extending throughout the dermis to the subcutis (Fig. 2). A chest X-ray showed several soft-tissue masses at the left upper lung field (Fig. 3A). Cultures for bacteria, fungi, and mycobacteria of the skin and the lung lesions were all negative. A bone marrow aspirate confirmed the diagnosis of chronic myelomonocytic leukemia (CMMoL).

The skin and lung lesions improved promptly after pulse therapy with methylprednisolone (Fig. 1C, 1D, 3B). However, the patient died on acute leukemic transformation of the CMMoL and sepsis 17 months later.

Fig. 1
(A) Multiple destructive ulcers with elevated borders on the face.
(B) Ulcerative lesions on the chest wall. Note the halo of erythema surrounding the ulcer.
(C, D) The ulcer healed gradually 2 months after the treatment, leaving some cribriform scars.

Fig. 2
(A) Skin biopsy from the border of the ulcer shows dense inflammatory infiltrate consisting of neutrophils and a few lymphocytes in the dermis. The infiltrate extends from the base of ulcer to the undermined epidermis (H&E, x40).
(B) Extensive hemorrhagic necrosis of the dermis, and thrombosis of the vessels (H&E, x100).

Fig. 3
(A) Several lobulated soft-tissue masses are demonstrated at the left upper thorax.
(B) The pulmonary lesion resolved after steroid treatment revealed by the follow-up chest X-ray 2 months later.
DIAGNOSIS: Pyoderma Gangrenosum with Lung Involvement in Association with Chronic Myelomonocytic Leukemia

DISCUSSION

Pyoderma gangrenosum (PG) is a rare, non-infectious, reactive neutrophilic dermatoses which typically starts with pustules that rapidly evolve into painful ulcers with undermined violaceous borders. Since its first description in 1930, the pathogenesis of PG has remained poorly understood even though a widening range of associated systemic diseases have been described.

Approximately 50% of PG patients have an associated systemic disease, most often inflammatory bowel disease, arthritis, paraproteinemia, hematologic disorders, and malignancy. For patients with hematologic disease, the appearance of PG may herald a more aggressive phase or malignant transformation of the underlying hematologic disease, indicating the need for full systemic review on these patients.

Although extracutaneous manifestations are unusual in PG, visceral involvement may occur, including the lungs, bones, spleen, liver, central nervous system, muscles, lymph nodes, and eyes. Lung is the most frequently involved organ. The visceral involvement may appear before, simultaneously or after cutaneous manifestations. Extracutaneous manifestations may be a sign of associated pathology, in particular hematologic disorder. The diagnosis of extracutaneous involvement of PG is usually difficult, requiring extensive investigations to eliminate a neoplastic or infectious process. In our patient, the pulmonary lesion was noted concurrently with the cutaneous involvement of PG. The prompt diagnosis of cutaneous PG in our case allows an appropriate therapeutic approach, i.e., the initiation of corticosteroid therapy. Both the cutaneous and extracutaneous manifestations promptly resolve. Accordingly, we concluded that the pulmonary lesion in our patient was an extracutaneous manifestation of PG.

The histopathological finding of PG is non-specific, which shows a dense neutrophilic infiltrate, often with abscess formation. The diagnosis of PG is based on the clinical and pathological features and requires exclusion of other causes of cutaneous ulceration. Systemic steroids have been the mainstay in the therapy of PG. Other treatment modalities include minocycline, sulfasalazine, dapsone, immunosuppressive agents. Topical therapies include hydrocolloid dressings, intralesional corticosteroid injections, topical steroids and topical tacrolimus. The management of PG remains a challenge because of its persistent and recurrent nature.

In summary, we report a case of PG presenting with both cutaneous and pulmonary lesions, and is associated with CMMoL. Considering the risk of progressing toward acute leukemia, a close follow-up of patients with PG and hematologic disorder is warranted. In addition, physicians should be aware of the potential visceral involvement in patients with PG that justifies an appropriate work-up.

REFERENCES