

Ulcerative Sarcoidosis: An Atypical Cause of Leg Ulcers.

Kazuki Miyaue¹, Testuya Hoshi², and Hiroki Isono³

¹HITO Medical Center

²Teine Keijinkai Hospital

³HITO medal center

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CASE

An 83-year-old Japanese woman presented with multiple chronic painful pretibial ulcers on both extremities that first appeared 3 years earlier, as diagnosed by a dermatologist. She reported generalized edema and blurred vision, without fever, night sweats, weight loss, or arthralgia. Four years prior to presentation, she had bilateral hilar adenopathy, which was diagnosed as an epithelioid cell granuloma through biopsy. Upon examination, the ulcers displayed asymmetric features with violaceous borders and necrotic bases, while the surrounding tissues appeared erythematous and edematous (Figure 1). The patient also exhibited erythematous plaques on both arms and bilateral uveitis. Laboratory tests revealed an elevated eosinophil count of 2630 μ L (reference range: 70-440) and an elevated serum angiotensin-converting enzyme (ACE) concentration of 25.5 U/L (reference range: 8.3–21.4). Antinuclear antibody test, antineutrophil cytoplasmic antibody test, interferon gamma release assays, and serological tests for syphilis yielded negative results. Deep vein thrombosis, vascular insufficiency and cardiac abnormalities were ruled out upon ultrasonography. Plaque biopsy of the right arm revealed perivascular dermatitis with giant cells. As further evaluation ruled out other possibilities, the patient was diagnosed with sarcoidosis and initiated on systemic prednisolone (0.7 mg/kg/day). Subsequently, the prednisolone dose was gradually tapered to 5 mg/day, leading to complete healing of the ulcers within 12 months (Figure 2).

DISCUSSION

In this case, the ulcers exhibited atypical characteristics. Clinical signs of atypical ulcers include a necrotic wound bed, a purple border, surrounding inflammation, an unusual site such as the proximal calf, asymmetry sites and severe pain¹. The differential diagnosis of atypical ulcers includes external, neoplastic, vasculopathic, hematologic, infectious, drug-induced, and inflammatory etiologies, including sarcoidosis.¹ Atypical ulcers are differentiated from typical ulcers caused by venous insufficiency, diabetes, ischemia, or pressure¹.

Sarcoidosis is a granulomatous disease affecting multiple organs. Cutaneous involvement occurs in approximately 20%–35% of patients with sarcoidosis². Common skin manifestations include maculopapules, nodules, plaques, infiltrative scars, lupus pernio (specific lesions), and erythema nodosum (nonspecific lesion)². Only 4.8% of patients with skin lesions develop skin ulcers. The diagnosis of sarcoidosis is established based on clinical and histopathological findings after excluding other potential causes. Differential diagnoses for ulcerative sarcoidosis include necrobiosis lipoidica, atypical mycobacterial infection, malignancy, and trauma^{2,3}. Although a biopsy of the ulcerative lesions was not performed due to concerns about delayed skin healing, we suspected sarcoidosis based on the ulcer features, such as necrotic ulcers with violaceous rolled borders in the pretibial area². Additionally, the patient's history of hilar lymphadenopathy, bilateral uveitis, elevated ACE concentration, and histopathological analysis of the lymph nodes supported the diagnosis. The patients was finally diagnosed with ulcerative sarcoidosis based on her rapid response to corticosteroids.

References

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