Received: 16 June 2023 | Revised: 8 February 2024 | Accepted: 8 February 2024

DOI: 10.1002/ccr3.8592

CASE IMAGE

Clinical Case Reports Open Access WILEY

Ulcerative sarcoidosis: An atypical cause of leg ulcers

Kazuki Miyaue[1] | **Tetsuya Hoshi**[2] | **Hiroki Isono**[1]

[1]Department of General Medicine, HITO Medical Center, Ehime, Japan

[2]Department of General Internal Medicine, Teine Keijinkai Hospital, Sapporo, Japan

Correspondence
Kazuki Miyaue, Department of General Medicine HITO Medical Center, 788-1, Kamibuncho, Shikokuchuo Shi, Ehime, 799-0121, Japan.
Email: miyauekazuki@gmail.com

Key Clinical Message

We present the case of an 83-year-old woman with leg ulcers who was diagnosed with sarcoidosis. This case highlights the importance for clinicians to consider ulcerative sarcoidosis when encountering patients with leg ulcers along with hilar adenopathy, uveitis, elevated serum angiotensin-converting enzyme, and histopathological findings of epithelioid cell granulomas.

KEYWORDS

corticosteroids, differential diagnosis, leg ulcers, sarcoidosis, skin

1 | INTRODUCTION

Sarcoidosis is a complex, multisystemic disease characterized by the formation of non-necrotizing granulomas affecting any organ.[1] It manifests in genetically predisposed individuals, triggered by environmental factors, leading to an immune response marked by granuloma formation.[1] Herein, we report a case of Japanese woman presenting with atypical leg ulcers, who was eventually diagnosed with sarcoidosis.

2 | 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. Her medical history was significant for hypertension and cholelithiasis. Her medications included amlodipine, spironolactone, azosemide, lansoprazole, loxoprofen, famotidine, and edoxaban (for possible DVT).

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 μL (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 (ANA) test, antineutrophil cytoplasmic antibody (ANCA) test, interferon gamma release assays (T-SPOT), and serological tests for syphilis yielded negative results. Deep vein thrombosis, vascular insufficiency, and cardiac abnormalities were ruled out upon ultrasonography. A plaque biopsy of the right arm revealed perivascular dermatitis with giant cells.

A history of hilar adenopathy, leg ulcers, histopathology of the lymph nodes (epithelioid cell granulomas) and the skin of the right arm (perivascular dermatitis with giant cells), uveitis, eosinophilia, and elevated serum ACE levels suggested sarcoidosis. The patient was 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).

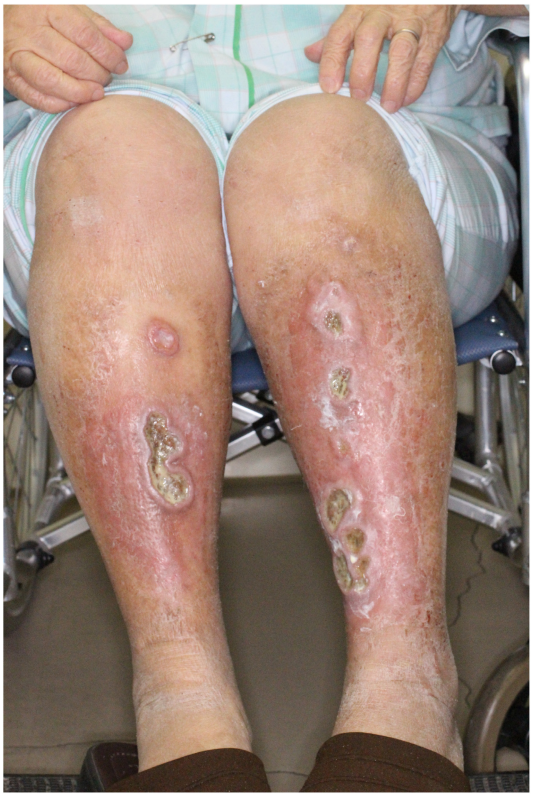

FIGURE 1 The ulcers exhibited asymmetry and displayed violaceous borders with necrotic bases, accompanied by erythematous and edematous surrounding tissues.

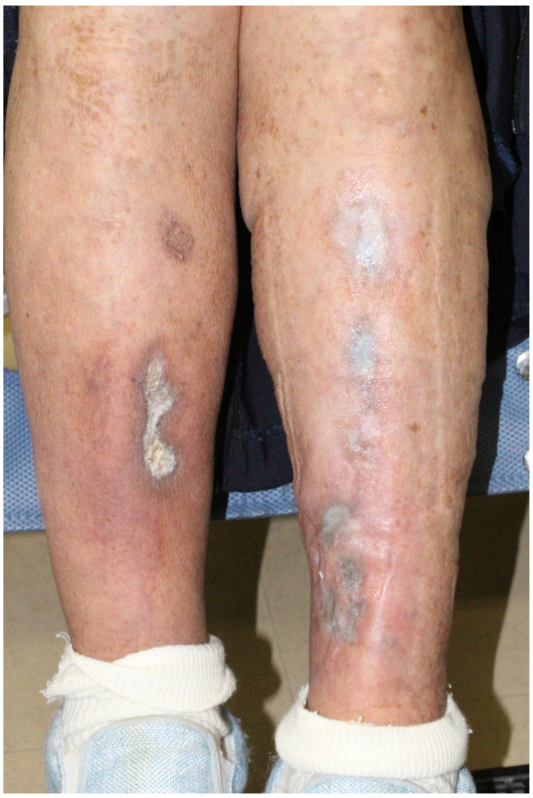

FIGURE 2 The ulcers were healed within a 12-month period.

3 | 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 pain.[2] The differential diagnosis of atypical ulcers includes external, neoplastic, vasculopathic, hematologic, infectious, drug-induced, and inflammatory etiologies, including sarcoidosis.[2] Atypical ulcers are differentiated from typical ulcers caused by venous insufficiency, diabetes, ischemia, or pressure.[2]

Cutaneous involvement occurs in approximately 20%–35% of patients with sarcoidosis.[3] Common skin manifestations include maculopapules, nodules, plaques, infiltrative scars, lupus perio (specific lesions), and erythema nodosum (nonspecific lesion).[3] A review of the literature[4] showed that only seven out of 147 patients with sarcoidosis developed skin ulcers. Four of the seven had hilar adenopathy, and four had elevated ACE levels, which the patient in this case had. None of the seven patients had uveitis, which was present in this case. The diagnosis of sarcoidosis is established based on clinical and histopathological findings after excluding other potential causes. Histopathological features of cutaneous sarcoidosis include noncaseating granulomas with epithelioid macrophages and multinucleated giant cells.[5] Differential diagnoses for ulcerative sarcoidosis include necrobiosis lipoidica, atypical mycobacterial infection, malignancy, and trauma.[3,6]

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 erea.[3] Additionally, the patient's history of hilar lymphadenopathy, bilateral uveitis, elevated ACE concentration, and histopathological analysis of the lymph nodes and the skin of the right arm supported the diagnosis. Tuberculosis, syphilis, ANCA-associated autoimmune diseases, ANCA-associated vasculitides, and vascular insufficiencies were ruled out by T-SPOT, serologies, and ultrasound. We couldn't completely rule out rare infectious diseases such as leishmaniasis, malignancies, and some dermatological conditions such as Kyrle's disease, but the patient was finally diagnosed with ulcerative sarcoidosis because of her rapid response to corticosteroids.

AUTHOR CONTRIBUTIONS

Kazuki Miyaue: Conceptualization; data curation; project administration; writing – original draft; writing – review and editing. **Tetsuya Hoshi:** Supervision. **Hiroki Isono:** Supervision; writing – review and editing.

ACKNOWLEDGMENTS

The authors acknowledge the use of ChatGPT (GPT-4 September 25 version) for initial brainstorming and generating preliminary drafts. All data interpretations and final manuscript revisions were done by human researchers.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

None.

DATA AVAILABILITY STATEMENT

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

DISCLAIMER

The views expressed in the submitted article are the individual's own and not the official position of the institution or the funder.

ETHICS STATEMENT

All procedures performed were in accordance with ethical standards. The examination was made in accordance with the approved principles.

CONSENT

This article is published with the written consent of the patient.

ORCID

Kazuki Miyaue https://orcid.org/0000-0002-7735-9308
Hiroki Isono https://orcid.org/0000-0002-6593-8740

REFERENCES

1. Llanos O, Hamzeh N. Sarcoidosis. *Med Clin North Am*. 2019;103(3):527-534.
2. Nickles MA, Tsoukas MM, Sweiss N, Ennis W, Altman IA. Atypical ulcers: a stepwise approach for clinicians. *Wounds*. 2022;34(10):236-244.
3. Fernandez-Faith E, McDonnell J. Cutaneous sarcoidosis: differential diagnosis. *Clin Dermatol*. 2007;25(3):276-287.
4. Yoo SS, Mimouni D, Nikolskaia OV, Kouba DJ, Sauder DN, Nousari CH. Clinicopathologic features of ulcerative-atrophic sarcoidosis. *Int J Dermatol*. 2004;43(2):108-112.
5. Wanat KA, Rosenbach M. A practical approach to cutaneous sarcoidosis. *Am J Clin Dermatol*. 2014;15(4):283-297.
6. Wanat KA, Rosenbach M. Cutaneous sarcoidosis. *Clin Chest Med*. 2015;36(4):685-702.

How to cite this article: Miyaue K, Hoshi T, Isono H. Ulcerative sarcoidosis: An atypical cause of leg ulcers. *Clin Case Rep*. 2024;12:e8592. doi:10.1002/ccr3.8592