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Sarcoidosis with the livedo reticularis-like appearance

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Abstract. Sarcoidosis is a multisystem inflammatory disease manifesting as noncaseating epithelioid cell granulomas. 25 to 30% of individuals with systemic sarcoidosis show variable cutaneous manifestations. A 59-year-old female was seen with reticular purplish-red nodules and plaques on the legs for three months. A skin biopsy of the livedo area revealed non-caseating epithelioid cell granulomas surrounding blood vessels in the dermis. She was diagnosed with sarcoidosis livedo, and cutaneous lesions subsided with oral prednisone. Sarcoidosis livedo (SL) assumes a uncommon livedo reticularis-like presentation. This is the first Chinese patient with SL, and more patients are needed to unveil the unique characters of SL.

Key words: sarcoidosis, reticular, nodule, plaque, livedo reticularis

Introduction

Sarcoidosis is a multisystem inflammatory disease of unknown cause that manifests as noncaseating epithelioid cell granulomas. 25 to 30% of individuals with systemic sarcoidosis show variable cutaneous manifestations (1), and only 15 cases have been reported with livedo reticularis-like appearance. We herein describe the first Chinese case with similar presentations.

Case report

A 59-year-old female was referred for reticular purplish-red nodules and plaques on the legs for three months. She had bilateral uveitis for two years. On examination, there were purplish-red nodules and plagues in a reticular configuration, giving the skin a livedo reticularis-like appearance (Figure 1).

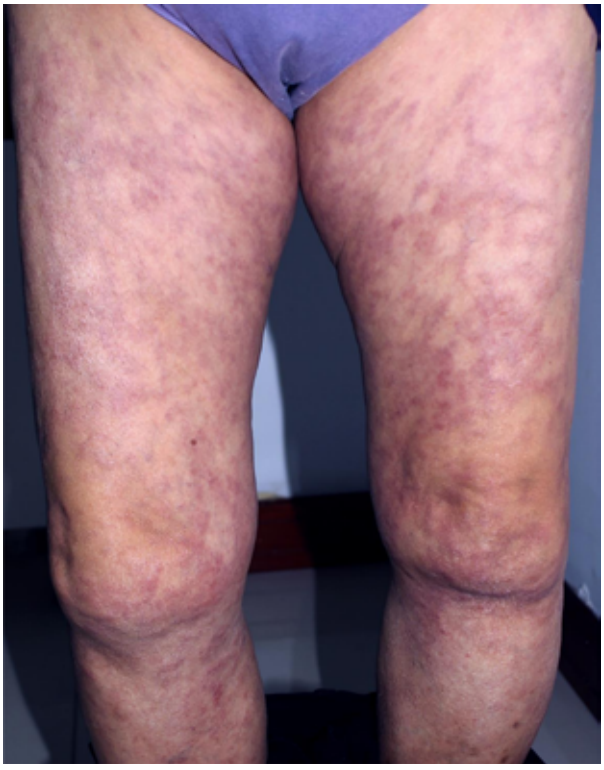

Figure 1. Purplish-red nodules and plagues with mild tenderness on both legs, giving the skin a livedo reticularis-like appearance.

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Skin specimen of the lesion revealed non-caseating epithelioid cell granulomas composed of histiocytes and giant cells with a sparse lymphocytic infiltrate at the margins (Figure 2 and 3). Ziehl-Neelsen and Periodic acid-Schiff staining did not identify any bacilli. The tuberculin test and the antinuclear antibody were negative. The serum angiotensin-converting enzyme was slightly elevated. Computed tomography of the chest revealed a diffuse bilateral distribution of multiple centrilobular nodules in the left lung's inferior lobe. She was diagnosed with sarcoidosis livedo (SL), and the lesions subsided with oral prednisone 30mg/day for two months.

Discussion

The cutaneous manifestations of sarcoidosis are notoriously diverse and may classically arise to be erythematous, papular, maculopapular, nodular, plaque, atrophic, angiolupoid, ichthyosiform, lichenoid, annular lesions. It may mimic a broad range of other skin disorders consisting of lupus pernio, scar, erythema nodosum, alopecia, erythroderma, and hypopigmentation (2). Sarcoidosis that atypically assumes a livedo reticularis-like change is defined as sarcoidosis livedo (SL) (3,4). As far as I knew, there are only 16 such cases (including this report) have been reported in English literature, and all of them were females. Many SL cases are Japanese and usually showed ocular and neurological comorbidities (3). There is a report that SL may be shifted from erythema nodosum-like lesions.

Because there is no pathognomonic sign, a histopathological investigation is crucial for excluding other granulomatous diseases. The diagnosis of sarcoidosis is established with the presence of noncaseating granuloma on a biopsy specimen (2). Blood examination showed elevated serum levels of angiotensin II-converting enzyme. Other skin disorders that can manifest as livedo reticularis include polyarteritis nodosa, vasculitis, systemic lupus erythematosus, rheumatoid arthritis, and other

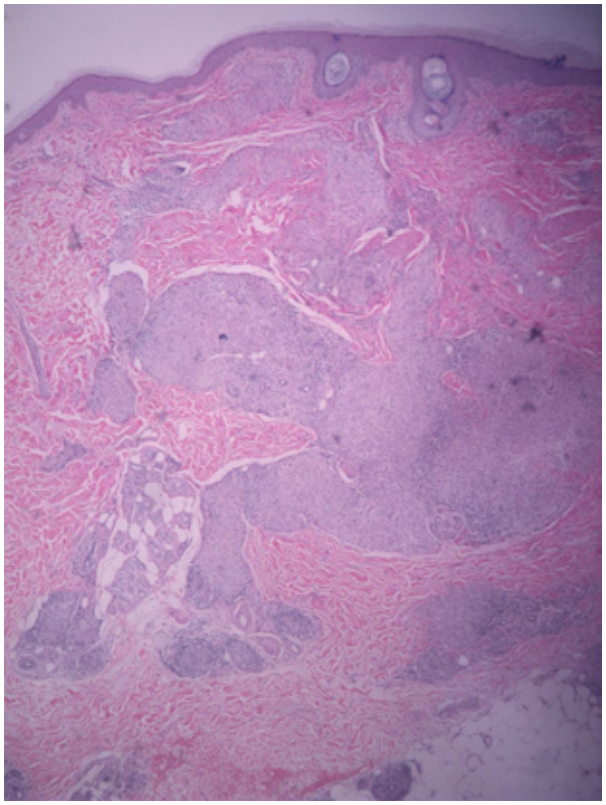

Figure 2. Non-caseating epithelioid cell granulomas surrounding blood vessels throughout the dermis and the subcutaneous tissues (H&E stain, original magnification ×40).

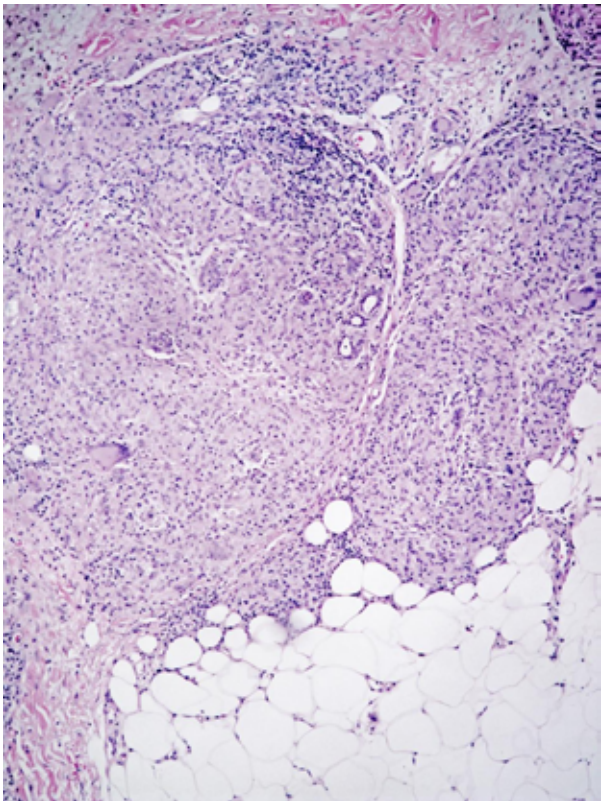

Figure 3. The granulomas were composed of histiocytes, and giant cells with a sparse lymphocytic infiltrate at the margins (H&E stain, original magnification ×200).

connective tissue disease (2,3). The clinical and histological features in our patients made them unlikely.

Macrophages play a pivotal role in sarcoidal granuloma formation via cytokines such as Interferon-γ, tumor necrosis factor-α, interleukin-12, and interleukin-18 after its activation (2). As for SL, it is proposed that damage to blood vessels or obstruction of its lumen may compromise the circulation caused by the infiltration of sarcoid granuloma and may ultimately attribute to the clinically apparent livedo (3).

The treatment regimen for sarcoidosis is tailored based on the severity, comorbidities, and patient preference. Glucocorticoid therapy may efficiently control the symptoms (2,5).

Conclusions

Sarcoidosis livedo is a uncommon skin presentations of sarcoidosis, We report the first Chinese patient with SL to highlight its unique characters.

Conflicts of Interest: All authors have seen and agree with the contents of the manuscript and there is no financial interest to report. We certify that this submission is original work and is not under review at any other publication. There is no conflict of interest.

References

1. Tripipitsiriwat A, Komoltri C, Ruangchira-Urai R, Ungprasert P. Clinical Characteristics of Sarcoidosis in Asian Population: A 14-year Single Center Retrospective Cohort Study from Thailand. Sarcoidosis Vasc Diffuse Lung Dis. 2020;37:e2020011.
2. Caplan A, Rosenbach M, Imadojemu S. Cutaneous Sarcoidosis. Semin Respir Crit Care Med. 2020;41:689-99.
3. Bittencourt MJS, Anjos ABD, Brito F, Unger DAA. Cutaneous sarcoidosis with livedo-like manifestation. An Bras Dermatol. 2019;94:493-4.
4. Hayashi S, Hatamochi A, Hamasaki Y, et al. A case of sarcoidosis with livedo. Int J Dermatol. 2009;48:1217-21.
5. Mana J, Marcoval J, Rubio M, Labori M, Fanlo M, Pujol R. Granulomatous cutaneous sarcoidosis: diagnosis, relationship to systemic disease, prognosis and treatment. Sarcoidosis Vasc Diffuse Lung Dis. 2013;30:268-81.