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CASE REPORT

Sarcoidosis with bilateral leg lymphedema as the initial presentation: a review of the literature

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ABSTRACT

Sarcoidosis is a granulomatous disease, characterized microscopically by noncaseating granulomas, which may involve multiple organs; however, the lung, skin, and lymph nodes are commonly affected. Sarcoidosis is a great imitator; in the skin, it presents with different cutaneous manifestations including lupus pernio, infiltrated plaques, maculopapular eruptions, infiltration of old scars, and subcutaneous sarcoidosis. Lymphedema as an initial presentation is extremely rare; cases are reported in African-American but not Asian patients. Lymphedema associated with sarcoidosis may result from lymphatic obstruction by infiltrating sarcoidosis. We present a case where the symptoms and signs of sarcoidosis were improved after treatment with systemic steroids.

Introduction

Sarcoidosis is a systemic granulomatous disease of unknown etiology. The skin and lungs are the organs most commonly affected by sarcoidosis. In the skin, typical manifestations are infiltrated plaques, maculopapular eruptions, or subcutaneous involvements. Sarcoidosis with initial presentation of edema in bilateral lower extremities is extremely rare. This report presents the first case of sarcoidosis with initial presentation of leg edema in an Asian woman.

Case report

The patient was a 48-year-old housewife with no prior systemic diseases. She visited our outpatient department due to bilateral leg edema (Figure 1A). The edema was noted to gradually progress from both feet to both legs, and was accompanied by joint pain over the shoulders, ankles, and knees. The leg edema was mild pitting without significant surface change. An indurated noduloplaque lesion approximately 2 cm × 4 cm and associated with itching and tenderness was present on her right forearm (Figure 1B). There was no fever, dyspnea, blurred vision, or mucosal involvement. Physical examination showed bilateral clear breathing sounds and a regular heartbeat. No lymphadenopathy was palpable. However, multiple subcutaneous asymptomatic nodules developed over both legs in the subsequent examination (Figure 1C). Biopsy of the right forearm plaque revealed multiple noncaseating granulomas with fibrosis and cell infiltration in the subcutaneous fat. The granulomas were composed of multinucleated giant cells and a few histiocytes with chronic inflammatory cells (Figure 2). Special stains for foreign body, acid-fast bacilli, and fungi were all negative, rendering the pathologic diagnosis of sarcoidosis. Skin biopsy of another nodule on the leg also revealed multiple similar, dense, uniform, circumscribed nests of noncaseating granulomas infiltrating the dermis.

Blood examinations showed a twofold increase in the level of angiotensin-converting enzyme (45.85 IU/L, normal range < 22.5 IU/L). The tests for antinuclear antibody and rheumatoid factor were negative. The levels of complement components C3 (126.00 mg/dL) and C4 (29.80 mg/dL) were within the normal ranges. Complete blood cell counts and blood levels of urea nitrogen, creatinine, liver enzymes, and electrolytes were all within the normal ranges. Chest radiography revealed a left hilar nodule of approximately 0.8 cm. Additional chest computed tomography showed mildly enlarged lymph nodes (all < 1.3 cm) in the mediastinal and bilateral hilar regions, confirming the diagnosis of

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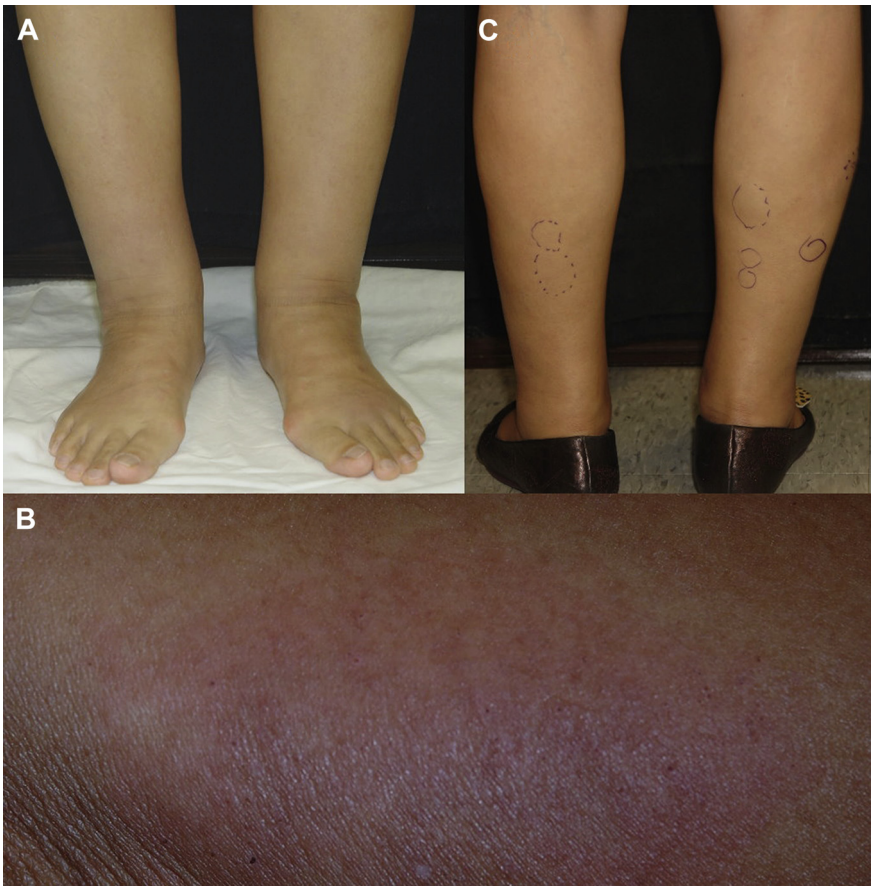

Figure 1 (A) Initial edema and ankle swelling on both lower legs. (B) Indurated and erythematous noduloplaque at the right forearm. (C) Multiple asymptomatic subcutaneous nodules over bilateral lower legs.

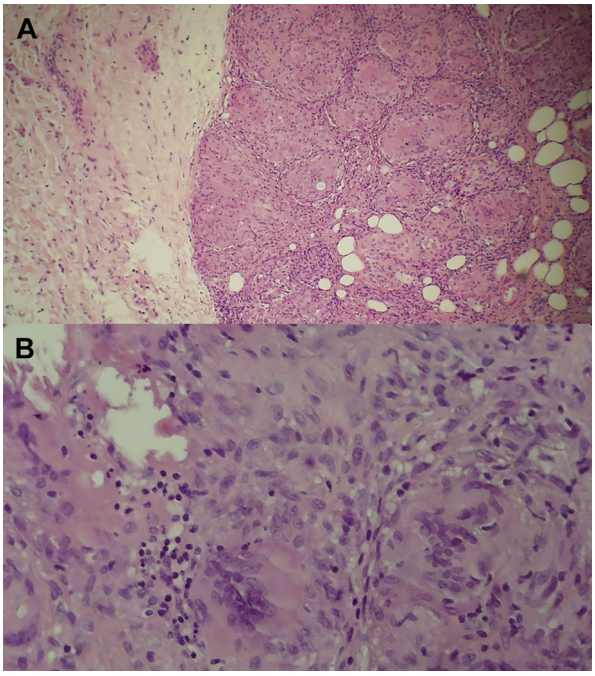

Figure 2 (A) Multiple noncaseating granulomas associated with fibrosis and mild infiltration in the subcutaneous fat (hematoxylin and eosin, ×100). (B) High-power field shows typical noncaseating naked granuloma (hematoxylin and eosin, ×200).

sarcoidosis (Figure 3). The patient was treated with methylprednisolone at 0.5 mg/kg/d. The bilateral leg edema was significantly improved within 10 days and the subcutaneous nodules over the legs and right forearm were also resolved.

Discussion

Sarcoidosis is a common systemic, noncaseating granulomatous disease of unknown etiology. There is a higher incidence rate among African Americans and in women compared to men.[1] Cutaneous sarcoidosis manifests itself in approximately 20–35% of patients with sarcoidosis; however, lymphedema is a rare initial manifestation of sarcoidosis.

Currently, few cases of sarcoidosis with initial presentation of leg edema have been reported in African Americans[2,3]; however, no cases have been reported in Asians. The mechanisms of sarcoidosis-related leg edema are unknown; suggested etiologies include sarcoidosis-related lymphadenopathy (direct sarcoidosis infiltration or lymph duct obstruction by soft tissue sarcoidosis) and tenosynovitis. In the case presented here, the leg edema may be related to multiple soft-tissue sarcoidosis that led to upstream lymphatic duct obstruction.

Furthermore, elevated serum angiotensin-converting enzyme (ACE) levels were observed in our case. ACE converts angiotensin I into angiotensin II and inactivates bradykinin via the kallikrein-kininogen system. Elevated serum ACE levels are present in sarcoidosis; elevated levels may result from the production of ACE