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Subcutaneous Sarcoidosis of the Distal Lower Leg in a Middle-Aged Woman Associated with Pulmonary Sarcoidosis: a Case Report

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Case Report

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Sarcoidosis is a systemic disease of unknown etiology that can involve almost any organ systems, characterized by the presence of non-caseating granulomas in affected organs. Typically, the lungs and mediastinal lymph nodes are the most-affected sites, with cutaneous manifestations being the second. Subcutaneous nodules are a rare manifestation of cutaneous sarcoidosis, and it is even rarer for subcutaneous sarcoidosis to be associated with pulmonary sarcoidosis. Here, we present a case of subcutaneous sarcoidosis of the distal lower leg associated with pulmonary sarcoidosis.

Keywords: Subcutaneous; Pulmonary; Sarcoidosis; Leg; Middle Age; Adult

INTRODUCTION

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology characterized by a development of non-caseating granulomas in various organs, such as lung, brain, kidneys, heart, skin, lymph nodes, and joints (1). Skin involvement is the second most common following pulmonary involvement in sarcoidosis, which may develop in 20-35% of patients (1).

A subcutaneous nodule is an uncommon manifestation of skin involvement of sarcoidosis, with incidence ranging from 1.4 to 6% (2). It is defined clinically by asymptomatic, non-tender, and flesh-colored nodules that typically range in size ranging from 0.5 to 2.0 cm (2). This infrequent form of cutaneous sarcoidosis is usually associated with bilateral hilar lymphadenopathy, but rarely with other organ systems, especially the lungs (3-5). Here, we report a unique case of subcutaneous sarcoidosis where lung, mediastinal, and bilateral hilar lymph nodes are involved at the same time, with initial manifestation of lower-leg soft-tissue masses.

CASE REPORT

A 48-year-old woman presented with palpable masses in her left distal lower leg,

noticed about a month before the hospital visit. She also had a two-year history of dyspnea and cough. On physical examination, multiple non-tender, fixed nodular masses were present in her left distal lower leg (Fig. 1). Inspiratory crackles were heard in her right lower lung field. She had no history of trauma, and laboratory investigations were within normal range, including complete blood cell count, erythrocyte sedimentation rate, C-Reactive protein, and serum angiotensin-converting enzyme level. Her initial plain chest radiograph revealed fine reticular opacities in both apical and lower lungs with bulging opacity in both hilar areas (image not shown). Her thoracic computed tomography (CT) scan showed multiple discrete nodules in both upper lungs, ground-glass opacities (GGOs), reticular densities in the peripheral portion of both lungs with lower dominancy, and relatively symmetric mediastinal and bilateral hilar lymphadenopathies (Fig. 2). On extremity magnetic resonance imaging (MRI), we noted multiple nodules with inhomogenous T1, T2 iso to slightly high signal intensity (SI) compared to skeletal muscle, with infiltrating margins and internal reticular patterns within a subcutaneous fat layer of the left distal lower leg. These nodules were diffusely well enhanced with internal non- to less-enhancing septa-like structures, and showed adjacent deep fascial thickening and infiltration (Fig. 3). They were very closely abutting the extensor digitorum tendon at the anterior ankle, but we could see no definite extension to adjacent muscles. For the differential diagnosis, we first considered a chronic inflammatory disorder involving lymph nodes, such as Kimura's disease, and then lymphoma. The patient underwent an excisional biopsy at the left lower-leg mass. The histopathological findings were consistent with those of sarcoidosis, given the presence of non-caseating granulomas packed with epithelioid cells and lymphocytes on hematoxylin and eosin stained images (Fig. 4). We also did a bronchoscopic biopsy on subcarinal lymph nodes in the mediastinum, which also revealed non-caseating granulomas (image not shown). Thus, we diagnosed the patient as having subcutaneous sarcoidosis associated with pulmonary sarcoidosis. We prescribed 1 mg/kg/day prednisone, which resolved the dyspnea and cough. The palpable masses also shrank, and the thoracic imaging abnormalities were normalized.

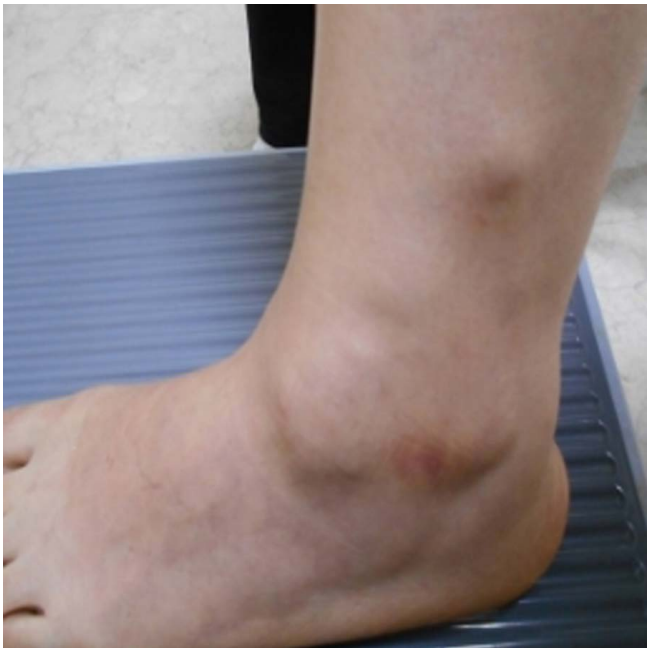


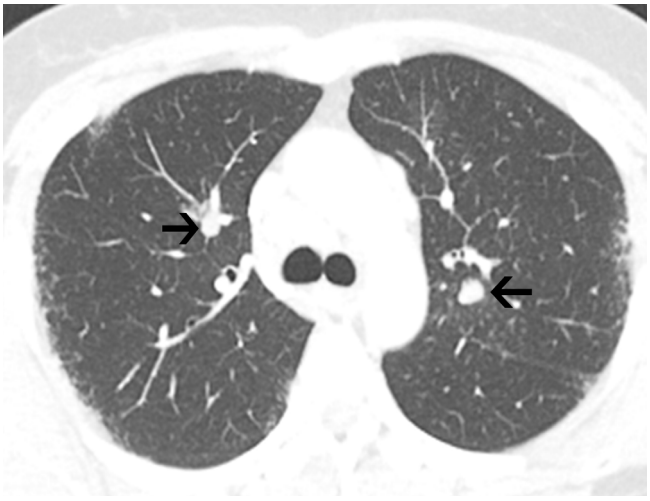
Fig. 1. Subcutaneous nodules with normal overlying skin on the left distal lower leg.

DISCUSSION

Sarcoidosis is a systemic disease characterized by non-caseating granulomas. Usually, the lungs are the primary site of the disease, with skin being the second most common site (3). Cutaneous sarcoidosis can be classified as specific and nonspecific types. The lesions specific for cutaneous sarcoidosis include nodules, maculopapules, plaque, subcutaneous nodules, infiltrative scar, and lupus pernio. Nonspecific types include erythema nodosum, prurigo, calcifications, erythema multiforme, and nail changes, such as clubbing, onycholysis, and subungual hyperkeratosis. Unlike the specific types, nonspecific types do not display non-caseating granulomas histologically (2).

Subcutaneous sarcoidosis is a rare form of the cutaneous sarcoidosis and is found in less than 6% of patients with sarcoidosis (2). It is most commonly associated with bilateral hilar lymphadenopathies, reaching 94% of cases in some studies (6). However, its association with the pulmonary system is exceptionally rare, with only 20 such cases reported in the literature published on PubMed (3, 4). It is thought to appear in the early stage of sarcoidosis, which makes it useful for diagnosis of systemic sarcoidosis (1). Yet this was not case with our patient, who might have had pulmonary manifestations prior to cutaneous lesion, considering the precedent pulmonary symptoms.

Imaging findings of subcutaneous sarcoidosis were as follows. On CT, it can appear as either a nodular or a diffuse pattern. The nodular form is well-defined and homogeneous, with a density similar to that of the muscle.



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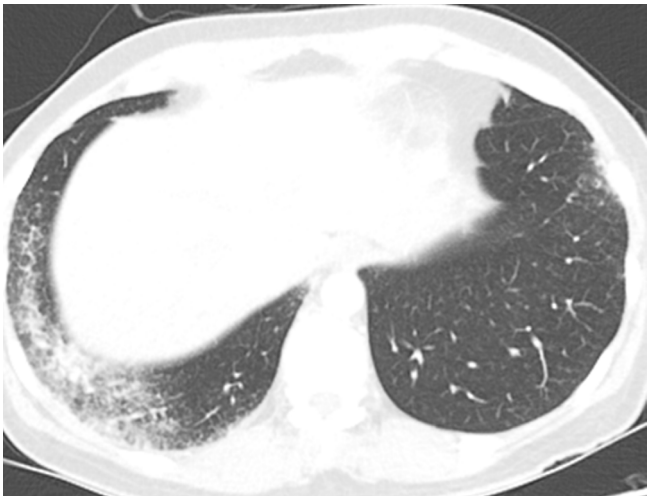

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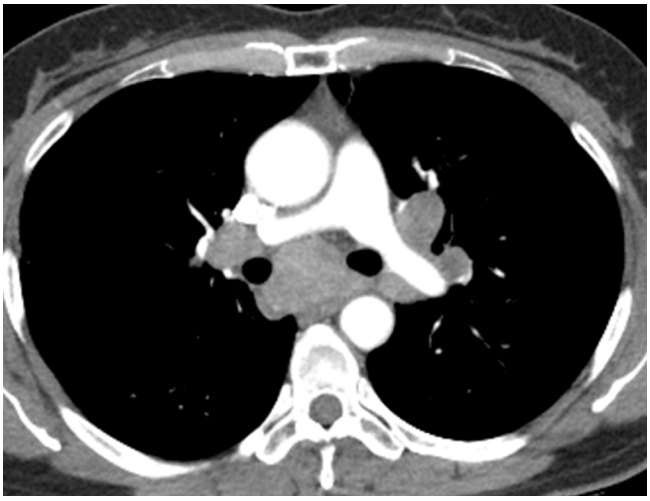

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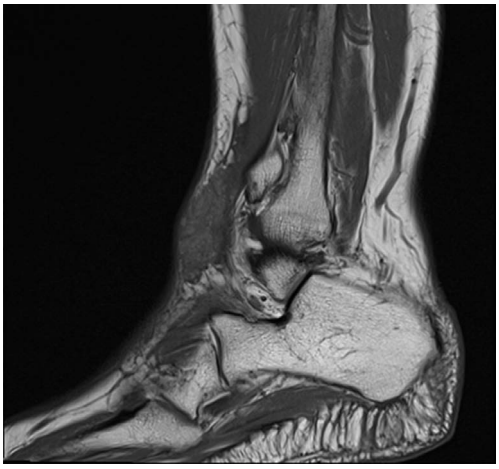
Fig. 2. Thoracic computed tomography (CT) findings of sarcoidosis in lung parenchyme and mediastinal lymph nodes. (a) The parenchymal window setting shows multiple discrete nodules in both upper lungs (arrows). (b) GGOs and reticular densities are also seen in both lower lungs. (c) In the mediastinal window, multiple enlarged LNs are noted in mediastinum and both hilar areas, in relatively symmetric fashion.

The diffuse form appears as ill-defined and heterogeneous, with a honeycomb-like pattern.

On MR, the nodular form shows a homogeneous iso-signal intensity like that of muscle on T1-weighted images and a heterogeneous hyper-signal intensity on T2-weighted images, with homogeneous enhancement. The diffuse form manifests a mesh or stripe pattern of intermediate SI on both T1 and T2 WI with slight enhancement on contrast-enhanced images (5). Although sarcoidosis rarely involves fascia, there are two reports where a nodular form of subcutaneous sarcoidosis involved both subcutaneous tissue and adjacent fascia (7, 8). In the reports, both authors considered the possibility of the presence of a malignant subcutaneous tumor, such as malignant fibrous histiocytoma or sarcoma. Therefore, it should be differentiated from malignant soft-tissue tumors, because it can breach the fascia.

For the differential diagnosis, other superficial soft-tissue masses, such as rheumatoid nodules, nodular fasciitis, vascular tumors, peripheral nerve-sheath tumors, and pathology involving lymph nodes such as lymphoma, can be considered. Rheumatoid nodules can be seen as a predominantly solid nodule or a nodule with a cystic portion. A rim-enhancing nodule in a subcutaneous tissue over prominent bones that are exposed to repetitive mechanical stress can be a clue to the diagnosis of a rheumatoid nodule (9). For nodular fasciitis, the linear extension of the lesion along the fascia, known as a 'fascial tail sign,' is an important diagnostic feature at MR imaging. A vascular tumor, such as hemangioma, may present as an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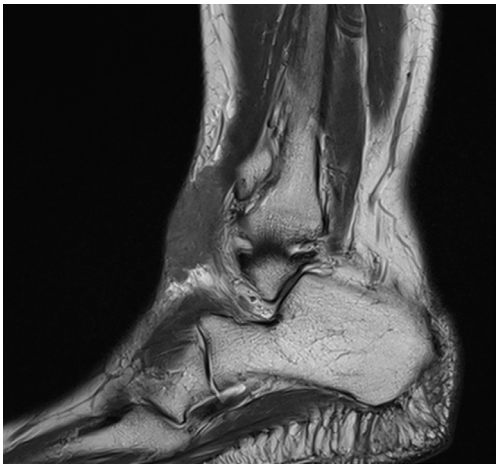
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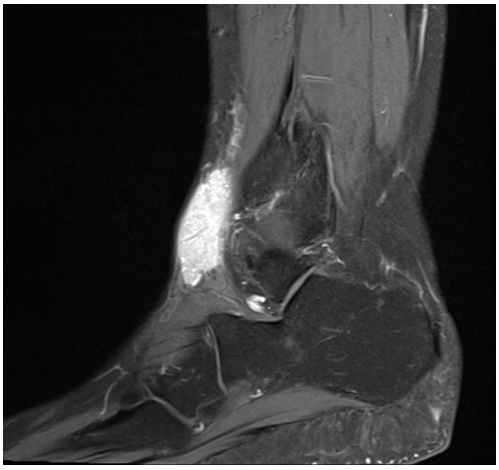
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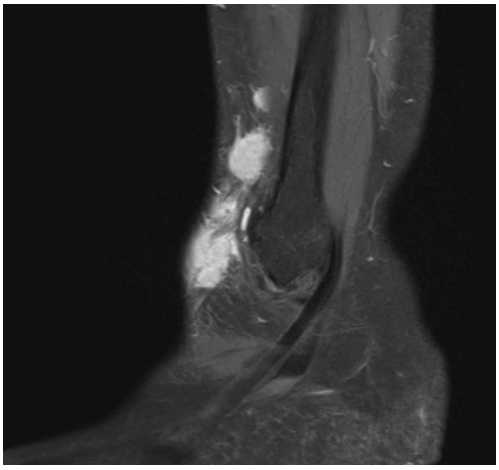
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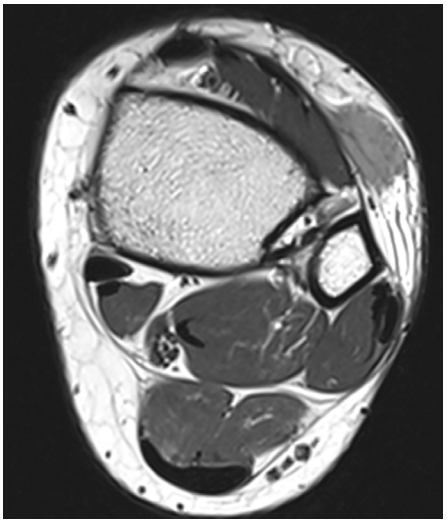
Fig. 3. Magnetic resonance (MR) findings of subcutaneous sarcoidosis. (a-d) Sagittal T1-weighted, T2-weighted, fat suppression T1-weighted contrast-enhanced images and (e-g) axial T2-WI, FS T2-WI, FS T1-CE images show multiple irregularly shaped, T1 and T2 intermediate-high signal relative to skeletal muscle with reticular patterns, well-enhancing masses within the subcutaneous fat layer of the left distal lower leg, with adjacent deep fascial thickening and edema. Internal non- to less-enhancing linear structures are also noted within the lesion.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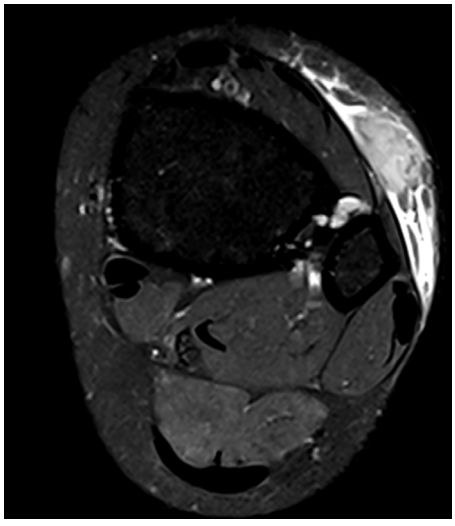
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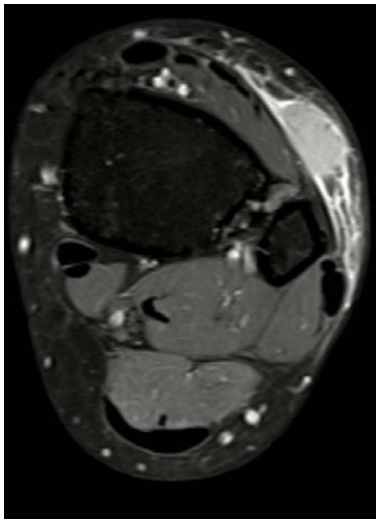
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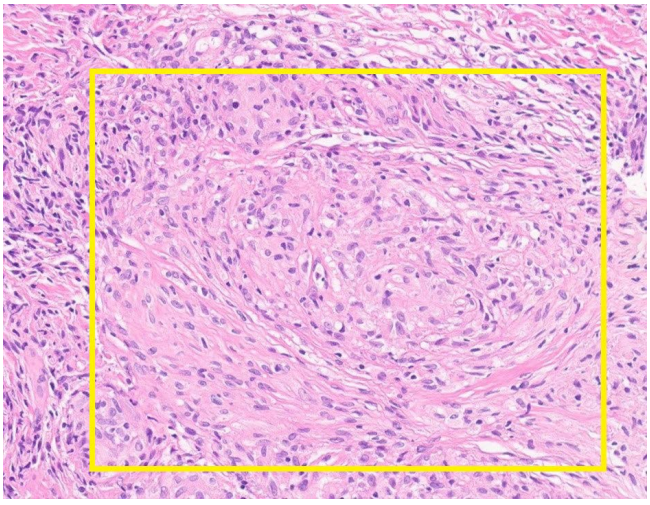

Fig. 4. High-magnification image of the biopsy specimen of a subcutaneous nodule. Histopathological findings showed a characteristic granuloma seen in sarcoidosis (box); a non-caseating granuloma consisted of epithelioid histiocytes admixed with lymphocytes (Hematoxylin and Eosin stain, × 200).

infiltrative lesion in which serpentine vessels interdigitate with fibroadipose tissue. For a peripheral nerve-sheath tumor, such as a schwannoma, a typical fusiform shape, signal intensity, and enhancement pattern are often present. It is isointense compared to skeletal muscle on T1 WI and hyperintense on T2 WI, with various degrees of enhancement (10).

In our patient, the lesions showed iso to slightly higher signal intensity than did muscle on both T1 and T2 WI with an internal reticular pattern. These had irregular shapes, showing marked enhancement with surrounding fat edema. The findings were consistent with the reported findings of subcutaneous sarcoidosis in nodular form. Within the lesions, we recognized none to less-enhancing linear structures, which we interpreted as vascular structures or septa. Although we first suspected pathologies such as Kimura's disease or lymphoma, we then found it to be subcutaneous sarcoidosis.

There is currently no consensus on the treatment for subcutaneous sarcoidosis with lung involvement. Systemic corticosteroid therapy is typically indicated because of extra cutaneous involvement or because of symptomatic or severe lung involvement. For the skin, only severe cutaneous lesions are indicated for systemic corticosteroid therapy (4).

In conclusion, we report on a rare case of subcutaneous sarcoidosis in the lower extremity, involving the lungs at the same time, which makes it almost unique. Therefore, subcutaneous sarcoidosis should be considered in patients with subcutaneous nodules in extremities that present with pulmonary sarcoidosis symptoms, as being as likely as are other soft-tissue masses.

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