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Case Report

Rare Presentations of Sarcoidosis: Cases of Non-Pulmonary Involvement

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Abstract

Sarcoidosis, an inflammatory disease characterized by non-caseating granulomas of unknown etiology, typically manifests with pulmonary involvement. However, presentations without pulmonary manifestations are exceedingly rare. We present 2 cases illustrating unusual presentations of sarcoidosis without pulmonary involvement. A 60-year-old male presented with chronic joint pain and swelling in the left hand. Despite recurrent hospital visits and inconclusive evaluations, granulomatous tenosynovitis was confirmed through histopathological examination. Systemic involvement was ruled out, and the patient responded well to corticosteroid therapy. A 36-year-old female exhibited persistent subcutaneous nodules on the left hand for 6 years. A biopsy confirmed non-caseating granulomas consistent with sarcoidosis. Despite the absence of pulmonary findings, systemic screening revealed no further involvement. Intralesional corticosteroid therapy yielded significant improvement. Although sarcoidosis typically presents with pulmonary involvement, cases without pulmonary manifestations are rare. Our cases highlight the diagnostic challenges and delayed recognition of such presentations. Increased awareness of atypical presentations is crucial for timely diagnosis and management to prevent potential systemic complications.

Keywords: Sarcoidosis, tenosynovitis, nodular sarcoidosis, isolated sarcoidosis

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Introduction

Sarcoidosis is an inflammatory disease of unknown etiology characterized by non-caseating granulomas, with widespread involvement in the body, most commonly affecting the lungs and thoracic lymph nodes.[1,2] The phenotypic and symptomatic manifestations of sarcoidosis can be unpredictable, with pulmonary involvement being the most common presentation. It is often asymptomatic or accompanied by constitutional symptoms or symptoms related to the affected organ.[3-5] In cases where sarcoidosis presents with pulmonary involvement and hilar lymphadenopathy, which are observed in approximately 90% of cases, diagnosis may take longer than 6 months.[6,7] Herein, we present 2 cases of sarcoidosis with various organ involvements, representing a rare presentation without pulmonary involvement.

Case Presentation

Case 1

A 60-year-old male presented with pain, limited mobility, and swelling in the third metacarpophalangeal joint of the left hand. The patient had experienced recurrent hospital visits for joint pain complaints over the past 2 years, without improvement with symptomatic treatment. A recent exacerbation of joint stiffness, swelling, and hypoesthesia prompted re-evaluation. On examination, a palpable, painless, non-warm, and movement-restricting mass was palpated extending from the palm of the left hand to the proximal interphalangeal joint of the third finger. Rheumatological, pulmonary, abdominal, and cardiovascular system examinations revealed no additional pathology. Laboratory tests, including complete blood count, creatinine, liver function tests, coagulation profile, sedimentation rate, and C-reactive protein, were within normal limits. Radiographic imaging of the left hand revealed no bone lesions. Magnetic resonance imaging (MRI) showed hyperintense fluid surrounding the flexor tendon of the third finger consistent with inflammatory tenosynovitis (Figures 1 and 2). A tenosynovectomy was performed, excising a 2 × 2 cm mass for pathological examination, which confirmed granulomatous tenosynovitis (Figure 3). Screening for systemic sarcoidosis was negative, with no evidence of granulomas on computed tomography (CT) imaging (Figure 4). Serum angiotensin-converting enzyme (ACE) level was elevated at 55.37 U/L (normal range:

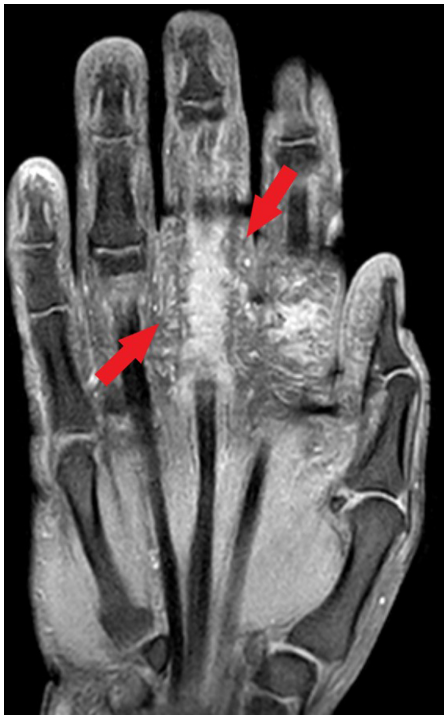

Figure 1. Left middle finger showing marked tendon sheath thickening (Turbo spin echo T1-weighted MRI).

8–52 U/L). The patient was managed with oral methylprednisolone therapy and is being followed up intermittently.

Case 2

A 36-year-old female presented with multiple 3-4 mm nodules on the flexor surface of the first digit of the left hand, persisting for 6 years (Figure 5). She had no known comorbidities or medication use. A biopsy of a nodule revealed non-caseating granulomas consistent with sarcoidosis (Figure 6). Screening for systemic involvement was negative, and chest CT showed no evidence of granulomas (Figure 7). Serum ACE levels were slightly elevated at 42 U/L. The patient experienced a significant reduction in nodule size following intralesional methylprednisolone injection, followed by oral methylprednisolone therapy (Figure 8). Informed consent was obtained from both the patients for publication purposes.

Main Points

- Although sarcoidosis typically affects the lungs, this article presents two rare cases without pulmonary involvement.
- In the first case, a 60-year-old male was diagnosed with granulomatous tenosynovitis in the third metacarpophalangeal joint of the left hand and responded well to corticosteroid therapy.
- In the second case, a 36-year-old female had persistent subcutaneous nodules in her left hand for six years, was diagnosed with sarcoidosis through biopsy, and showed improvement with intralesional corticosteroid treatment.
- Since non-pulmonary forms of sarcoidosis are rare, the diagnostic process may be delayed, leading to potential systemic damage.
- Increasing awareness of non-pulmonary sarcoidosis is crucial for early diagnosis and treatment.

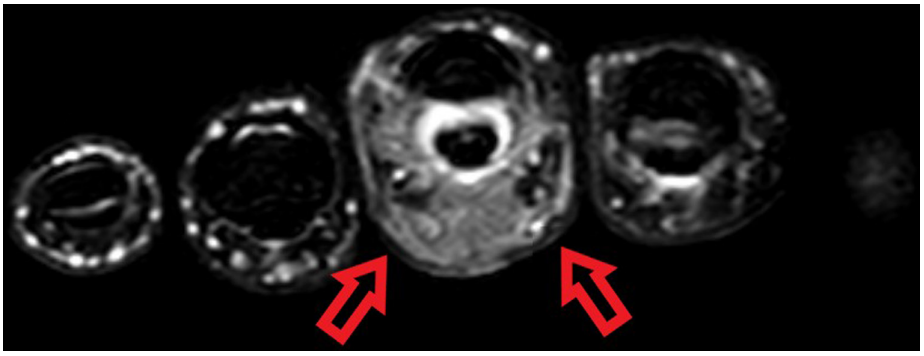

Figure 2. Left middle finger showing marked tendon sheath thickening (axial fat-saturated T2-weighted MRI).

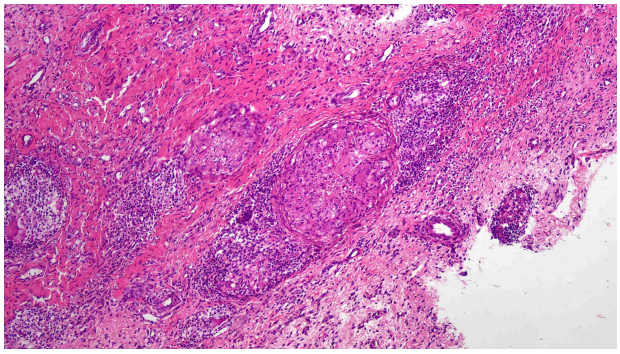

Figure 3. Noncaseating granuloma, operation material (hematoxylin and eosin, ×100).

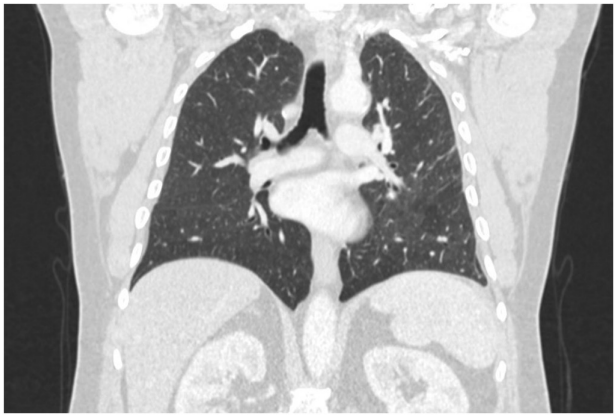

Figure 4. Chest CT shows clear lungs with no evidence of interstitial changes, hilar, or mediastinal lymphadenopathy.

Discussion

Although pulmonary involvement is observed in approximately 90% of sarcoidosis cases, cases without pulmonary involvement, such as those presenting with tenosynovitis and subcutaneous nodules, are exceedingly rare.[6,7] To our knowledge, there are fewer than 5 reported cases of sarcoidosis presenting with tenosynovitis without pulmonary involvement.[8,9] Despite typical symptoms and

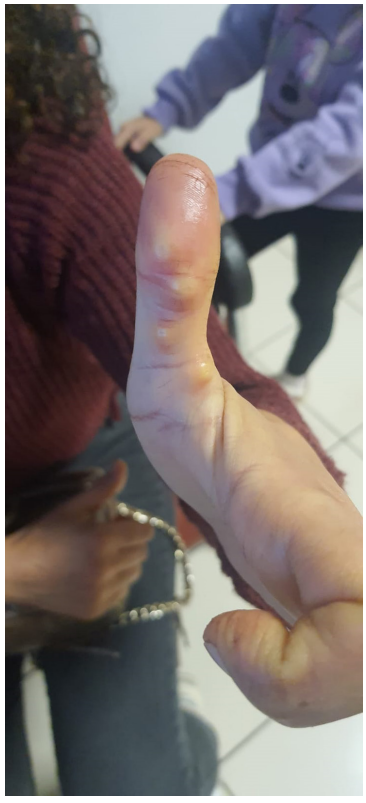

Figure 5. Nodules flexor surface of the first digit of the left hand.

presentations, diagnosis can take longer than 6 months.[6] As illustrated by the cases presented herein, the accurate diagnosis of rare presentations of sarcoidosis may take years. Given the potential for systemic and irreversible damage with delayed diagnosis and treatment, maintaining awareness of sarcoidosis as a differential diagnosis is essential in such cases.

Informed Consent: Informed consent was obtained from the patients who agreed to take part in the study.

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Declaration of Interests: The authors have no conflict of interest to declare.

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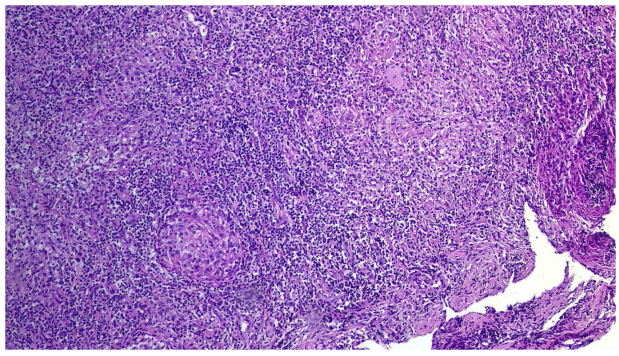

Figure 6. Non-caseating granuloma, nodule biopsy (hematoxylin and eosin, ×100).

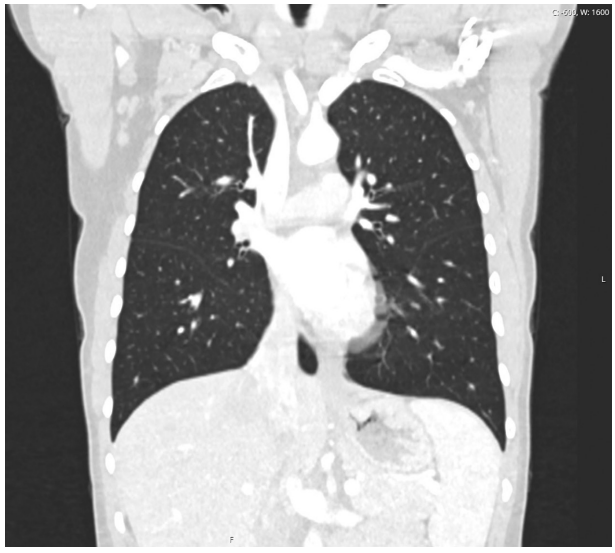

Figure 7. Chest CT shows clear lungs with no evidence of interstitial changes, hilar, or mediastinal lymphadenopathy.

Figure 8. After intralesional methylprednisolone injection, followed by oral methylprednisolone therapy.

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