Nail Sarcoidosis with and without Systemic Involvement: Report of Two Cases

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Established Facts

- Sarcoidosis is a multisystemic disorder characterized by the formation of noncaseating epithelioid granulomas.
- Nail involvement in sarcoidosis is rare.

Novel Insights

- We describe 2 cases of sarcoidosis that developed nail involvement – one with and the other without any evidence of systemic involvement.

Key Words

Nail disorder · Nail pathology · Sarcoidosis

Abstract

The most commonly seen nail change in sarcoidosis, often associated with chronic systemic disease, is onychodystrophy. Imaging of hands, feet and chest is of paramount importance when nail sarcoidosis is suspected, for evaluation of bone cysts and pulmonary involvement, respectively. We report 2 cases of sarcoidosis that developed nail involvement – one with pulmonary disease, and the other without any evidence of systemic involvement, which is very rare.

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Introduction

Sarcoidosis is a multisystemic disorder characterized by the formation of noncaseating epithelioid granulomas that can affect any organ of the body, leading to a wide variety of clinical manifestations [1, 2]. It most commonly involves pulmonary structures, ranging from alveolitis to fibrosis [3].

Nail involvement in sarcoidosis is rare, and the main change found is nail dystrophy [1]. In such cases, an important feature is an association with long-term disease [1, 2].

We report 2 cases of sarcoidosis developing nail involvement – one with pulmonary disease, and the other without any evidence of systemic involvement, which is very rare.

Fig. 1. Nail of the right fourth finger.
Fig. 2. Left fourth finger with onycholysis and subungual hyperkeratosis.

Fig. 3. Histopathology of the nail bed showing a noncaseating epithelioid granuloma in the dermis (HE). **a** Low magnification. **b** High magnification.

Fig. 4. Erythematous and infiltrated papules involving the columella.

Case Reports

Case 1

A 67-year-old Caucasian male patient with an 18-year history of infiltrated lesions located on his ear helix was diagnosed with cutaneous sarcoidosis. Oral prednisone and hydroxychloroquine were introduced with good response, leading to remission after 2 years of treatment. He had been asymptomatic for 2 years when he developed nail dystrophy on his right fourth finger (fig. 1). The results of routine hematological and blood biochemistry tests were all normal. The chest and hand roentgenograms were normal, full body gallium scintigraphy was not characteristic of sarcoidosis, and chest tomography was not suggestive of pulmonary involvement; therefore, systemic sarcoidosis was ruled out. Nail clippings were obtained for mycological examination, with negative results. A shave biopsy of the nail bed was taken, revealing sarcoid granulomas in the dermis and a lymphocytic infiltrate. Some of the granulomas presented central fibrinoid necrosis. Since nail dystrophy was the sole clinical presentation at that time, we opted to use topical occlusive clobetasol, to which the patient showed good response.

Fig. 5. Radiograph of the left hand: bone cysts of the fourth distal phalanx.

Case 2

A 58-year-old African-descendent woman with a 4-month history of pain and swelling of her left fourth finger presented with onycholysis and subungual hyperkeratosis (fig. 2). No periungual edema was observed. Nail clippings for mycological examination had negative results. A punch biopsy taken from the nail bed showed sarcoid granulomas in the dermis and a chronic inflammatory infiltrate (fig. 3a, b). In face of the nail sarcoidosis diagnosis, a full-body dermatological examination was performed, revealing few infiltrated papules on the nose columella (fig. 4) and erythematous annular macules with a hypochromic halo and slight desquamation located on her arms and breasts. The cutaneous lesions also showed histopathological findings of sarcoidosis. A left hand X-ray demonstrated bone cysts of the fourth distal phalanx (fig. 5). Despite the absence of respiratory symptoms, a chest X-ray and a chest CT were performed, which showed bilateral hilar lymphadenopathy. Routine blood tests were all normal. After 6 weeks of treatment, the patient had a good response to 1-gram methylprednisolone pulse therapy administered weekly, but had to discontinue treatment due to adverse events.

Discussion

Skin involvement occurs in as many as one third of all patients with sarcoidosis and is usually an early manifestation of the disease. In contrast, nail changes are more unusual, ranging from 1 in 400 to 3 in 188 cases, and they are often associated with chronic systemic disease [1, 2, 4, 5].

The most commonly seen nail change in sarcoidosis is onychodystrophy, but other findings may include nail thickening, longitudinal ridging, discoloration, splinter hemorrhages, onychorrhexis, onycholysis, pterygium, subungual hyperkeratosis, lamellar splitting, clubbing, nail pitting, cracking, and brittleness of the nails [1, 2].

A review of 33 cases described in the literature revealed a greater association of nail involvement with digital bone cysts, dactylitis, and lupus pernio. In this review, all cases were associated with systemic or cutaneous involvement. Systemic involvement includes, in decreasing order of prevalence, lymphadenopathy (especially hilar), pulmonary fibrosis, arthritis, sinusitis, and splenomegaly [1]. About 79% of the patients had skin lesions, with 36% exhibiting lupus pernio and 6% having erythema nodosum [1]. These two types of lesions may have prognostic significance – the former being related to chronic systemic involvement and the latter being related to acute and benign disease [6].

Commonly, the diagnosis of nail sarcoidosis is delayed due to its clinical similarity to other prevalent diseases such as onychomycosis, psoriasis, lichen planus, trauma, drug eruption, and subungual wart [7, 8]. Nail thickening, discoloration, onycholysis, and onychodystrophy may be present in onychomycosis [9]; discoloration, onycholysis, splinter hemorrhages, and onychodystrophy may be present in trauma [10]; onycholysis, subungual hyperkeratosis, and nail pitting are typically present in psoriasis [11], and the pterygium classically occurs in lichen planus, although this is usually associated with skin or mucosal lesions [12]. A detailed clinical history and a thorough dermatologic examination drive the diagnostic investigation, as it may reveal systemic symptoms or demonstrate characteristic skin lesions of sarcoidosis [1, 6]. The histopathological study of the nail bed – or eventually of the nail fold – makes the diagnosis clear by demonstrating noncaseating epithelioid granulomas [7, 13]. Losada-Campa et al. [7] propose an interesting theory related to the pathogenesis of nail changes. They claim that there is a strong correlation of such changes with the compressive effects created by sarcoid granulomas between the phalanx and the nail plate.

Treatment options for nail dystrophy in sarcoidosis include systemic treatment with oral prednisone, hydroxychloroquine sulfate, high-potency topical steroids, and steroid injections into the nail fold [13].

In spite of being rare, nail changes in sarcoidosis are usually associated with the systemic form of the disease [2]; therefore, imaging of hands, feet and chest are of paramount importance when nail sarcoidosis is suspect-

ed [14]. The first patient in this report shows a very uncommon manifestation, where cutaneous lesions are exclusive. Even with negative tests for systemic sarcoidosis, the patient has been monitored for lesions in other organs.

We conclude that sarcoidosis should be considered a differential diagnosis of nail lesions, allowing an early diagnosis of systemic sarcoidosis.

Statement of Ethics

Both patients were informed and gave informed consent to the publication of this paper.

Disclosure Statement

There is no conflict of interest.

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