

A CASE OF NAIL SARCOIDOSIS WITH RICH CLINICAL FINDINGS

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To the editor,

As skin lesions are observed in 10-35% of sarcoidosis patients, nail involvement is very rare which generally develops in the setting of chronic disease (1-4). Nail dystrophy is the most frequent nail change among several described findings (1,2,4-7). Herein, we report a case of sarcoidosis showing rich nail findings some of them rarely reported or not well-described.

A 36-year-old man with pulmonary sarcoidosis (stage one) (Figure 1a,b) presented with multiple subcutaneous nodules on the face and extremities (Figure 2a). Histopathologic examination confirmed cutaneous sarcoidosis. Median splitting was noticed on one fingernail plate accompanied by a subungual reddish nodule and increased convexity on the proximal portion of the nail plate (Figure 2b). Magnetic resonance imaging of hand demonstrated soft tissue expansion at the location with increased convexity. In addition, bone cysts were detected in adjacent thumb and middle fingers radiologically (Figure 3a,b). Systemic methylprednisolone therapy resulted in significant improvement of the nail changes after four months of treatment (Figure 2c), along with the skin lesions that resolved completely. However, two months after cessation of the therapy, skin lesions recurred and distal notching associated with onycholysis and increased

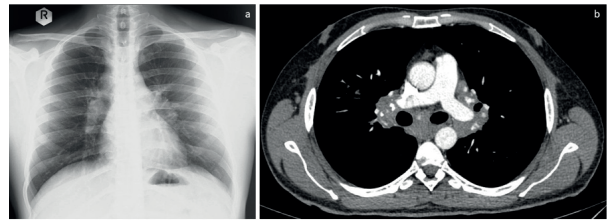


FIGURE 1. (A) Symmetrical hilar lymph nodes are observed on chest radiograph, (B) Axial computed tomography section of thorax shows mediastinal and hilar lymph nodes with punctate and amorphous calcifications.

convexity of the nail plate appeared (Figure 2d). All lesions regressed after readministration of systemic corticosteroid (Figure 2e).

Three years later, he was admitted with ulcerated plaques and subcutaneous nodules on arms (Figure 4a) and severe nail involvement (Figure 4b). Increased convexity, longitudinal ridging and subungual hyperkeratosis were observed on the previously affected third digit of the right hand (Figure 2f), while a subungual reddish hyperkeratotic nodule on the thumb-nail was detected which was associated with partial (median) nail plate loss (Figure 4c). Pitting, mild subungual hyperkeratosis, erythronychia and onycholysis were present on the third digit of the left hand (Figure 4d). Furthermore, total dystrophy (opaque, fragile, irregularly thickened nail plate) and subungual hyperkeratosis were observed on both big toenails accompanied by red-violaceous periungual discoloration (paronychia involvement) and tiny papules evaluated as lupus pernio of digit (Figure 4b,e). All lesions including periungual erythema, papules and nail findings regressed dramatically (Figure 4f) after moderate-dose systemic corticosteroid.

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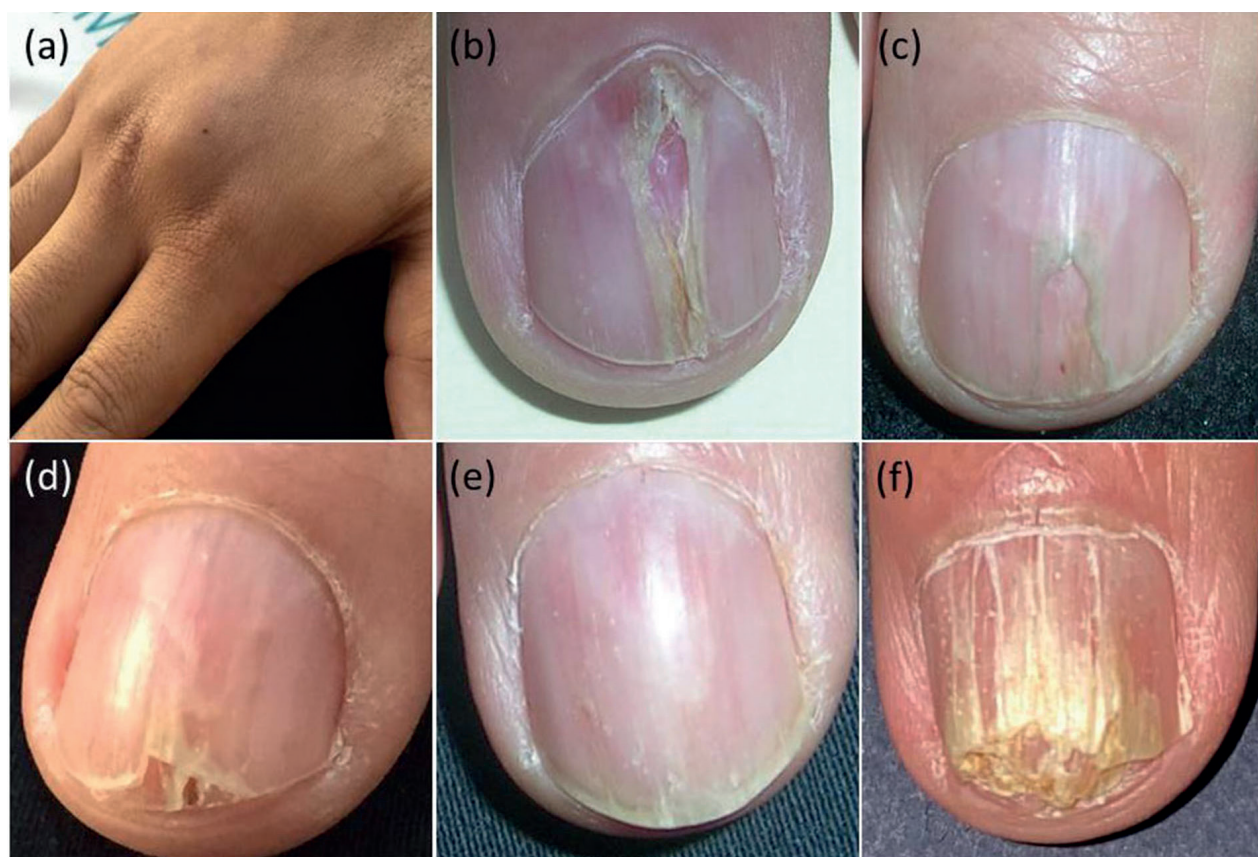


FIGURE 2. (A) A subcutaneous nodule on the dorsum of the right hand. (B) A subungual smooth-surfaced, ill-defined reddish nodule on the proximal portion of the nail bed associated with leukonychia, increased convexity and median splitting in the nail plate of the right third finger. (C) Regression of the nail findings resulted in superficial nail plate loss on the distal portion of the nail after four months of systemic corticosteroid therapy (initial dose of 24 mg/day). (D) Relapse on the same nail as distal notching accompanied by onycholysis and increased nail plate convexity occurred two months after treatment cessation. (E) Totally healed nail except leukonychia following a second course of systemic corticosteroid therapy (initial dose of 24 mg/day). (F) Longitudinal ridging and subungual hyperkeratosis on the distal part of the same finger nail appeared after three years remission period in association with the involvement of four other nails in Figure 4.

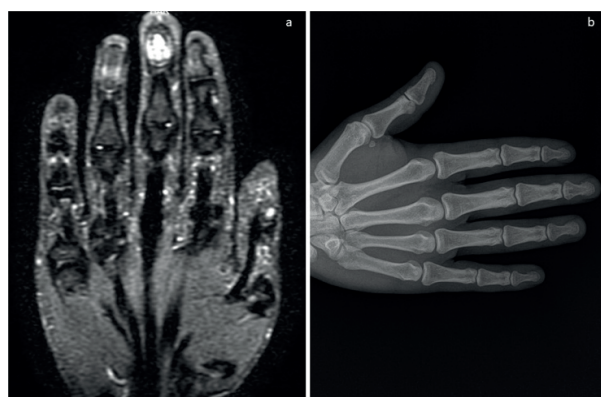


FIGURE 3. (A) T2-weighted coronal magnetic resonance image shows well-circumscribed cystic bone lesion in the distal phalanx of the third finger and on the thumb. (B) Radiograph of the hand shows minimal cystic and lytic destruction of phalanges of the fingers.

Nail involvement in sarcoidosis has a prevalence ranging from 0.3% to 1.6% in various antecedent series (3). Information concerning the clinical features of the disease is still based on case reports. To the best of our knowledge, only 10 sarcoidosis cases with nail findings have been reported from 2011 to 2021 (1-3,5,6,8-12) following a review article including formerly reported 33 cases (4) (Table 1). Nail dystrophy (30%), longitudinal ridging (24%), onycholysis (21%), subungual hyperkeratosis (21%) and nail hyperkeratosis (6%) were the leading features in this article (4). A few new clinical presentations including longitudinal erythronychia (3) were also described thereafter (Table 1) (1-3,5,6,8-12). In our patient, we have encountered the well-known features of nail sarcoidosis in



FIGURE 4. (A) A violaceous infiltrated plaque with superficial ulceration on the left upper arm. (B) Nail involvement of three fingers and two big toes associated with periungual skin changes of big toes. (C) Subungual reddish hyperkeratotic nodule on the right thumbnail with partial nail dystrophy and median nail plate loss. (D) The third fingernail of the left hand showing pitting, onycholysis, erythronychia, leukonychia and mild subungual hyperkeratosis on the distal part of the nail. (E) Total dystrophy and pronounced subungual hyperkeratosis on the right big toenail with red-violaceous periungual discoloration and multiple reddish tiny papules on the digit pulp. (F) Regression of periungual involvement, nail dystrophy of the right big toenail resulted in nail plate loss and mild subungual hyperkeratosis in 2.5 months after administration of systemic corticosteroid therapy (initial dose of 40 mg/day).

addition to peculiar findings such as splitting on the median of nail plate and subungual reddish nodules which occurred at different stages of the disease. Although longitudinal splitting was formerly described (7), “median splitting” may be a more convenient term due to midline location as seen in our case.

As nail findings were more frequently associated with lupus pernio typically located on the face, our case showed subcutaneous nodules, plaques on various parts of the body and lupus pernio on digits toes (4). Especially bone involvement on terminal phalanges was incidentally detected in sarcoidosis patients presenting with nail dystrophy as in our case (4). Rich nail findings are possibly related to granulomatous infiltration on the nail matrix or nail bed. The micro-

compressive effect of microgranulomas between the nail plate and phalanx was held responsible for nail dystrophy (13). Santoro *et al* reported a case presenting with subungual hyperkeratotic papules associated with longitudinal splitting and distal notching (7). In addition to hyperkeratotic subungual nodule, the reddish nodular lesion in the same area noted in our case was quite unique, with smoother surface. These nodules presenting on the nail bed might be accepted as macroscopic counterparts of the aforementioned granulomatous lesions causing secondary changes in the nail plate, such as median splitting by means of direct mechanical pressure. Longitudinal ridging, distal notching, onycholysis and partial nail plate loss were other features seen on the fingernails of our patient.

Paronychia involvement was rarely observed in sarcoidosis (4,9,10,12). In our case, both big toes with periungual involvement including nail matrix area showed total nail dystrophy.

Main therapeutic options of nail sarcoidosis are topical, intralesional or systemic corticosteroids, doxycycline and methotrexate (Table 1) (1-3,5,6,8-12). In our case, systemic corticosteroid therapy primarily

TABLE 1. Patients with nail involvement of sarcoidosis reported in the literature and present case

| AUTHOR / YEAR | AGE (YEARS) / GENDER | NAIL FINDINGS / AFFECTED NAILS | SYSTEMIC INVOLVEMENT | TREATMENT RESPONSE OF NAIL INVOLVEMENT |
|--------------------------|--|--|--|--|
| Momen*† (4) / 2013 | 24-80, not specified in 8 patients / 13 females, 9 males, not specified in 11 patients | Red-purple periungual discoloration, painful and swollen nail fold, infiltration of proximal folds, erythematous subungual hyperkeratotic papules, atrophic matrix, trachyonychia, fragility, longitudinal stripes/ridges/cracking/grooves, splitting, pits, subungual hyperkeratosis, hyperkeratosis of nail, dystrophy, nail destruction, thickening, onycholysis, splinter haemorrhages, pterygium, lateral deviation, discoloration, leukonychia, hyperpigmentation, erosion of free edges, ulceration, complete nail loss / Some or all of fingernails/toenails | Pulmonary (n=15), upper airway (n=3), bone (n=17), cutaneous (n=26), dactylitis (n=11) | Not specified in detail |
| Rajan (1) / 2014 | 44 / female | Purplish soft tissue swelling of the digit pulp beneath the free edge of nails / Big toes | Bone | Intradermal corticosteroid injections/ Regression |
| Kawaguchi (5) / 2014 | 51 / female | Longitudinal ridging, thickening, fragility / One big toenail | Pulmonary | High-potency topical corticosteroids/ No response |
| Bekkali (6) / 2014 | 29 / female | Subungual hyperkeratotic verrucous lesion / One toenail | Pulmonary, bone, cutaneous | High-potency topical corticosteroids/ Regression |
| Noriega (2) / 2015 | 67 / male | Nail dystrophy / One fingernail | Cutaneous | High-potency topical corticosteroids/ Regression |
| | 58 / female | Onycholysis, subungual hyperkeratosis / One fingernail | Pulmonary, bone, cutaneous | Systemic pulse corticosteroid/ Regression |
| Albers (8) / 2016 | 59 / female | Periungual erythema, onychoschizia, onycholysis, cuticular loss / Nine fingernails sparing only one thumbnail | Pulmonary, bone | Doxycycline/ Regression |
| van Lümmig (3) / 2018 | 37 / female | Subungual erythematous macules, longitudinal erythronychia, onychorrhexis / Multiple toenails including big toe (number not stated) | Pulmonary, upper airway | High-potency topical corticosteroids/ No response Intradermal corticosteroid injections/ Regression |
| Moulouguet (9,10) / 2018 | 40 / male | Violaceous paronychia, pterygium, nail atrophy / Thumb and one other fingernail | Pulmonary, bone, dactylitis | Systemic corticosteroid/ Regression |
| Cartayrade (11) / 2019 | 28 / female | Nail splitting / One fingernail | Bone, cutaneous, dactylitis | Systemic corticosteroid and methotrexate/ Not stated |
| Maouni (12) / 2019 | 57 / female | Paronychia, nail hyperkeratosis and longitudinal ulcero-verrucous lesions on nail plate / Four toenails including one big toe | Pulmonary, upper airway, bone, cutaneous, dactylitis | Systemic corticosteroid/ Regression |
| Present case / 2022 | 36 / male | Periungual erythema and papules (lupus pernio), subungual reddish (smooth-surfaced or hyperkeratotic) nodule, subungual hyperkeratosis, nail dystrophy, pitting, onycholysis, distal notching, median splitting, longitudinal ridging, leukonychia, erythronychia, increased nail plate convexity, nail plate loss (median and total) / Third fingernails of both hands, right thumbnail and both big toes | Pulmonary, bone, cutaneous | Systemic corticosteroid (on each exacerbation)/ Regression (three times on one fingernail) |

*A review article including 33 patients with nail involvement of sarcoidosis reported between 1981 and 2010

†Main systemic involvements of sarcoidosis mentioned in the review article were noted in this table.

indicated for multiple cutaneous lesions showed dramatic effect for all types of nail lesions. However, the lesion on one fingernail showed two recurrences.

In conclusion, our case indicates the heterogeneity of nail sarcoidosis related with location of granulomas. Median splitting and smooth-surfaced subungual nodule seen in our case are previously not well described findings. Even severe nail involvement responds well to systemic corticosteroids but recurrence on the same nails is possible.

CONFLICTS OF INTEREST: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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