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Osseous sarcoidosis with Fingers distal phalanx autoamputation in a treatment-naïve case

Sarcoïdose osseuse avec autoamputation de la phalange distale des doigts chez un patient naïf de traitement

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ABSTRACT

We report a case of a 55-year-old female, with 3 years history of a treatment-naive sarcoidosis involving lungs with second-stage sarcoidosis, skin in form of lupus pernio, and bones with multiple cystic areas of the digital bones, soft tissue swelling, nails dystrophy and missed phalanges. Uveitis frequently associated with bone sarcoidosis, was absent in our patient. Prior to attend to our clinic, she had been treated with short courses of nonsteroidal anti-inflammatory drugs and analgesics. No long-term corticosteroids therapy had been reported. Osseous lesions led to autoamputation of distal phalanges of two fingers. It is a very rare complication of unclear mechanism; however, they have been attributed by Several authors to long-term corticosteroid therapy, hypothesis not supported by our findings. Systemic corticosteroids are considered as standard medication. There are no firm guidelines for dosage and period of the treatment. Steroid-sparing agents are often added mainly methotrexate, in some patients, biologics can be considered. Unfortunately for our patient there was not enough time for management as she died lately.

Keywords: autoamputation; hand sarcoidosis; Corticosteroids; treatment naïve.

RESUME

Nous rapportons le cas d'une femme de 55 ans, avec 3 ans d'antécédents de sarcoïdose naïve de traitement atteignant les poumons avec une sarcoïdose de deuxième stade, une atteinte de la peau sous la forme de lupus pernio et une atteinte osseuse avec de multiples zones kystiques des os digitaux, un gonflement des tissus, une dystrophie des ongles et des phalanges manquées. L'uvéite fréquemment associée à une sarcoïdose osseuse était absente chez notre patient. Avant de se présenter à notre clinique, elle avait été traitée avec des traitements de courte durée (anti-inflammatoires non stéroïdiens et d'antalgiques). Aucune corticothérapie au long cours n'a été rapportée. Les lésions osseuses ont conduit à l'autoamputation des phalanges distales de deux doigts. C'est une complication très rare de mécanisme peu clair ; cependant, ils ont été attribués par plusieurs auteurs à une corticothérapie au long cours, hypothèse non étayée par nos résultats. Les corticoïdes systémiques sont considérés comme des médicaments de référence. Il n'y a pas de lignes directrices fermes concernant la posologie et la durée du traitement. Des agents épargneurs de stéroïdes sont souvent ajoutés principalement du méthotrexate, chez certains patients, des produits biologiques peuvent être envisagés. Malheureusement pour notre patiente, il n'y avait pas assez de temps pour la prise en charge car elle est décédée au début du suivi.

Mots-clés : autoamputation ; sarcoïdose de la main ; corticoïdes ; naïf de traitement.

INTRODUCTION

Sarcoidosis is a multi-system disease of unknown aetiology that predominantly affects the lungs and intrathoracic lymph nodes. In approximately 5% of patients, sarcoidosis involves the bones [1].

Dactylitis is one of the most familiar appearances of musculoskeletal involvement in sarcoidosis. Most clinicians are familiar with this pattern of disease, which is almost exclusively associated with chronic systemic involvement. It has been described in coexistence with lupus pernio and other forms of chronic cutaneous sarcoidosis [2]. It typically develops in a symmetrical pattern, most often affecting the second and third phalanges, preserving the metacarpo-phalangeal (MCP) joints [3]. Dactylitis is associated with swelling and erythema and is similar in appearance to psoriatic arthritis [4]. It is more common in those of African descent. Radiological findings most frequently show cystic lesions, punched-out lesions, osteolysis, reticularization of cortical bone or cortical defects and, rarely, sclerotic, periostitis or destructive lesions [5].

There are three radiological types of sarcoid bone disease: type I, characterized by big cystic lesions, which is quite rare and can be associated with a stress fracture from a pathological fracture; type II, characterized by multiple small, circumscribed cysts, occasionally conflicting, and polycyclic; and type III, characterized by tunnelling of the cortex of the phalanx, which leads to

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remodelling of the cortical and trabecular architecture. All three forms can coexist in the same bone. Acro-osteolysis, presenting as nodular densities in the terminal phalanges, can also occur [6].

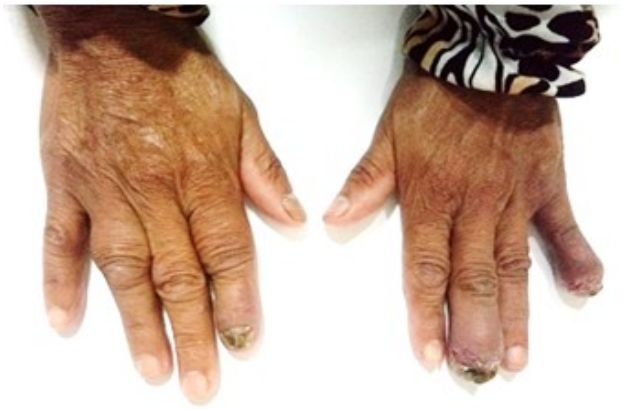

Figure 1. Both hands overview

CASE REPORT

A 55-year-old housewife, without a previous medical history, non-smoker, presented with a 3-year history of pain, swelling and severe disability of both hands. Clinical examination revealed a swollen proximal interphalangeal (PIP) joint of the right fourth finger, nail dystrophy of right second finger, tenderness and swelling of the right wrist with restricted range of motion. The left hand showed third and fifth finger shortening deformities with dysmorphic nails (Figure 1), left middle and little fingers acquired shortening deformity and right wrist swelling getting worse over time. Her skin examination showed pinkish papules and nodules of the apex and rims of the alae of her nose, with scattered lesions on her lips and right malar cheek area (Figure 2).

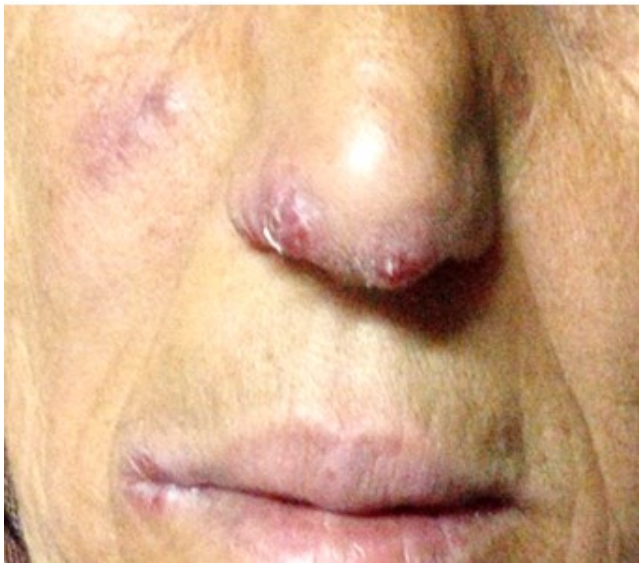

Figure 2. Face cutaneous sarcoidosis

She denied cough, fever, shortness of breath and weight loss. There was no history of trauma or injury. Lab tests revealed a normal peripheral blood count, positive C reactive protein levels and a high erythrocyte sedimentation rate (57 mm).

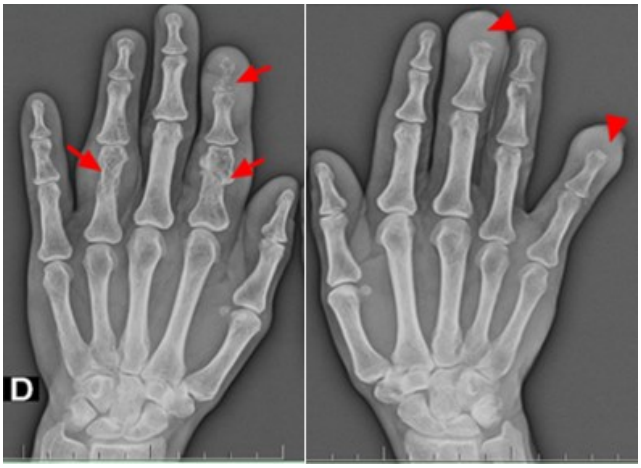

Figure 3. X-ray film exhibiting multiple cystic areas of the digital bones(arrow), soft tissue swelling and missed phalanges (arrowhead).

Blood chemistry was normal (including calcium and alkaline phosphatase levels) and the serology revealed a negative rheumatoid factor and anti-cyclic citrullinated peptide antibodies. PPD test, the serum angiotensin converting enzyme level were not evaluated. Chest X-ray revealed bilateral reticulonodular opacities of the mid and lower zones of the lungs and hilar lymphadenopathy, consistent with second-stage sarcoidosis. Both hands plain X-ray showed multiple cystic areas of the digital bones, soft tissue swelling and missed phalanges (Figure3). The patient was treated symptomatically, with non-steroidal anti-inflammatory drugs and topical antibiotics. Unfortunately, there was not enough time for specific management as she died lately.

DISCUSSION

The first case of bone sarcoidosis was reported by Besnier in 1898 in a patient with lupus pernio. The characteristic bone involvement in sarcoidosis as 'osteitis tuberculosa multiplex cystic' was described by Jungling in 1928 [7]. Jaffe in 1972 was the first to report bone sarcoidosis with distal phalanx autoamputation. To our knowledge this complication is still very rare, few cases only were found on literature.Due to the absence of prospective trials, based on the retrospective data, bone involvement was detected in 1–15% of the patients with sarcoidosis. Patients with bone sarcoidosis have other organ involvement, most commonly pulmonary (80–90%), lupus pernio (48–70%), and uveitis (30–50%). The risk of bone sarcoidosis was shown to be 21-fold higher in patients with lupus pernio [8], similar to our patient who conversely didn't present ocular involvement.In rare cases, mutilation and autoamputation of fingers, due to destruction of the terminal phalanges, has been reported [8]. Cetinkaya and co-workers [9] in 2006 wondered whether sarcoidosis can cause fingers' autoamputation; their assumptions seem to be realistic, confirmed by our findings. The reasons for the lesions of sarcoidosis developing and their particular location are unknown. Many theories have been suggested to explain bone involvement. One of them is the high levels of 1,25(OH)2 D3 and a rise in osteoclastic activity, leading to bone resorption. Furthermore, it is possible that granulomas can induce local osteoclastic reaction. Sarcoid granulomas can be a source of osteoclastic factor (9). Several authors have suggested the role of long-term systemic corticosteroids in the genesis of osteolysis; in contrast, our findings do not support what was thought previously regarding their role. Many bone disorders can mimic osseous sarcoidosis. Differential diagnosis includes

neoplastic lesions: enchondroma, chondrosarcoma, metastases especially bronchial carcinoma, renal, breast and prostate, epidermal inclusion cyst. Non neoplastic lesions include Infection (Osteomyelitis, leprosy), osteoarthritis, metabolic (hyperparathyroidism, renal osteodystrophy, gout), autoimmune (scleroderma) conditions…… etc.Due to the fact that it can mimic rheumatologic diseases, delayed diagnosis and/or misdiagnosis can occur. Therefore, in patients presenting to a rheumatologist with musculoskeletal system findings, it should definitely be considered in the differential diagnosis. Sarcoidosis should not be considered only as a mimicking but also a Th1-mediated, primary rheumatologic pathology [8].Treatment of skeletal sarcoidosis is not standardized due to a lack of guidelines from the literature and due to variability in clinical course, including reported cases of improvement with no treatment at all [10]. In general, it is estimated that between 20% and 70% of all patients need systemic therapy. Despite lack of symptoms directly associated with bone involvement, most of the patients should be treated. Similarly, all symptomatic osseous involvements are indications for specific treatment even when the patient has no other manifestations [11]. Systemic corticosteroids are considered as standard medication. There are no firm guidelines for dosage and period of the treatment. In most cases, the initial dose of 20–40 mg of prednisone daily is followed after 6–12 weeks with dose reduction [12]. Higher doses are recommended in some life-threatening situations. Steroid-sparing agents are often added, mainly methotrexate, but also leflunomide, azathioprine, Antimalarial drugs. In some patients, cyclophosphamide, mycophenolate mofetil and biologics can be considered. Unfortunately for our patient there wasn't enough time for management as she died lately.

DECLARATION D'INTERETS

Les auteurs ne déclarent pas de conflits d'intérêts en rapport avec cet article.

Patient consent: Has been obtained orally and detail has been removed from this case description to ensure anonymity.

REFERENCES

1. Siltzbach LE, James DG, Neville E, Turiaf J, Battesti JP, Sharma OP, et al. Course and prognosis of sarcoidosis around the world. Am J Med [Internet]. 1974;57(6):847–52. Available from: http://dx.doi.org/10.1016/0002-9343(74)90160-0
2. Bechman K, Christidis D, Walsh S, Birring SS, Galloway J. A review of the musculoskeletal manifestations of sarcoidosis. Rheumatology (Oxford) [Internet]. 2018;57(5):777–83. Available from: http://dx.doi.org/10.1093/rheumatology/kex317
3. Awada H, Abi-Karam G, Fayad F. Musculoskeletal and other extrapulmonary disorders in sarcoidosis. Best Pract Res Clin Rheumatol [Internet]. 2003;17(6):971–87. Available from: http://dx.doi.org/10.1016/j.berh.2003.09.005
4. Rothschild BM, Pingitore C, Eaton M. Dactylitis: implications for clinical practice. Semin Arthritis Rheum [Internet]. 1998;28(1):41–7. Available from: http://dx.doi.org/10.1016/s0049-0172(98)80027-9
5. Jansen T, Geusens P. Sarcoidosis: joint, muscle and bone involvement. Eur Respir Monogr. 2005;32(10).
6. Nessrine A, Zahra AF, Taoufik H. Musculoskeletal involvement in sarcoidosis. J Bras Pneumol [Internet]. 2014;40(2):175–82. Available from: http://dx.doi.org/10.1590/s1806-37132014000200012
7. Jungling O. Uber ostitistuberculosa multiplex cystoides. Bruns KlinChirurg. 1928;143(401).
8. Kobak S. Sarcoidosis: a rheumatologist's perspective. Ther Adv Musculoskelet Dis [Internet]. 2015;7(5):196–205. Available from: http://dx.doi.org/10.1177/1759720X15591310
9. Cetinkaya R, Kavak A, Parlak AH, Tuzuner T, Arbak PM, Alper M. Can sarcoidosis cause autoamputation of a finger phalanx? J Hand Surg Br [Internet]. 2006;31(4):413–5. Available from: http://dx.doi.org/10.1016/j.jhsb.2006.03.161
10. Johnson AK, Johnson JM, Ames E, Filippi C. Spontaneous clinical and radiological resolution of vertebral sarcoidosis: a case report: A case report. Spine. 2012;37:E414-416.
11. Kucharz EJ. Osseous manifestations of sarcoidosis. Reumatologia [Internet]. 2020;58(2):93–100. Available from: http://dx.doi.org/10.5114/reum.2020.95363
12. Gunasekaran K, Ahmad MR, Dalal B, Edmonds L. Resolution of osseous sarcoidosis with steroids. BMJ Case Rep [Internet]. 2018;bcr-2017-224064. Available from: http://dx.doi.org/10.1136/bcr-2017-224064

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