CASE REPORT

Dactylitis and bone lesions at the onset of sarcoidosis: a case report

Daniela Fodor[1], Laura Poanta[2], Liliana Rogojan[2]

1 2nd Internal Medicine Clinic, University of Medicine and Pharmacy, Cluj-Napoca, Romania
2 Histopathology Department, University of Medicine and Pharmacy, Cluj-Napoca, Romania

KEY WORDS

bone cysts, dactylitis, sarcoidosis

ABSTRACT

Dactylitis and bone lesions are rare complications of sarcoidosis that occur in the chronic disease and they are unusual features of the disease at presentation. The present paper describes a case of a 28-year-old woman with dactylitis (due to tenosynovitis and soft tissue granulomas) and phalangeal bone lesions in 2 fingers at the onset of sarcoidosis. She also had asymptomatic pulmonary type I sarcoidosis (bihilar lymph node enlargement with no involvement of the lung parenchyma). The response to treatment (prednisone 30 mg/day, tapered to 5 mg over 2 months for a 12-month period) was very good, with no relapse at 6 months after the end of systemic treatment.

INTRODUCTION Sarcoidosis is an inflammatory disorder of unknown cause, characterized by the presence of non-caseating granulomas in the tissues. The disease affects multiple organs, most commonly the lung, the lymph nodes, the skin and the eyes, but any other organ involvement is possible. Skeletal involvement has been reported in 1–13% of patients with sarcoidosis, with an estimated average of 5%. The musculoskeletal involvement in sarcoidosis usually occurs in patients with a generalized (systemic) disease and it is believed to be a sign of a chronic and prolonged clinical course.[1] While bone lesions of the phalanges are more common, dactylitis is a rare rheumatologic complication of sarcoidosis, and an association of sarcoidosis of soft tissues and bones of the hand is infrequent.

We present a case of sarcoidosis with soft tissue manifestations (dactylitis due to tenosynovitis and soft tissue granulomas) and phalangeal bone lesions that appeared as early as at the onset of the disease.

CASE REPORT The patient, a 28-year-old Caucasian woman, working as a PC operator, without a previous medical history, nonsmoker, presented with a 2-month history of pain, swelling and severe disability of the right index and left thumb, with progressive worsening and no response to non-steroid anti-inflammatory treatment. Prior to the onset she had never had symptoms about the hands or other joints. She had no constitutional symptoms, like cough, fever, shortness of breath, and weight loss.

The clinical examination revealed the swelling of the right index, from metacarpophalangeal joint to the finger tip on the flexor part and along the second phalanx on the extensor part. At the flexor side of the left thumb there was a swelling distal to the metacarpophalangeal joint. The involved areas were slightly warm and tender and there was an important restriction of the joint motion in the affected joints (FIGURE 1). The findings of the rest of clinical examinations (the remaining musculoskeletal system, the respiratory and the cardiac systems, the abdomen and the skin) were normal.

The antero-posterior and lateral radiographs of the patient's hands showed the presence of cysts in the thumb's 1st phalanx head and the index's 1st and 2nd phalanx head, with no significant cortical disruption, a minor narrowing of the proximal interphalangeal spaces of both fingers and the swelling of soft tissue (FIGURE 2).

The ultrasonography showed severe tenosynovitis of the flexor sheath of both fingers, with hypervascularisation in, and mostly around, the tendon, and a clear definition of the tendon sheath

Correspondence to:
Assistant Professor Laura Poanta, MD, PhD, II Internal Medicine Clinic, University of Medicine and Pharmacy, "Iuliu Hatieganu", 2–4 Clinicilor Street, 400006, Cluj-Napoca, Romania,
phone: +40-744-894-190,
fax: +40-264-596-912,
e-mail: laurapoanta@yahoo.com
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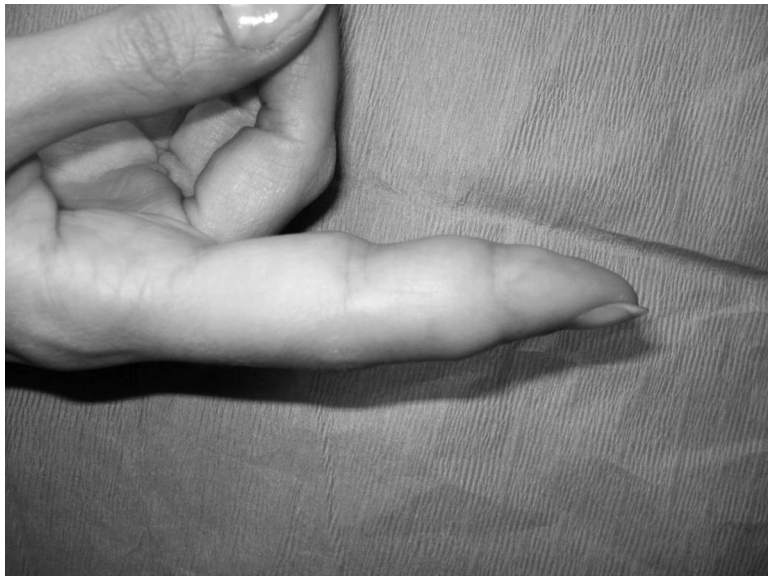

FIGURE 1 Clinical aspect of the right index

from the surroundings tissues. At the extensor part of the index phalanx a hypoechoic mass situated between the bone and the extensor tendon was identified. Some irregularities of bone contours were present, but with no fluid or synovitis inside the joints.

Laboratory tests revealed a normal peripheral blood count, C-reactive protein and serum fibrinogen levels and a normal erythrocyte sedimentation rate. Blood chemistry was normal (including calcium and alkaline phosphatase levels) and the serology revealed a negative rheumatoid factor and anti-cyclic citrullinated peptide antibodies. The tuberculin skin test, the serum angiotensin-converting enzyme level and the human leukocyte antigens type were not evaluated.

A surgical biopsy from the extensor part of the index was performed. The pathologic tissue had a yellowish appearance with a good delineation from the skin, the extensor tendon and the bone. Histopathological findings were of non-caseating granuloma composed of epithelial cells, lymphocyte and occasional Langerhans giant cells.

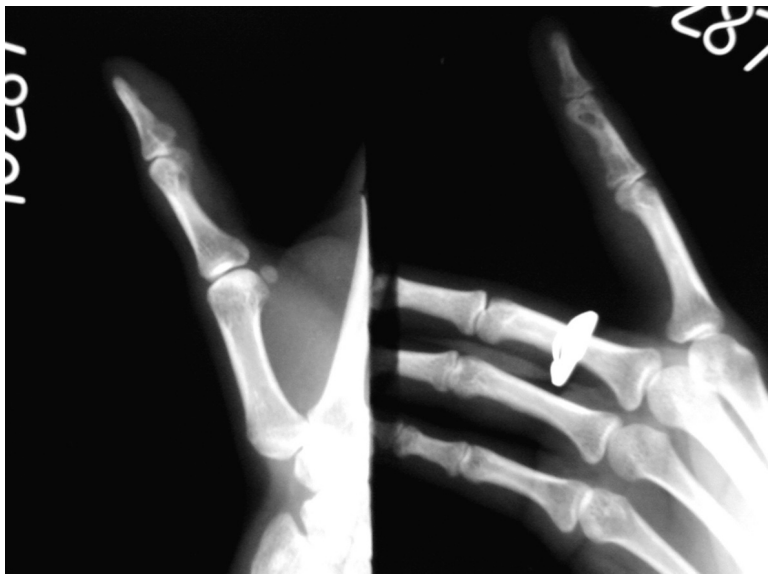

FIGURE 2 Radiography of the left thumb and right index. Bone cysts, slightly narrowing of proximal interphalangeal joints spaces, soft-tissue tumefaction are seen

The pulmonary radiography showed bihilar lymph nodes enlargement with no radiological involvement of the lung parenchyma, interpreted as type I sarcoidosis. The absence of pulmonary parenchyma involvement was confirmed on the computed tomography examination.

The patient received systemic corticosteroids, prednisone 30 mg/day, tapered to 5 mg over 2 months for a 12-month period. She was called for a check-up on every 3 months. The fingers returned to the normal clinical appearance and the pulmonary aspect became normal. Six months after the end of the systemic treatment there was no relapse.

DISCUSSION Joint involvement in sarcoidosis often manifests itself as arthralgia, but deforming arthritis is rare.[1,2] There are 2 patterns of sarcoidal arthropathy. The early pattern is encountered in Löfgren syndrome, the acute type of the onset of sarcoidosis occurring in the first 6 months of symptoms. Polyarthralgia (of ankles, knees, proximal interphalangeal joints, wrists, and elbows) is common, but conventional radiographs of the symptomatic joints show normal joints or only osteoporosis and the swelling of soft tissue[2]. The 2nd form is observed after 6 months or more from the diagnosis and it usually involves 2 or 3 joins (ankles, knees, proximal interphalangeal joints, wrists or shoulders). In this form, dactylitis can occur. Radiographic changes usually consist of cystic bone lesions.[2,3] Bone involvement is reported to have a worse prognosis, with 4 times higher mortality rate compared to the patients with normal bone findings, but almost half of the patients have no symptoms related to bone involvement in spite of abnormal radiographic findings.[3]

In the discussed patient's case the onset of the disease, with asymmetric dactylitis that developed in a short period of time and with no constitutional complains, was the starting point of the diagnosis.

The term of dactylitis is used to describe the inflammation of a finger or a toe. In clinical practice, dactylitis is considered a hallmark feature of spondyloarthritis especially of psoriatic arthritis. Dactylitis is so specific for spondyloarthritis that it was included in the clinical criteria for the classification and the diagnosis of the whole disease group.[4] In spondyloarthritis the aspect of "sausage-like" digit is due to the flexor tenosynovitis and sometimes to the enlargement of the joint. In spite of this high specificity of dactylitis for spondyloarthritis a differential diagnosis should be carefully considered. In tuberculosis dactylitis, short tubular bones of hands and feet are affected by tuberculous osteomyelitis (the aspect of *spina ventosa* on the radiography). Syphilitic dactylitis is encountered in congenital syphilis and the manifestations are bilateral and symmetric. Blistering distal dactylitis is attributed to infection with group A β-hemolytic *Streptococcus* or *Staphylococcus aureus*, involving the volar fat pad

of the distal phalanx of the digits, and it usually presents as a fluid-filled blister. In sickle cell dactylitis, ("hand-foot syndrome") bone marrow infarction of the carpal and tarsal bones and phalanges develops. Gouty dactylitis is a rare finding, seen in a chronic polyarticular disease.[4,5]

The current patient did not fulfill the diagnostic criteria for all these diseases. Thus the decision was made to perform a surgical biopsy from the extensor part of the index, and this area was chosen for 2 reasons: first, because it was the most painful region, with a great difficulty in extension of the finger, and second, in order to avoid an unnecessary tendon sheath dissection. The histological aspect was typical of the sarcoidosis diagnosis. Search for other organ involvement (despite the absence of other symptoms) revealed bihilar lymph node enlargement with no lung parenchyma involvement.

The diagnosis of sarcoidosis is in fact established by exclusion. Since respiratory and systemic symptoms were absent, normal laboratory tests and detection of the most important histological findings consistent with the diagnosis (non-caseating granulomas and the presence of bilateral hilar lymphadenopathy) other causes of granulomatous diseases, including tuberculosis, were excluded. The main limitations of this exclusion were lack of evaluation of the tuberculin skin test and of the *Mycobacterium tuberculosis* cultures from a histological sample. Failure to assess the serum angiotensin-converting enzyme level was another limitation in our study.

Sarcoid dactylitis results from typical non-caseating granulomas invading the phalanges and the adjacent soft tissue. Tenosynovitis or tendonitis can be easily depicted by imaging techniques, the magnetic resonance imaging or ultrasonography, but the findings are unspecific to both methods, and in some cases the soft-tissue biopsy is required to establish the diagnosis. In the current patient, there were no abnormalities on clinical examination (except for the involved fingers) and in blood tests, however tenosynovitis with bone cysts indicated the likelihood of sarcoidosis.

Although cutaneous sarcoidosis lesions have been reported to be associated with skeletal lesions,[5] the described patient did not present any skin lesions during the year before the appearance of dactylitis.

Dactylitis is a rare manifestation of sarcoidosis and it even more rarely occurs as a feature of sarcoidosis. There have been few cases described in the literature. The bone involvement in sarcoidosis is associated with a chronic course of the disease, poor prognosis, and fibrotic lesions in all organ systems.[1] Pitt et al[6] reported 4 cases of sarcoidosis complicated with dactylitis, all with management problems and complications, and Wright[7] described a case complicated with resorption of phalanges. Leibowitz et al[8] described 3 cases of black South Africans with dactylitis as the 1st manifestation of sarcoidosis and assumed that these patients might form a subgroup with bone involvement and a good prognosis. The patient discussed here had dactylitis as the 1st manifestation of the disease, her response to the treatment was very good and she also had a good prognosis. The different course of dactylitis at the onset compared to dactylitis that develops during sarcoidosis is more likely to be related to causative factors than to the immunologic pathogenesis of the disease.

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