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Case report

Leprosy simulating systemic sclerosis: a case report☆

Hanseníase que simula esclerose sistêmica: relato de caso

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Introduction

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*, being highly contagious but with low morbidity due to the resistance of a good part of the population to this disease. Its worldwide prevalence has declined since the introduction of multidrug chemotherapy; but despite this, Brazil has not achieved the goal of eliminating the disease, being second only to India in the absolute number of cases.[1] Leprosy presents a broad spectrum of clinical manifestations and can be a diagnostic challenge, especially in the early years of the disease. Some of these manifestations resemble the clinical picture of certain rheumatologic diseases.[2,3] We report a case of leprosy with skin thickening, distal phalanx resorption, and facial telangiectasia, simulating systemic sclerosis.

Case report

Male patient, 69 years, with clinical manifestations that started 4 years ago with sclerodactyly associated with an unrelated-to-cold cyanosis and pallor in his extremities, with no history of digital ulceration, but with progressive resorption of distal phalanges evidenced in the clinical progression. The patient still had symptoms of sensitive polyneuropathy in his hands and feet and telangiectasias on the face. A year before diagnosis, hypoesthesic skin lesions on elbows appeared. The patient had no joint complaints, dyspnea or dysphagia. There was a history of intimate contact with a family member with a history of leprosy 6 years before the onset of the patient's symptoms. A prior research for leprosy with a skin biopsy resulted negative.

☆ Study conducted at Universidade Federal do Paraná (UFPR), Hospital de Clínicas, Serviço de Reumatologia, Curitiba, PR, Brazil.

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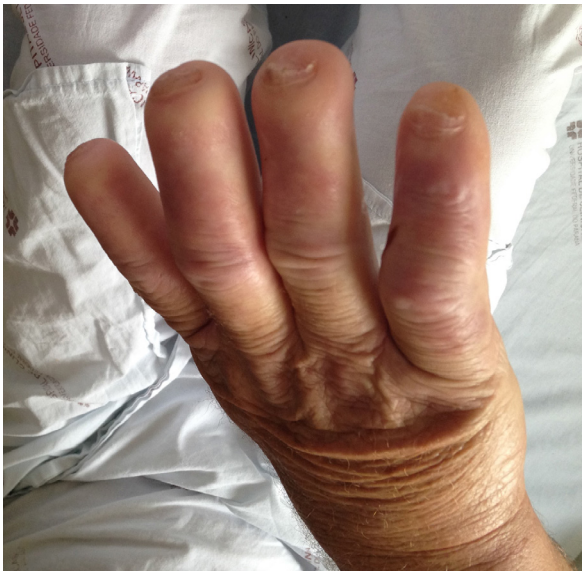

Fig. 1 – Skin thickening and resorption of distal phalanges.

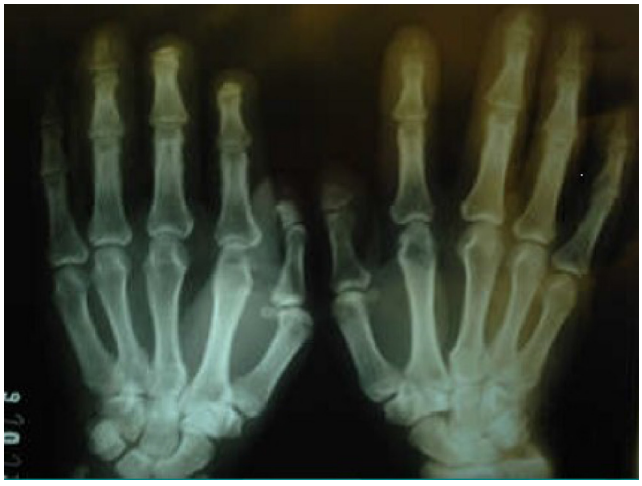

Fig. 2 – Radiography of the patient's hands showing acro-osteolysis.

In the current evaluation, distal skin and ulnar nerve bilateral thickening, resorption of distal phalanges (Fig. 1), telangiectasias on the face, as well as hypoesthesic lesions with infiltration and erythema on both elbows were evidenced. Laboratory tests were normal and a radiography of his hands showed bone resorption of distal phalanges (Fig. 2). Capillaroscopy was not performed. The patient underwent a new biopsy, which confirmed the diagnosis of tuberculoid leprosy. The patient was instructed with regard to subsequent treatment and that his close contacts should be evaluated, and proceeds with his dermatological treatment.

Discussion

Leprosy results in several clinical manifestations that depend on the host immune response. In tuberculoid leprosy, there is an efficient cell-mediated response, while lepromatous (Virchowian) leprosy is characterized by humoral immunity. Between these two extremes, intermediate forms are seen that reflect the gradual variation in resistance against the bacillus.[4]

In the early stages, the patient may present with only skin and neurological symptoms, showing no signs of involvement of other organs.[2] The musculoskeletal involvement is the third most common symptom.[5] In a study of 70 patients with leprosy, Vengadakrishnan et al. showed that 61.42% of patients showed rheumatological manifestations, the most common of them being arthritis.[6] On the other hand, a Brazilian study showed that only 6.1% of 1257 leprosy patients had joint involvement.[7]

Systemic symptoms and the presence of autoantibodies can also occur.[2] Ribeiro et al. determined the antibody profile of 158 leprosy patients, with and without joint involvement. The frequency of anticardiolipin and anti-beta 2 glycoprotein I antibodies was significantly higher in leprosy patients *versus* healthy controls.[8] Elbeialy et al. demonstrated that the presence of anticardiolipin antibodies was associated with the presence of Raynaud's phenomenon in patients with leprosy.[9]

In the literature, leprosy cases misdiagnosed as systemic lupus erythematosus, vasculitis, rheumatoid arthritis,[3] and systemic sclerosis have been described. The latter is an autoimmune disorder that causes excessive accumulation of collagen, causing skin sclerosis and fibrosis of internal organs. Lee et al. also reported a case of leprosy simulating systemic sclerosis. The patient was mistakenly diagnosed with this disease, due to the similarity of skin lesions, the presence of Raynaud's phenomenon, and the positivity of some antibodies. The skin thickening was subsequently interpreted as a longstanding skin edema secondary to leprosy plates, and not as true scleroderma.[2]

Initially, the patient described in this study was diagnosed as a systemic sclerosis carrier by presenting skin thickening in his hands, Raynaud's phenomenon, telangiectasias, and resorption of the distal phalanges, and also due to the fact an initial skin biopsy has not shown the presence of acid-fast bacilli. Resorption of distal phalanx occurs in 20–25% of patients with systemic sclerosis, and this finding is strongly associated with severe digital ischemia, suggesting that this is due to ischemic atrophy.[10] In leprosy, the changes of the distal phalanx can be of various types, for instance, a focal defect, irregularity, complete destruction, or concentric narrowing, occurring in 19–45% of patients.[11]

Despite several symptoms common to these two conditions presented by our patient, we also found anesthesia of "Stocking-Glove" Pattern, and the involvement of signals from other organs, as evidenced by the absence of dysphagia and dyspnea, and by pulmonary function tests which were within the normal range. Thus, leprosy was shown as the most likely diagnosis, which was confirmed by another skin biopsy.

Leprosy still has considerable prevalence in Brazil, and the delay in its diagnosis and treatment can result in deformities and functional impairment. Besides arthritis, the disease has many other laboratory and clinical similarities with autoimmune diseases. Thus, rheumatologists should consider leprosy as a diagnostic hypothesis in patients with musculoskeletal involvement, particularly if such patients are showing an involvement of peripheral nerves.

Conflicts of interest

The authors declare no conflicts of interest.

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