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VIRCHOWIAN LEPROSY MIMETIZING JUVENILE SYSTEMIC SCLERODERMA

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Abstract: BACKGROUND Leprosy, a chronic infectious disease caused by *Mycobacterium leprae*, affects the skin, peripheral nerves, upper respiratory tract, musculoskeletal system and eyes. As it presents a wide spectrum of clinical manifestations, it can be a diagnostic challenge, especially in early stages of disease. Some of these manifestations resemble pictures of rheumatic diseases that affect adults and children. Among the musculoskeletal manifestations of childhood leprosy are inflammatory chronic arthritis, mimicking juvenile idiopathic arthritis or spondyloarthritis, inflammatory swelling of the hands and feet, neuropathic arthritis, septic arthritis, arthralgias/myalgias, soft tissue rheumatism and multisystem involvement similar to collagenases, including vasculitis and myositis. CASE REPORT We report the case of a previously healthy 8-year-old boy admitted to the pediatric rheumatology unity due to the presence of puffy hands and fingers and hard and shiny edema of the lower limbs, suggesting systemic scleroderma diagnosis. He had an 11-month history of recurrent episodes of intermittent fever, plaque-like, erythematous, nonpruritic facial skin lesions and joint pain in the knees and ankles. On physical examination, he presented, in addition to the aforementioned findings, purpuric lesions on the toes, hepatosplenomegaly, arthralgias (wrist, small joints of the hands, knees and ankles) and erythematous plaques infiltrated in the bilateral malar region, nasal region and auricular pavilion. Complementary tests showed normocytic and normochromic anemia, leukocytosis, neutrophilia, thrombocytosis, elevation of inflammatory tests and negativity of ANA, RF, ANCA, anti-RNP and anti-Scl70. Due to the infiltrated skin lesions, lymph smear testing was performed, which was strongly positive, and a diagnosis of Virchowian leprosy associated with mixed leprosy reaction was made. There was clinical and laboratory improvement after initiation of multidrug therapy and systemic corticosteroid therapy. CONCLUSION Leprosy is known as a great mimic of rheumatic diseases, often fulfilling diagnostic criteria for many of them. The multibacillary forms present greater musculoskeletal involvement. It should be considered in differential diagnosis of children with musculoskeletal symptoms, autoantibody positivity and cutaneous and/or neurological involvement.

Keywords: leprosy**,** musculoskeletal involvement, scleroderma, children,

BACKGROUND

Leprosy, a chronic infectious disease caused by the bacillus *Mycobacterium leprae*, affects the skin, peripheral nerves, upper respiratory tract, musculoskeletal system and eyes and is one of the main causes of permanent disability in undeveloped countries. Most cases result of close contact with infected and untreated people, with transmission occurring through the upper airways (LIMA NETO et al., 2020). The incubation period is long, lasting from three to five years, but in children it can be reduced to weeks (MORAES et al., 2021). Although rare in childhood, particularly in those aged less than five years, due to their immature immune system and intrafamilial contacts, children are more susceptible to this infection (JHA; SUCHANA MARAHATTA, 2021).

Brazil is the only country in the Americas that has not reached the leprosy control goal. Worldwide, it is the second country in number of new leprosy cases and the third one with the highest rates of leprosy in children under 15 years (DA PAZ et al., 2022). The number of cases in those aged < 15 years is an important indicator for determining the level of disease transmission, and it indicates the need to intensify or implement specific prevention and control measures for this age group

(MORAES et al., 2021). The early diagnosis of leprosy is essential in the prevention of deformities, the repercussions of which are still more catastrophic when treating children younger (JHA; SUCHANA MARAHATTA, 2021).

Because its broad-spectrum clinical manifestations, leprosy in infancy can represent a diagnostic challenge, especially in early stages of the disease. The hallmark of leprosy is the presence of lesions or areas with hypo or anesthesia, that can be accompanied by hypohidrosis, alopecia, nerve thickening, and/or ache (BARRETO et al., 2017), but skin lesions may vary from an asymptomatic patch to diffuse infiltration of the entire skin (JHA; SUCHANA MARAHATTA, 2021). The definition of clinical cases is based on the number of lesions: up to five cutaneous lesions in the paucibacillary pole (represented by the indeterminate and tuberculoid forms), and above five lesions in the multibacillary pole (represented by dimorphic and Virchowian forms) (MOTA et al., 2021).

Indeterminate leprosy is characterized by a restricted macular condition, with a few hypopigmented macules, sometimes erythematous, of unclear outer limits, diverse sizes, with change in the thermal and/ or pain sensitivity, alopecia, and hypo or anhydrous, but all clinical forms might have hypopigmented lesions (MOTA et al., 2021). Usually affects children under 10 years old and rarely adolescents. The tuberculoid form manifests as lesions in a fully infiltrated plaque or of erythematous-papular sharp borders, which are externally defined, with change in the thermal, pain, and tactile sensitivity (in this order), hypo or anhydrous areas, and alopecia (DE OLIVEIRA; DINIZ, 2016).

Dimorphic leprosy, the most common form in adults, presents with erythematous or hypopigmented macules or tuberculoid-like lesions with raised, poorly defined borders (BARRETO et al., 2017). The Virchowian form can course with erythemato-violaceous plaques, livedo reticularis, nodules, infiltration, madarosis and xeroderma (GOMES et al., 2021). All of the above classifications are also used to establish the clinical forms in children and adolescents. However, in this age group, the studies show a predominance of the paucibacillary forms (DE OLIVEIRA; DINIZ, 2016). Reaction episodes, which are characterized by exacerbation of inflammatory processes, either localized or systemic, will be present in about 5–20% of children with leprosy at some point before, during, and/or after the end of treatment (BANDEIRA; PIRES; QUARESMA, 2019).

Some of leprosy manifestations resemble pictures of rheumatic diseases that affect adults and children. Musculoskeletal involvement occurs predominantly in multibacillary forms and leprosy reactions, which are somewhat less observed among pediatric cases (NEDER et al., 2014). It is second to the cutaneous and neurologic manifestations (CHOPRA, 2014). Inflammatory arthritis, often a component of leprosy reactions, can be the first manifestation of the disease and is one of the most frequent rheumatologic symptoms in adults, although not usually significant (SALVI; CHOPRA, 2013).

Among the musculoskeletal manifestations of childhood leprosy are inflammatory chronic arthritis, mimicking juvenile idiopathic arthritis (mostly the poliarticular form) or spondyloarthritis, inflammatory swelling of the hands and feet, neuropathic arthritis, septic arthritis, arthralgias/ myalgias, soft tissue rheumatism and multisystem involvement similar to collagenases, including vasculitis and myositis (CHOPRA, 2014). In addition to systemic symptoms, the presence of autoantibodies may also occur (NEDER et al., 2014).

Cases of leprosy misdiagnosed as systemic

lupus erythematosus, vasculitis, rheumatoid arthritis and systemic sclerosis have been described in the literature (CHU et al., 2017), mostly in adults. Juvenile systemic sclerosis is a multisystem connective tissue disorder characterized by fibrotic skin changes and abnormalities of internal organs in children under 16 years old (ZULIAN, 2017). It should be considered in a child with moderate to severe Raynaud phenomenon, especially if it is associated with digital ulcers and/or abnormal nailfold capillaroscopy, and limited range of finger motion from edema or skin thickening (sclerodactyly) (LI, 2018).

CASE REPORT

An 8-year-old boy, with no comorbidities, was admitted to the Pediatric Rheumatology Unit with a 12-day history of daily fever associated with edema on the dorsum of the hands, fingers and lower limbs, with overlying, shiny skin (Images 1 and 2) and arthralgia of the knees and ankles. In the last three days, it evolved with the presence of erythematous lesions on the face. He had an 11-month history of episodic fever, erythematous plaque-like, non-pruritic lesions on the face, and arthralgia in the knees and ankles, remaining asymptomatic between attacks. The parents denied family history of rheumatic diseases and contact with endemic infectious diseases.

He was hospitalized in the first month of symptoms presenting laboratory abnormalities with normocytic and normochromic anemia, thrombocytosis and elevation of erythrocyte sedimentation rate, aspartate amino transferase and D-dimer, in addition to hypoalbuminemia and positive PCR for SARS-CoV-2. After ruling out other infectious and neoplastic diseases, he was diagnosed with multisystem inflammatory syndrome associated with COVID-19 at the time, evolving with favorable response to corticosteroid therapy and acetylsalicylic acid. However, after discharge, the family did not maintain medical follow-up and discontinued use of medications.

In the physical examination of the current hospitalization, in addition to the condition already described, he presented purpuric lesions on the toes (image 3), hepatosplenomegaly and erythematous plaques infiltrated in the malar region, nose and ear lobe (image 4 and 5). Tests showed anemia, leukocytosis, neutrophilia, thrombocytosis, and elevated inflammatory markers. Due to the diagnostic possibility of juvenile systemic scleroderma (edematous phase), although the patient did not present with other signs or symptoms compatible with such hypotheses, autoantibodies (antinuclear antibody, rheumatoid factor, Anti-RNP and anti-Scl70) were performed with negative results.

Due to the lesions on his face, he underwent a lymphatic bacilloscopy which was positive for *Mycobacterium leprae*. After dermatological evaluation dermatology, Virchowian leprosy with mixed leprosy reaction was diagnosed. He received multidrug therapy and systemic corticosteroid therapy at an immunosuppressive dose presenting excellent clinical and laboratorial improvement.

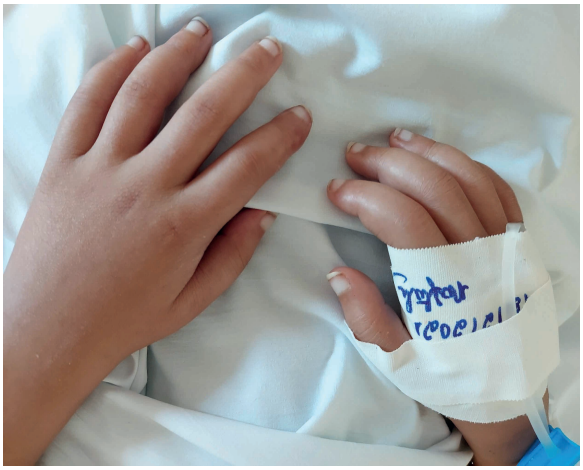


Image 1: Puffy hands and fingers.

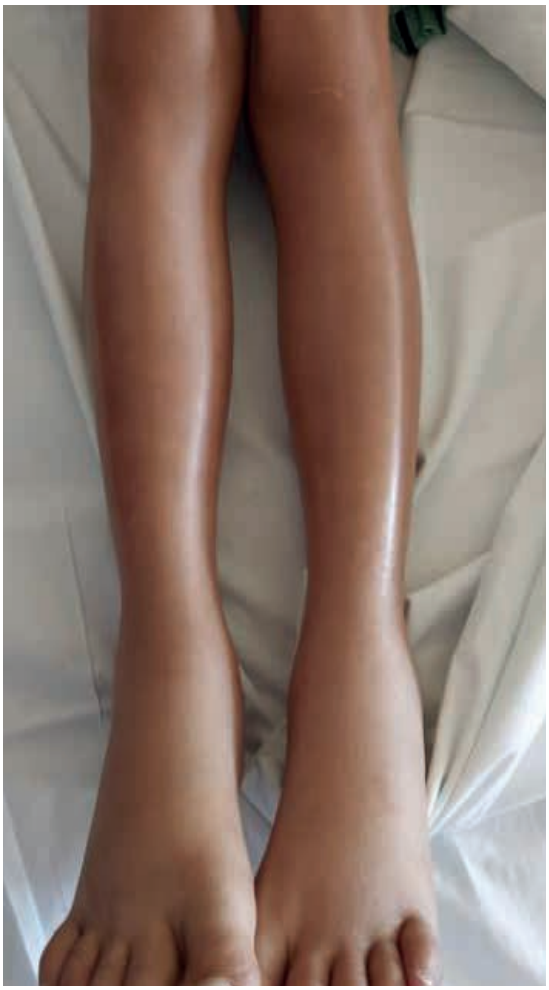


Image 2: Lower limbs edema with shiny and warm skin.

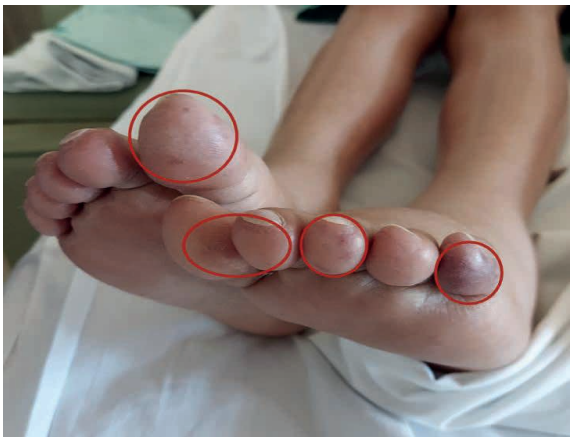
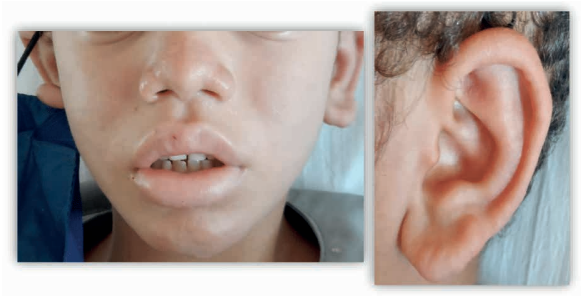


Image 3: Purpuric lesions on toes.



Images 4 and 5: Erythematous and infiltrated plaques in the bilateral malar region, nasal region and ear lobe.

CONCLUSIONS

Leprosy is an infectious disease capable of mimicking rheumatic diseases, sometimes fulfilling diagnostic criteria for many of them. The present report describes a case of childhood leprosy with musculoskeletal manifestations suggestive of juvenile systemic scleroderma in its initial (edematous) phase. The multibacillary forms of leprosy present greater musculoskeletal involvement, and despite the positivity of autoantibodies described in the various forms of the disease, these were not found in this patient.

In pediatrics, leprosy with musculoskeletal involvement has been rarely described in the literature, which makes its diagnosis even more difficult. It is important to include leprosy in the differential diagnosis in children with a clinical condition suggestive of rheumatological diseases and/or with positive autoantibodies and cutaneous/neurological involvement, especially in endemic areas, such as Brazil.

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