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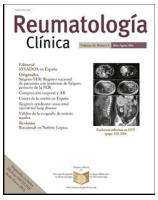

Case report

Sarcoidosis: An unusual presentation

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ABSTRACT

A 35-year-old man presented with a 3-year history of arthralgia and purple coloration of the skin of his fingers and feet.

Hand and foot radiography showed cystic bone lesions on phalanges suggestive of sarcoidosis. Lab tests revealed increased liver enzymes. Liver MRI evidenced an enlarged liver and retroperitoneal lymphadenopathy. Histological analysis of the finger skin, lymph nodes and liver demonstrated the presence of granulomas, confirming the diagnosis of sarcoidosis. The patient started prednisolone with rapid improvement of the symptoms.

Skin lesions are divided into two groups: specific for sarcoidosis (with granulomas, lupus pernio-like) and nonspecific (without granulomas, erythema nodosum-like). Specific cutaneous lesions usually cause no other symptoms beyond cosmetic changes. Lupus pernio stands out for having distinctive features but, to the best of our knowledge, the simultaneous involvement of both hands and feet has never been reported.

Sarcoidosis: un cuadro clínico inicial poco frecuente

Palabras clave:
Sarcoidosis
Lupus pernio
Lesiones óseas

RESUMEN

Se presenta el caso de un hombre de 35 años con una historia de artralgia y con la piel de los dedos y los pies de color violáceo, de 3 años de duración.

La radiografía de pies y manos mostró lesiones quísticas óseas en las falanges, indicativas de sarcoidosis. Las pruebas de laboratorio revelaron una elevación de las enzimas hepáticas. La resonancia magnética hepática puso de manifiesto hepatomegalia y linfadenopatía retroperitoneal. El análisis histológico de la piel de los dedos, los ganglios linfáticos y el hígado mostró la existencia de granulomas, lo que confirmó el diagnóstico de sarcoidosis. El paciente comenzó el tratamiento con prednisolona con una rápida mejoría de los síntomas.

Las lesiones de la piel se dividen en 2 grupos: específicas de la sarcoidosis (con granulomas y lupus característico del eritema pernio) e inespecíficas (sin granulomas y de tipo eritema nudoso). Las lesiones cutáneas específicas generalmente no causan más síntomas que los cambios estéticos. El lupus pernio destaca por presentar características distintivas, pero no nos consta que se haya descrito nunca la afectación simultánea de ambas manos y pies.

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Clinical observation

A 35 years-old man was sent to the rheumatology consultation with a 3-year history of arthralgia and purple coloration of the skin of the fingers of the hands and feet (Fig. 1). The patient complained of purple swollen of the skin with joint pain and reduced range of motion.

The lab tests showed an increase of the liver enzymes: AST 95 U/L (10–37), ALT 130 U/L (10–37), gamma-GT 317 U/L (10–49), alkaline phosphatase 561 U/L (30–120), hypergammaglobulinemia, a low titer positive rheumatoid test and a positive anti-smooth muscle antibody. Abdominal ultrasound and a liver MRI evidenced an enlarged liver (without focal lesions) and retroperitoneal lymphadenopathy. A lymphoproliferative disease was suspected.

Liver and lymph node biopsies showed the presence of epithelioid granulomas. Infectious causes associated with granulomatous processes were excluded.

Hands and feet radiography evidenced the presence of bone cystic lesions on phalanges suggestive of sarcoidosis.

We also assessed the involvement of other organs. Full body scintigraphy with gallium showed hyperfixation in several mediastinal and abdominal lymph nodes and high resolution chest-CT confirmed these findings, in a pattern suggestive of lymph node sarcoidosis. No lesions were found in the pulmonary parenchyma. Angiotensin converting enzyme (ACE) level was significantly elevated (117 U/L, range 20–70).

The patient was submitted to a skin biopsy, in which were also visible epithelioid granulomas with rare giant multinucleated cells.

Confirmed the diagnosis of sarcoidosis with bone, skin, liver and lymph node involvement, the patient started prednisolone 40 mg/day, with rapid improvement of the joint pain, skin lesions (Fig. 1) and analytical abnormalities.

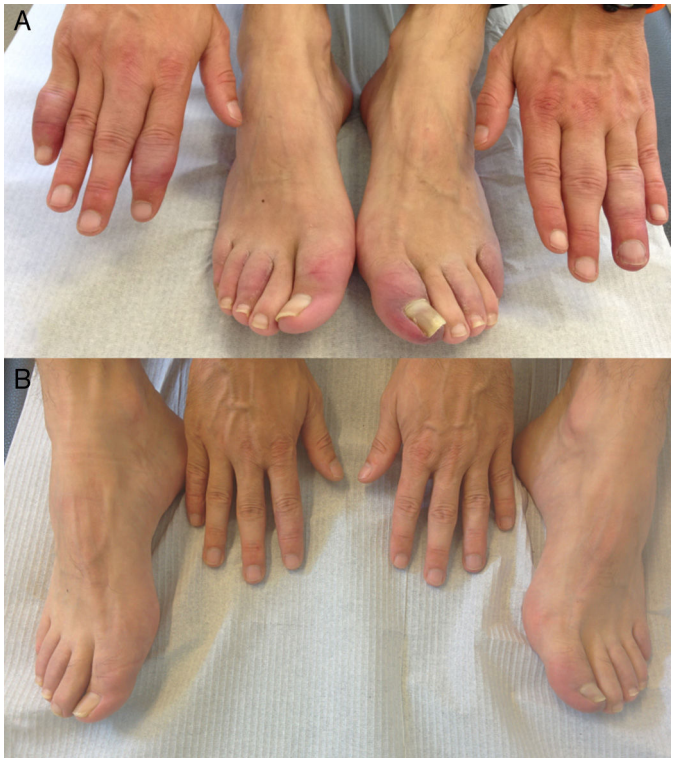

Fig. 1. Hand and feet skin lesions at presentation (A) and after 10 months of treatment (B).

Discussion

Sarcoidosis is a rare disease with heterogeneous presentation characterized by the formation of non-caseating granulomas.

Bone involvement is usually associated to lung, skin and lymph node involvement,[1,2] as in the case described. Bone lesions are asymptomatic in half of the cases, although they may present with bone or joint pain or edema. In conventional X-ray, bone lesions usually appear as cystic, lytic or permeative lesions, and are usually visible on phalanges.[1,3] Asymptomatic patients usually do not require specific treatment.[1] Symptomatic patients may need treatment: NSAIDs, corticosteroids, methotrexate, hydroxychloroquine, and infliximab are described in the literature for this purpose.[1,2]

Skin lesions have a predilection for black women.[4] They are divided into two large groups: those specific of sarcoidosis (with evidence of granuloma) and nonspecific (with inflammatory changes but no identifiable granulomas).[5,6] Erythema nodosum is the most common non-specific injury of sarcoidosis, and is a sign of the Löfgren syndrome.[7] The specific cutaneous lesions of sarcoidosis usually cause no other symptoms beyond the cosmetic changes and are very heterogeneous in their appearance.[5] In this group stands out lupus pernio for being one of the few lesions with some distinctive features: violet papules and nodules that coalesce on plaques, with predilection for face, especially nose, ears and perioral region.[4] They can also appear in the fingers,[4,6] but the simultaneous involvement in both hands and feet never was described to the best of our knowledge, as seen in this case presented.

Most patients remain asymptomatic and have only a slight increase in liver function enzymes.[8] Treatment with corticosteroids usually normalize liver enzymes, but is not recommended for asymptomatic patients with no involvement of other organs, since delay in treatment rarely leads to significant liver damage.[9]

Conclusion

Our clinical case emphasizes the difficulty in diagnosis with an atypical presentation. It is very important to keep a high level of awareness to the diagnosis of sarcoidosis. Joint and bone involvement is uncommon, the skin lesions will be variable and it is important to know them.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Conflicts of interest

The authors declare no conflict of interest.

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