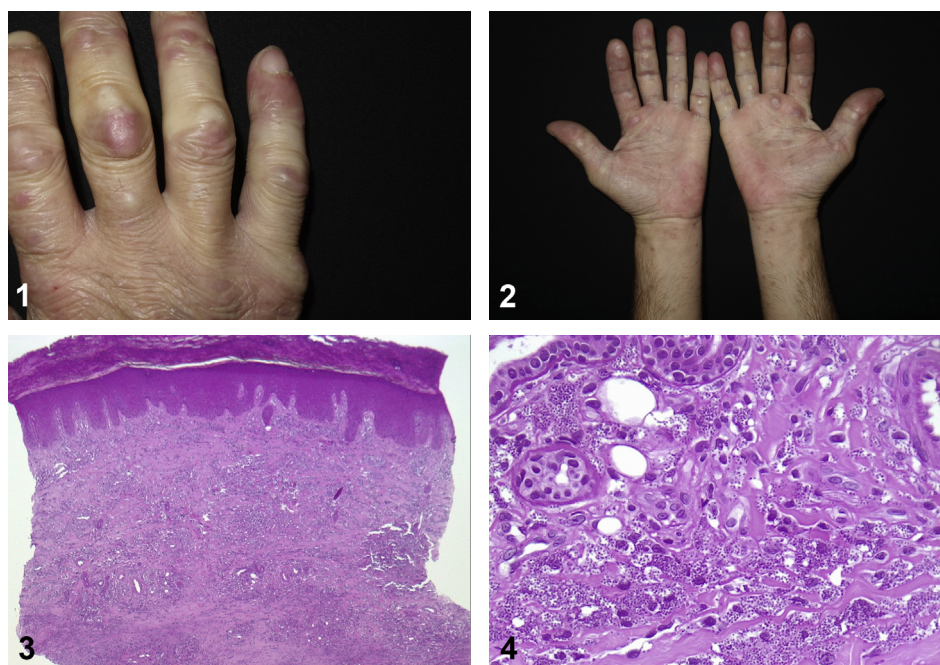


Violaceous nodules over dorsal interphalangeal joints in a patient with HIV



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CASE PRESENTATION

A 49-year-old HIV-positive patient with a history of intravenous drug use had painful erythematous-violaceous papules and nodules on the dorsum of his hands, especially over the interphalangeal joints (Figs 1 and 2). He also had some hyperkeratotic nodular lesions in his palms. Hepatomegaly was present. He was hepatitis B and C positive and had liver fibrosis. The CD4 cell count was $38/\text{mm}^3$ but had an undetectable HIV viral load. His medications included antiretroviral therapy, methadone and antipsychotic drugs. He had no fever, headache, sore throat, cough, weight loss or muscular weakness. Laboratory tests found anemia, leukopenia, and thrombocytopenia. A skin biopsy was performed (Figs 3 and 4).

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Question 1: Considering clinical and histopathologic images, what is the most likely diagnosis?

- A. Nodular secondary syphilis
- B. Amyopathic dermatomyositis
- C. Atypical leishmaniasis—associated HIV
- D. Erythema elevatum diutinum
- E. Granuloma annulare

Answers:

A. Nodular secondary syphilis — Incorrect. Typically known as the *great imitator*, secondary syphilis often mimics other diseases or leads to extremely rare skin lesions. Few cases of nodular syphilis have been reported. Clinically, the lesions appear as plaques or red-violaceous nodules, solitary or multiple, and partially infiltrated.¹ Palms and soles usually present with papules and plaques with a collaret of scale (collarette of Bielt). Histopathology found nonnecrotizing granulomas and an interface dermatitis, with a dense dermal lymphoplasmacytic infiltrate.

B. Amyopathic dermatomyositis — Incorrect. The features that are classically considered pathognomonic for dermatomyositis include the heliotrope sign and Gottron papules. Other distinctive features are the photodistributed, pink-violet poikiloderma involving the upper chest and back in addition to cuticle nail dystrophy and nail fold telangiectasias.² The characteristic changes seen in skin biopsy specimens can be very subtle and include epidermal atrophy and signs of interface vacuolar dermatitis.

C. Atypical leishmaniasis associated HIV — Correct. Cutaneous lesions are sometimes the first sign of visceral involvement. We have found only 3 AIDS patients with leishmaniasis presenting as a dermatomyositis-like eruption. They had lesions limited to the dorsum of hands, wrists, and elbows. The morphology and location of the lesions led us initially to make a misdiagnosis of dermatomyositis.³

D. Erythema elevatum diutinum — Incorrect. Erythema elevatum diutinum is a rare, chronic dermatosis characterized by red-violet papules or nodules over extensor surfaces. An earlier onset occurs more often in the setting of HIV infection.⁴ Histopathologic features consist of a fibrosing leukocytoclastic vasculitis with a neutrophilic infiltrate.

E. Granuloma annulare — Incorrect. Granuloma annulare is a benign, usually self-limited, cutaneous disease that presents as arciform to annular plaques on the dorsal hands or feet of young people. Deep dermal or subcutaneous granuloma annulare manifests as large, painless, skin-colored nodules, which may be mistaken for rheumatoid nodules.⁵ Interstitial pattern or nonnecrotizing granulomatous pattern with focal degeneration of collagen and elastic fibers and mucin deposition can be observed in skin biopsy.

Question 2: Which of the following complementary tests and techniques would be of additional use in this case?

- A. Rapid plasma reagin test and fluorescent treponemal antibody absorption (FTA-ABS)
- B. Montenegro skin test
- C. Bone marrow biopsy
- D. Anti-MDA5 (CADM-140)
- E. Muscular magnetic resonance imaging (MRI)

Answers:

A. Rapid plasma reagin test and fluorescent treponemal antibody absorption (FTA-ABS) — Incorrect. Syphilitic serologic tests are the gold standard in the diagnosis of primary and secondary syphilis. In secondary syphilis, both treponemic and nontreponemic tests are usually positive.¹

B. Montenegro skin test — Incorrect. This test uses leishmanial antigens to induce a cell-mediated response, and it has traditionally been an important diagnostic tool. However, it is invariably negative during the febrile phase of visceral leishmaniasis and in patients with diffuse cutaneous (anergic) leishmaniasis like in this case.⁶

C. Bone marrow biopsy — Correct. This sample may be used for culture and polymerase chain reaction–based assays. In our patient, culture and polymerase chain reaction of bone marrow were positive for *Leishmania donovani*.

D. Anti-MDA5 (CADM-140) — Incorrect. The autoantigen CADM-140 was subsequently found to be identical to 2 previously identified gene products, interferon induced with helicase C domain protein 1 (IFIH1) and melanoma differentiation–associated gene 5 (MDA5). In some large series, this autoantibody is positive in 65% of cases of clinically amyopathic dermatomyositis.²

E. Muscular MRI — Incorrect. MRI of proximal muscles is used in the evaluation for myositis in case of suspected dermatomyositis.

Question 3: Observing the histopathologic samples of Figs 3 and 4, which of the following is FALSE?

A. Many organisms within macrophages and among collagen fibers throughout the papillary and reticular dermis were observed.

B. Amastigotes can be seen around eccrine glands.

C. Inflammation is absent.

D. CD1a stain can be useful to detect amastigotes.

E. Tuberculoid granulomas with caseation necrosis is observed.

Answers:

A. Many organisms within macrophages and among collagen fibers throughout the papillary and reticular dermis were observed — Incorrect. It is observed in Figs 3 and 4.

B. Amastigotes can be seen around eccrine glands — Incorrect. It is observed in Fig 4.

C. Inflammation is absent — Correct. It is observed in Fig 3.

D. CD1a stain can be useful to detect amastigotes — Incorrect. CD1a (MTB1 clone) can be a useful stain to help detect amastigotes in cutaneous leishmaniasis and can be used in both Old World and New World leishmaniasis. The pattern of CD1a immunostaining of amastigotes is characteristic, with peripheral positivity, a negative nucleus in the center, and reinforcement of the kinetoplast in 1 pole. *Leishmania* amastigotes were langerin negative. Visceral *Leishmania amastigotes* also express CD1a.

E. Tuberculoid granulomas with caseation necrosis is observed — Correct. This feature is observed in mycobacterial infections but not in cutaneous leishmaniasis. Daudén et al³ observed that the histopathologic findings from their AIDS patients with leishmaniasis mimicking a dermatomyositis differed significantly from those observed in immunocompetent persons. The huge numbers of *Leishmania* organisms, the absence of an inflammatory reaction, the involvement of the reticular dermis, eccrine glands, and fat lobules, and the presence of *Leishmania* organisms inside keratinocytes are extremely rare events that have never been seen even in a large series of immunocompetent patients. Even, the presence of amastigotes within eccrine ducts suggest transepithelial elimination through eccrine sweat glands.³ Our patient was treated with intravenous liposomal amphotericin B with complete resolution of skin lesions in 4 weeks.

Abbreviation used:

MRI: magnetic resonance imaging

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