

## Extensor plaques with associated arthritis and neuropathy



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A 68-year-old man with family history of psoriasis presented with a 1.5-year history of neuropathic pain in his feet associated with sudden appearance of peripheral edema (Fig 1). After having an unremarkable electromyogram, he was referred to the rheumatology department for initiation of adalimumab for polyarthralgias and erythematous plaques on extensor surfaces. Physical examination found a painless erythematous and edematous left ear (Fig 2); oval erythematous plaques on extensor elbows, knees, and trunk (Fig 3); pitting edema bilaterally to mid shins; decreased sensation to light touch on bilateral plantar feet; and tender small joints in his hands. Autoimmune, vascular, cardiac, and imaging studies were unrevealing.

**Question 1: What is the most likely diagnosis?**

- A. Psoriasis
- B. Relapsing polychondritis
- C. Hansen disease (HD)
- D. Sarcoidosis
- E. Granuloma annulare (GA)

**Answers:**

**A.** Psoriasis — Incorrect. Although psoriasis commonly involves the extensor surfaces and has associated oligoarthritis of small joints, psoriasis does not cause an erythematous, edematous ear, bilateral peripheral edema, and sensory loss. Chronic arthritis involving distal joints has been identified as a presenting symptom of HD but may lead to misdiagnosis as a rheumatologic disorder.<sup>1,2</sup>

**B.** Relapsing polychondritis — Incorrect. Relapsing polychondritis presents with pain, edema, and erythema of cartilaginous ear and spares the lobe.

**C.** HD — Correct. HD is a chronic granulomatous infection caused by *Mycobacterium leprae* that presents with cutaneous and peripheral nerve involvement. HD is classified based on immunologic response: lepromatous, predominantly T helper cell 2 mediated, and tuberculoid, predominantly T helper cell 1 mediated. Lepromatous leprosy classically presents as multiple, poorly defined, erythematous macules, papules, nodules and plaques.<sup>2,3</sup> A biopsy of the right elbow, left ear, and back found a dermal noncaseating granulomatous infiltrate. A Fite

stain for mycobacteria was diffusely and strongly positive, indicating multibacillary leprosy.

**D.** Sarcoidosis — Incorrect. Cutaneous sarcoid commonly presents as red-brown or violaceous papules and plaques that favor the perinasal, perioral, and periocular regions of the face. Common systemic manifestations include dyspnea, lymphadenopathy, and uveitis. Arthritis and neuropathies can also be seen in sarcoidosis.

**E.** GA — Incorrect. GA presents as pink, asymptomatic annular plaques commonly on dorsal hands. There are possible associations with systemic diseases; however, joint pain, neuropathy, and peripheral edema are not commonly involved in GA.

**Question 2: Which of the following diagnostic tests has the highest specificity for leprosy?**

- A. Acid-fast bacilli
- B. Culture
- C. Serologic assays
- D. Polymerase chain reaction (PCR)
- E. Lepromin test

**Answers:**

**A.** Acid-fast bacilli — Incorrect. The bacilli on histology may not be detected. The cell wall of *M leprae* is less acid fast and alcohol fast compared with other mycobacterial species, thus unable to withstand the decolorization process. A Fite stain, which is the most commonly used stain, uses

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peanut oil to minimize the exposure of the cell wall to other solvents during the staining process.

**B.** Culture — Incorrect. *M leprae* cannot be cultured in vitro. It must be cultivated in mouse footpads or in armadillos.

**C.** Serologic assays — Incorrect. Serologic tests including natural disaccharide octyl — leprosy IDRI diagnostic 1 (NDO-LID) and Phenolic glycolipid 1 (PGL1) have limited capacity in the diagnosis of leprosy but may be useful as a supportive tool. These assays have a high negative predictive value, which could exclude leprosy in endemic areas. Both tests have a better capacity to detect individuals with multibacillary leprosy but were inefficient for the diagnosis of paucibacillary disease. This finding reflects the immune response developed by the host. Paucibacillary cases mount an effective cellular immune response to control bacterial replication, which mitigates antibody responses.<sup>4,5</sup>

**D.** PCR — Correct. Identification of *M leprae* is possible by using PCR, and this technique can be applied to skin biopsy samples, skin smears, nerves, urine, oral or nasal swabs, and blood. PCR confirms the diagnosis especially in difficult cases, detects resistance, and provides treatment follow-up.<sup>4</sup>

**E.** Lepromin test — Incorrect. The lepromin test is an intradermal injection of heat-killed *M leprae* that provides prognostic information. A nodule at the site of injection in 3 to 4 weeks indicates a positive test and ability to mount an immune response.

**Question 3: A patient with lepromatous leprosy has fever, myalgias, joint swelling, and nodular skin lesions. What is the treatment of choice?**

- A. Systemic steroids
- B. Thalidomide
- C. Clofazimine
- D. Dapsone
- E. Clarithromycin

**Answers:**

**A.** Systemic steroids — Incorrect. Oral prednisone is used for type I (reversal) reactions, which are caused by a delayed-type hypersensitivity reaction.

Type I reactions occur mostly in borderline tuberculoid, and existing skin lesions become more erythematous, edematous, and raised. However, systemic steroids are necessary in type II reactions when neuritis or iritis is present.<sup>3</sup>

**B.** Thalidomide — Correct. The patient is experiencing a type II reaction, which represents a cutaneous and systemic small vessel vasculitis. Thalidomide remains the treatment of choice for type II reactions even though teratogenicity is a major concern.<sup>3,4</sup>

**C.** Clofazimine — Incorrect. Although clofazimine has possible benefits in type II reactions, it is not the treatment of choice. With the use of clofazimine in the multidrug therapy, the frequency of type II reactions has decreased.<sup>3</sup>

**D.** Dapsone — Incorrect. Dapsone is part of the multidrug therapy for leprosy. For paucibacillary (2-5 skin lesions) leprosy, treatment consists of rifampin and dapsone. For multibacillary (>5 lesions), treatment consists of rifampin, clofazimine, and dapsone.

**E.** Clarithromycin — Incorrect. Clarithromycin is a macrolide antibiotic used to treat atypical mycobacterium infections such as *Mycobacterium fortuitum*, *Mycobacterium chelonae*, and *Mycobacterium abscessus*.

**Abbreviations used:**

GA: granuloma annulare  
HD: Hansen disease  
PCR: polymerase chain reaction

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