Leprosy, a Pleitropic infectious disease: a challenging diagnosis

Manal El Meniawy, Mervat Essam, Ymna Khaled

Department of Internal Medicine, Faculty of Medicine, Cairo University, Cairo, Egypt

Correspondence to Mervat Essam, MD Internal Medicine, Department of Internal Medicine, Faculty of Medicine, Cairo University, Cairo, 11562, Egypt. Tel: +20 011 2024 5071; fax: +20 26 28 884; e-mail: lambaa2020@yahoo.com

Received 4 July 2017
Accepted 23 September 2017

The Egyptian Journal of Internal Medicine 2018, 30:40–42

This is a case report of 22-year-old man who was suffering from epididymo-orchitis for more than 2 years. Several months after the onset of the condition, the patient developed bilateral upper-limb and lower-limb numbness and tingling sensation with hypothesia, which was further complicated by nonhealing foot ulcer, arthralgia, and generalized maculopapular skin rash. The patient was initially managed as rheumatoid arthritis associated with vasculitis, which was later diagnosed as lepromatous leprosy. Musculoskeletal complaints are not exclusive to only autoimmune diseases; it can also be observed in several disorders, such as infectious diseases. It is challenging for any physician to properly diagnose patients with leprosy as differentiating leprosy from other systemic rheumatic disease is pivotal.

Keywords:
arthralgia, epididymo-orchitis, leprosy, rash

Egypt J Intern Med 30:40–42

1110-7782

Introduction

Leprosy is a chronic granulomatous infectious disease caused by *Mycobacterium leprae* [1]. Interestingly, Egypt is one of most ancient places where leprosy was observed [2]; the oldest recoded leprous case was from Dakhleh Oasis [3].

Leprosy is a pleiotropic disease. It likens many dermatologic and neurologic disorders, rheumatologic diseases such as rheumatoid arthritis (RA), systemic lupus erythematosus, and systemic vasculitis [3], and affects multiple organs making its diagnosis challenging [4].

Many studies have revealed that testicular affection occurs mainly in the lepromatous type leprosy [5]. The constellation of evident neurological and dermatological with other rheumatic features increases the possibility of diagnosis of leprosy, especially in endemic area.

Case report

A 22-year-old male patient complained of recurrent, persistent, nonradiating, burning scrotal pain, with no precipitating or relieving factors, along with tenderness, redness, and hotness, for a period over 36 months. The patient had no history of penile discharge or genital ulcers or trauma. The episodes were associated with low-grade fever. The patient was diagnosed as epididymo-orchitis with partial response to antibiotics.

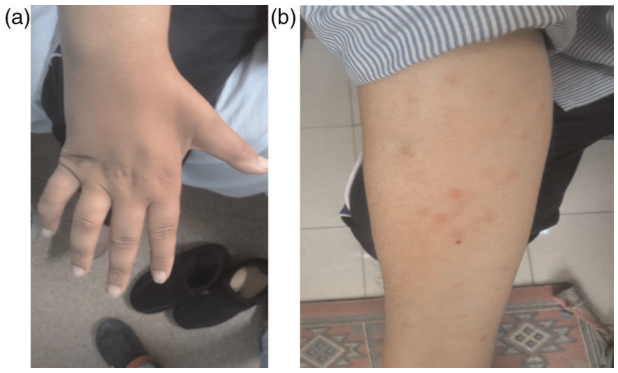
Eight months later, the patient developed bilateral upper-limb and lower-limb numbness and tingling sensation with hypothesia, which was further complicated by nonhealing foot ulcer, arthralgia of both elbows, knees, ankles, and small joints of hands proximal and distal interphalangeal joints (PIPs and DIPs), generalized nonpruritic maculopapular skin rash. He sought medical advice and was diagnosed to have RA along with Still' disease. He was prescribed steroids and immunosuppressive drugs before his admission to our hospital with no amelioration.

On hospital admittance, systematic examination revealed cushingoid facies, wasting of hypothenar muscles and 1st interossei of both hands with generalized non tender maculopapular rash over both forearms with flexion deformity of little and ring fingers of RT hand (Fig. 1a and b). There was deep ulcer on the left sole about 3×3 cm with clear base, inverted edges with skin macules allover both legs (Fig. 2a and b).

There was decrease in superficial sensations on both upper limbs up to elbow (C5, C6, C8, and T1) and both lower limbs along the distribution of L4, L5 and S1, S2. Ulnar nerve was thickened while common peroneal nerve was not felt. There was tenderness in both wrists. Cardiac, pulmonary, abdominal and genital review was normal. Fundus examination was normal. Complete blood count showed leukocytosis with absolute neutrophilia and lymphocytosis. Erythrocyte sedimentation rate

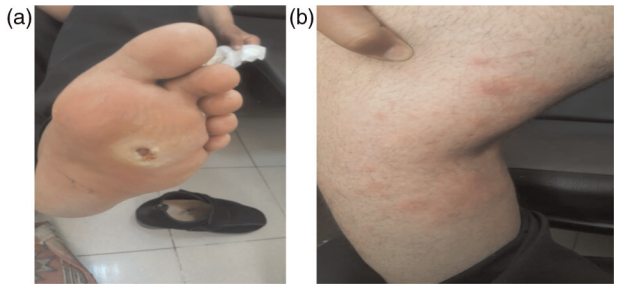
 DOI: 10.4103/ejim.ejim_44_17

Figure 1



(a) Wasting of small muscle of hands with flexion deformity of little and ring fingers of the right hand. (b) Maculopapular rash over the upper limb.

Figure 2



(a) Deep ulcer on left sole. (b) Maculopapular rash over the leg.

(70 mm/h) and C-reactive protein (76 u/l) level were elevated in addition to high ferritin level (558.1 ng/ml).

The liver and kidney function tests were normal and antinuclear antibody, rheumatoid factor, and anti-neutrophil cytoplasmic antibody tests were negative as well as viral markers for HIV, hepatitis C virus, and hepatitis B virus.

Chest radiograph and abdominal sonograph were normal; however, scrotal ultrasonography revealed bilateral diffuse abnormal testicular texture. Sensorimotor nerve conduction studies showed prolonged distal delay and slow conduction of both right and left median and ulnar nerves.

Ophthalmological examination showed no abnormality.

On the basis of cutaneous and neurological signs, leprosy was suspected, which was confirmed by dermatological consultation.

Six slit-skin smears were carried out at a leprosy sanatorium and the result was positive for leprosy (4+).

The patient was diagnosed to have lepromatous leprosy with type 2 reaction and was admitted to a leprosy sanatorium. He was prescribed multidrug therapy with steroids. On contacting the patients after 6 months of treatment, he claimed that his conditions have ameliorated with regard to the skin and joint affection.

Discussion

Leprosy is a chronic granulomatous infection caused by obligate intracellular, slowly growing, Gram-positive, acid-fast *M. leprae* [6]. Different classifications have been identified depending on the reaction of the mycobacteria with the host immunity, mainly T-cell reactivity [7], resulting into a spectrum of various pathological forms switching from one form to another [8]. The clinical manifestations include mainly skin and neuropathy, which were the clue in our case.

Leprotic immunological reactions are of two types: type 1 (T1R) (reversal), which occurs in paucibacillary leprosy and presents with exacerbation of already existing erythema and swelling of skin lesions along with acute neuritis and low-grade fever [6].

The type 2 (T2R) leprotic immune-mediated reaction is called 'erythema nodosum leprosum (ENL)'. It occurs in lepromatous (multibacillary) leprosy. This form is characterized by severe illness, which is associated with fever, fatigue, hepatosplenomegaly, lymphadenitis, arthritis, glomerulonephritis, neuritis, painful skin nodules, and orchitis [3].

Testicular involvement in leprosy is frequent. Khanolkat [9] had observed that the testes and their adnexa are preferentially affected in lepromatous leprosy and it can also occur during episodes of ENL reaction [10]. However, it needs to be emphasized that acute epididymo-orchitis in leprosy is not sexual and venereal in origin. It may be attributed to fact that the immune response to lepromatous leprosy is humoral mediated, in which immune cells directly attack the bacillus and infiltrates it to the surface of the testes and epididymis; and finally spermatocyte reaction due to testicular hypersensitivity [11].

Leprosy as an infectious disease can mimic symptoms of common, systemic autoimmune diseases or can clinically manifest as malignancy such as lymphoma. The coexistence of different dermatological and neurological symptoms along with joint affection

and epididymo-orchitis in our case raised the possibility of different rheumatic disorders such as RA, systemic lupus erythematosus, vasculitis, or adult Still' disease for which he was prescribed steroids. As there was very little improvement of our patient, we started to revise the presumed rheumatologic diagnosis and put leprosy disease into consideration. The case was assumed to have T2R leprosy despite absence of erythema nodosum (EN); as unusual attack of T2R without EN could be seen [12]. This matches with Marahatta *et al.* [13], who reported a case of histoid leprosy, which is an unusual variant of lepromatous leprosy. Their case had unusual type 2 lepra reaction with constitutional symptoms, perichondritis, and epididymo-orchitis.Tilak [14] had reported four cases of advanced lepromatous leprosy, of which three were frankly impotent; chronic inflammatory tissues with positive AFB were evident in the biopsies of testicular specimens of two of the cases. This was unlike our case, who presented with recurrent episodes of acute epididymo-orchitis for months before developing other defined neurological and dermatological symptoms.

Faten *et al*. [15] assessed 40 patients with lepromatous leprosy for testicular function and concluded that leprosy causes a pattern of primary testicular failure (hypergonadotropic hypogonadism) that is highly related to disease duration, degree of testicular involvement, severity and recurrence of orchitis resulting from ENL, and early management and treatment compliance.

Conclusion

Leprosy is a pleiotropic disorder. It can involve many organs and can be confused with other diseases such as connective tissue disease. Herein, the case presents the importance of inquiry to diagnose leprosy patients, the difficulty in detecting this infectious disease, and sustainability of elimination. Therefore, we would like to emphasize the importance of high level of suspicion for disease diagnosis.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1 Walker SL, Lockwood DNJ. The clinical and immunological features of leprosy. Br Med Bull 2006; 77–78: 103–121.

2 Cole ST, Singh P. History and phylogeography of leprosy. In: Nunzi E, Massone C, editors. Leprosy. Milano: Springer Milan; 2012. pp. 3–11.

3 El-Gendy H, El-Gohary R, Shohdy K, Ragab G. Leprosy masquerading as systemic rheumatic diseases. J Clin Rheumatol 2016; 22:164–271.

4 Britton WJ, Lockwood DNJ. Leprosy. Lancet 2004; 363:1209–1219.

5 Nigam P, Mukhija RD, Gupta AK, Dayal SG, Goyal BM. Gonadal involvement in leprosy: study of gynaecomastia, testicular and epididymal involvement and therapeutic efficacy of indigenous drugs. Hansenol Int 1984; 9:10–20.

6 Bartt RE. Leprosy (Hansen's disease). Curr Treat Options Neurol 2004; 6:95–103.

7 Ridley DS, Jopling WH. Classification of leprosy according to immunity. A five-group system. Int J Lepr Other Mycobact Dis 1966; 34: 255–273.

8 de Oliveira Fernandes TRM, Korinfskin JP, Espíndola MMM, de Oliveira Corrêa LM. Arthritis and diagnosis of leprosy: a case report and review of the literature. An Bras Dermatol 2014; 89:323–325.

9 Khanolkat VR. Pathology in leprosy. In: Leprosy in theory and practice. 1964; pp. 131–132.

10 Ghorpade A, Ramanam C. Primary penile tuberculoid leprosy. Indian J Lepr 2000; 72:499–500.

11 Muir E. Lepra reaction and the general adaptation syndrome. Lep Rev 1962; 33:240–251.

12 Mahajan R, Dogra S, Kaur I, Yadav S, Saikia UN, Budania A. Type II reaction without erythema nodosum leprosum masquerading as lymphoma. Lepr Rev 2012; 83:378–383.

13 Marahatta S, Agrawal S, Paudyal P. Difficult diagnosis and challenging treatment – a report on leprosy. Lepr Rev 2016; 87:543–547.

14 Tilak T. Acute epididymo-orchitis in lepromatous leprosy. Lep Rev 1968; 39:31–36.

15 Faten A, Seham A, Abeer M, Asmaa S, Omima M. Testicular function in male patients with lepromatous leprosy. Egypt J Dermatol Venereol 2014; 34:41–45.