Case Report

Lepromatous Hansen's Disease Simulating Still's Disease: A Parodying Case Report with Diagnostic Conundrum.

Safa Patrick[1], Sumit Kar[2], Nitin Gangane[3], Abhay Deshmukh[4], Ajinkya Sawant[5], Pooja Manwar[6], Subhor Nandwani[7]

1. Senior Resident, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
2. Professor and Head, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
3. Professor, Department of Pathology, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
4. Assisstant professor, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
5. Senior Resident, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
6. Resident, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
7. Resident, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.

Abstract

Leprosy, caused by Mycobacterium leprae, is an ancient chronic granulomatous infectious disease which is known to affect human beings and continues to be an important prevalent disease in several developing nations. Since leprosy mimics connective tissue disease, several case reports have mentioned that it mimics common rheumatologic diseases like rheumatoid arthritis (RA), seronegative spondyloarthritis (SpA) and systemic lupus erythematosus (SLE). After extensive literature search we failed to find case report of leprosy with type two reaction simulating still's disease. Hence we hereby report a case of an 18 year old male with lepromatous Hansen's disease with type 2 reaction misdiagnosed and mismanaged as Still's disease.

Keywords: Lepromatous Hansen's Disease, Still's Disease, Erythematous Nodules

Introduction

Leprosy is an ancient chronic granulomatous infectious disease which is known to affect human beings and continues to be an important prevalent disease in several developing nations.[1] Mycobacterium leprae is the causative organism for leprosy. It is globally distributed; however, it is especially prevalent in developing countries in Asia, Africa, and Central/South America, while it is quite rare in developed countries.[2] Since leprosy mimics connective tissue disease ,several case reports have mentioned that it mimics common rheumatologic diseases like rheumatoid arthritis (RA), seronegative spondyloarthritis (SpA) and systemic lupus erythematosus (SLE).[3-4] It is also known to present with clinical and laboratorial findings that mimic autoimmune connective tissue diseases and vasculitis.[5] One to five percent of leprosy patients are reported to develop arthritis of small joints of hands and feet akin to that seen in rheumatoid arthritis.[1] We hereby report a case in which leprosy presented with features similar and thus initially diagnosed and treated as still's disease. In areas endemic for leprosy it is imperative to keep in mind the myriad presentations of leprosy including its musculoskeletal manifestations, as consequences of misdiagnosis, mistreatment can be catastrophic, especially in the era of increasing use of biologic disease modifying agents.[6]

Corresponding author
Sumit Kar, MBBS, MD, Professor and Head, Dept. of Dermatology, Venereology and Leprosy, Mahatma Gandhi Institute of Medical Sciences, Sewagram, Maharashtra, 442102, India.
Email: karmgims@gmail.com
Received: 14 January 2022 Accepted: 22 January 2022 Available Online: 30 January 2022

Cite this Article:
Patrick S, Kar S, Gangane N, Deshmukh A, Sawant A, et al. Lepromatous Hansen's Disease Simulating Still's Disease: A Parodying Case Report with Diagnostic Conundrum. J Ban Acad Dermatol. 2022; 2 (1): 41-43

Available at: www.jbadbd.com
An official publication of Bangladesh Academy of Dermatology (B.A.D.)

Case Report :

An 18 year old male presented with multiple red colored nodules to plaques with erosions on few lesions of both upper and lower limb of size 1x2 cm to 2x3 cm on and off since 6 years and Joint pain on and off since 6 years. On detailed history patient had multiple joint pain involving large and small joints with high grade fever with multiple red colored raised lesions over bilateral upper limb and lower limb (Figure1). Patient was on treatment from rheumatologist diagnosing him to have Still's disease and accordingly he was treated with high dose corticosteroids during the period of exacerbation, which would give symptomatic relieve followed by reappearance of symptoms when patients is off steroids. Due to long term steroid use patient developed cushingoid facies and hypertension. He was also given disease modifying agents like methotrexate and mycophenolate mofetil. Disease severely affected the quality of life and not able to do his daily activities due to pain. On Examination submental, bilateral inguinal and right axillary lymph nodes were enlarged and with bilateral edematous hands. Pateint also had hepatospleenomegaly. On cutaneous examination erythematous nodules to plaques with few erosions was present over bilateral upper limb and bilateral lower limb. Multiple hyperpigmented macules were also present over bilateral upper limb and lower limb. Nerve examination revealed bilateral ulnar, superficial radiocutaneous and common peroneal nerve thickening with neuritis. Hence differential diagnoses were Hansen's disease with Type 2 lepra reaction and Still's disease. Routine investigations revealed anaemia, leucocytosis and raised liver enzymes. C reactive protein was positive. Slit skin smear for AFB was Positive. Diagnosis of Hansen's disease with type 2 reaction was further confirmed on histopathology and Fite-Faraco staining. Microscopically, H&E stained section of skin biopsy showed presence of unremarkable epidermis. Dermis shows presence of clusters of macrophages. These macrophages showed abundant pink to pale cytoplasm which corresponds to numerous intracellular organisms. There is also presence of diffuse lymphocytic infiltrate (Figure 2). Fite-Faraco stained sections showed presence of large aggregates of red coloured leprosy bacilli within the macrophages (Figure 3). Multibacillary antileprosy treatment, corticosteroids and thalidomide for type 2 reaction was started. Patient showed excellent response to treatment.

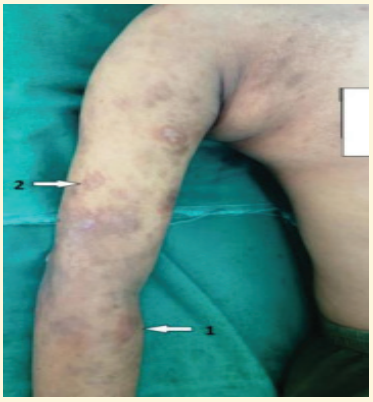

Figure 1: Multiple erythematous nodules (arrow no. 1) with multiple post inflammatory hyperpigmented patches seen (arrow no. 2)

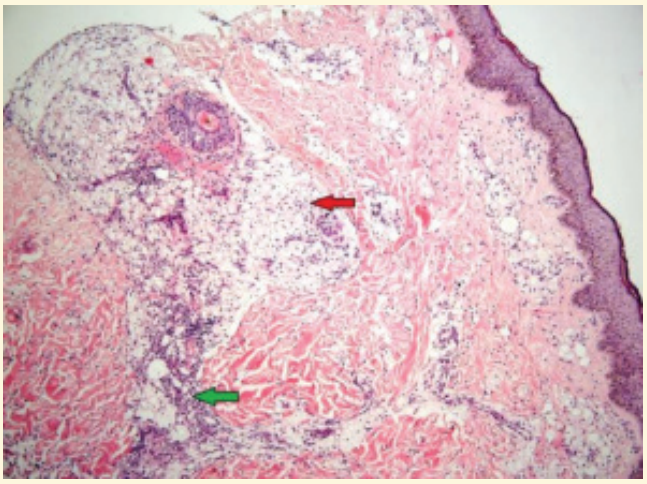

Figure 2: Large clusters of macrophages (red coloured arrow) within the dermis along with lymphocytic infiltrate (green coloured arrow) (H&E, x400).

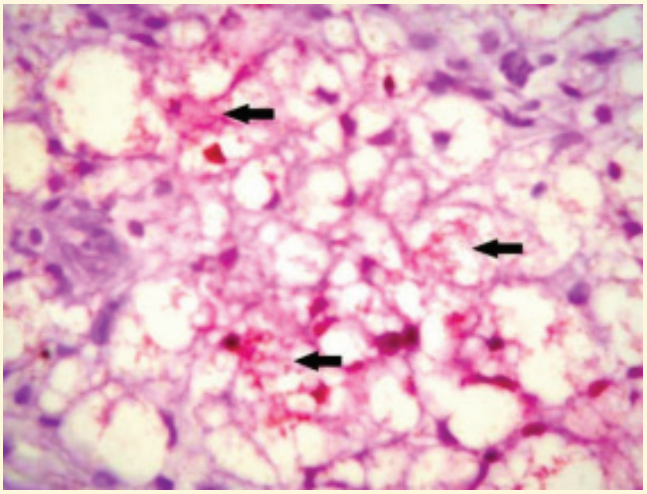

Figure 3: Large aggregates of red coloured leprosy bacilli (black coloured arrows) within the macrophages (Fite-faraco, x1000).

Discussion

Leprosy displays a wide spectrum of clinical and histological manifestation, reflecting the broad range of cellular immune response to Mycobacterium leprae. The disease spectrum ranges from a form with a strong immune response and fewer organisms as in case of tuberculoid or paucibacillary to a form with a weaker immune response and higher organisms as in case of lepromatous or multibacillary.[6] Articular involvement in leprosy is the third most frequent manifestation, after dermatological and neurological involvement.[2] Stills disease present with the skin lesions which are evanescent, salmon colored, macular or maculopapular cutaneous eruption, usually non pruritic in nature, occurs along with fever and disappears during a febrile period. The rash predominantly involves trunk, extremities but can also involve palms, soles and occasionally the face. Leprosy patients have various forms of skin lesions such as macules, papules, nodules, plaques, diffuse infiltration, and ulcers. Patients of leprosy have a number of auto antibodies which add to the confusion. These include ASO, RF, ANA, ANCA and even antiphospholipid antibodies, ds-DNA, and anti-CCP and C reactive protein.[3] Our patient had multiple erythematous nodules to plaques with fever, lymphadenopathy, anaemia, leucocytosis and positive C reactive protein which is common for both the diseases. In our case, histopathological and Fite Faraco staining helped us to confirm the diagnosis of Hansen's disease. Multibacillary leprosy may be initially misdiagnosed as Still's disease due to sharing of similar clinical manifestations. Hansen's disease is a great mimic and needs an astute clinical sense with a high index of clinical suspicion to capture it early enough for an effective and complete cure.[1] Moreover, this case report is aimed to create awareness for possibility of leprosy patient visiting rheumatology out-patient from endemic areas and getting misdiagnosed. We conclude that an unexplained high grade fever accompanied by skin rash from endemic areas should be actively ruled out clinically and if necessary with investigations for excluding mycobacterium infection before diagnosing, a rare, diagnosis of exclusion autoimmune disorder like Still's disease to save patients from needless anti-rheumatic treatment

References

1. Salvi S, Chopra A Leprosy in a rheumatology setting: a challenging mimic to expose. Clinical rheumatology. 2013 Oct 1;32(10):1557-63.
2. Hou Z, Lin L, Gong Y, Wang D, Xiao Z Leprosy mimicking connective tissue disease-a challenge for rheumatologists. Int J Clin Exp Med. 2016 Jan 1;9(9):18040-6.
3. Rath D, Bhargava S, Kundu BK Leprosy mimicking common rheumatologic entities: a trial for the clinician in the era of biologics. Case reports in Rheumatology. 2014;2014.
4. Horta-Baas G, Hernandez-Cabrera MF, Barile-Fabris LA, del S Romero-Figueroa M, Arenas Guzmán R Multibacillary leprosy mimicking systemic lupus erythematosus: case report and literature review. Lupus. 2015 Sep;24(10):1095-102.
5. Pandhi D, Chhabra N. New insights in the pathogenesis of type 1 and type 2 lepra reaction. Indian Journal of Dermatology, Venereology, and Leprology. 2013 Nov 1;79(6):739.
6. Silva EA, Iyer A, Ura S, Lauris JR, Naafs B, Das PK, Vilani-Moreno F Utility of measuring serum levels of anti-PGL-I antibody, neopterin and C-reactive protein in monitoring leprosy patients during multi-drug treatment and reactions. Tropical Medicine & International Health. 2007 Dec;12(12):1450-8