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LUCIO PHENOMENON AND DIFFUSE NON NODULAR LEPROMATOUS LEPROSY – AN UNCOMMON OCCURANCE IN INDIA

Dermatology

Dr. Akshika Mukhija Resident, Department of Dermatology, SAMC and PG Institute, Indore, Madhya Pradesh, India.

Dr. Kanchan Kumawat* Resident, Department of Dermatology, SAMC and PG Institute, Indore, Madhya Pradesh, India. *Corresponding Author

Dr. Chaitanya Namdeo Professor, Department of Dermatology, SAMC and PG Institute, Indore, Madhya Pradesh, India.

Dr. Oshin Jain Resident, Department of Dermatology, SAMC and PG Institute, Indore, Madhya Pradesh, India.

ABSTRACT

Lucio's phenomenon is a rare manifestation of untreated Leprosy. Patient usually presents late due to absence of constitutional symptoms. Its diagnosis is further confused with other diseases like Vasculitis which also presents with vasculonecrotic skin lesions. We present a case of Lucio Phenomenon in a previously undiagnosed non-nodular diffuse Lepromatous Leprosy.

KEYWORDS

Leprosy, Lucio phenomenon

INTRODUCTION:

Leprosy is a chronic infection caused by *Mycobacterium leprae which affects most commonly s*kin, nasal mucosa and peripheral nerves. In India current prevalence rate of leprosy was 0.66 per 10,000 population in 2016.[1] The clinical course of disease depends upon the immune response of the host which varies from Tuberculoid Leprosy to Lepromatous Leprosy.[2] Leprosy Reactions are immunologically mediated episodes of acute or subacute inflammation which interrupt, the relatively uneventful usual chronic course of disease. Generally two types of Lepra reactions are seen, Type 1 Lepra reaction showing type 4 hypersensitivity and Type 2 Lepra Reaction showing type 3 hypersensitivity.[3] Lucio Phenomenon is a special type of reaction observed in uniformly diffuse shiny infiltrative non-nodular form of pure Lepromatous Leprosy (LL) or Borderline Lepromatous Leprosy (BL).[4,5] Non nodular LL is also known as Lucio Leprosy, which is seen only in untreated cases of Leprosy and chiefly encountered in Mexicans.[6,7] Few cases have also been reported from other countries like Costa Rica, USA, Hawaii, Brazil and recently from India too.[8,9] It was first described by Lucio and Alvarado in 1852 in Mexico. Lucio's phenomenon was named in 1948 by Latapí and Zamoro.[10] Here we present a case who presented with large cutaneous infarcts which on first impression was diagnosed as vasculitic but subsequently were proved to be Lucio phenomenon.

Case presentation

A 58 year old South East Asian (Indian) male from a poor socio-economic class presented with spontaneous ulceration with dark crusted and ecchymotic lesions involving the extremities, supraorbital region, pinna and scrotum along with arthralgia, fever and chills for 15 days. Additionally, he complained of numbness and tingling over extremities with progressive loss of eyelashes and eyebrow during the preceding one year. He also had two episodes of epistaxis with nasal stuffiness. Cutaneous examination revealed multiple reticular dark violaceous purpura with angular, ragged margins and scabs over the trunk, gluteal regions and legs. Cilliary and supracilliary madarosis was present. Hyperpigmented crusted plaques were present over supraorbital margin, pinna, nipples, elbows, fingers, toes, penile shaft and scrotum. There were two painless ulcers over calcaneum and medial maleolus with undermined margins with non-foul smelling serosanguineous discharge.[FIGURE-1]

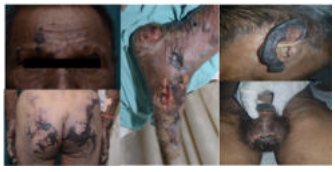

Figure-1: Cutaneous Features Of Patient

On neurological examination there was symmetrical, non-tender and non-nodular thickening of multiple nerves including transverse cervical, supraclavicular, radial and ulnar nerves of both sides. Patient was able to perform all motor functions but there was complete sensory loss over extremities showing glove and stocking anesthesia upto shoulders in upper limbs and upto knees in lower limbs.

A differential diagnosis of LL disease with Lucio Phenomenon, vasculitis and cryoglobinemeas were kept. Baseline investigations were done including CBC, LFT, RFT, Chest X-ray and Urine examination; which were within normal limits. D-dimer value was raised and cryoglobulin levels were within normal limits. A slit– skin smear examination of the eyebrows and earlobes demonstrated a bacterial index (BI) of 6+.[FIGURE-2]

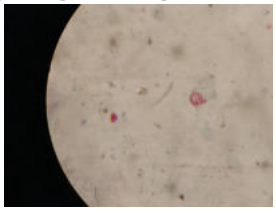

Figure-2: Bi = 6+ [acid Fast Bacilli (afb) Seen In Clusters]

Skin biopsy was performed which showed epidermal thinning, orthokeratosis, parakeratosis and perivascular & periadnexal infiltrate of histiocytes, foamy macrophages and few lymphocytes, extravasation of RBCs was also seen.[FIGURE-3a,b,c] Fite Faraco staining showed numerous AFBs in perivascular endothelial cells giving impression of Lucio Phenomenon.[FIGURE- 4a,b,c]

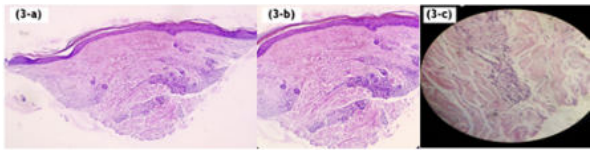

Figure-3 -histopathology Pictures – (a- Scanner View, B- 40x, C-100x)

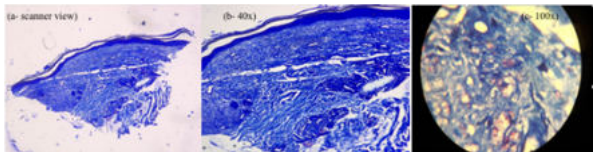

Figure-4 - Fite Faraco Staining (a- Scanner View, B- 40x, C- 100x)

DISCUSSION:

Type 1 Lepra reaction occurs with Tuberculoid Leprosy where cell mediated immunity (CMI) robust and characteristically presents as inflammation of pre-existing lesions or appearance of new lesions. Type 2 Lepra reaction occurs with Lepromatous or Borderline Leprosy due to weak CMI and high bacillary load, generally presenting as Erythema Nodosun Leprosum (ENL). Vasculonecrotic lesions in Leprosy can be seen either with Lucio phenomenon or with vasculonecrotic ENL. Lucio pchenomenon is a rare type of lepra reaction which manifests as bullae formation, necrotic areas followed by eschar formation and ulceration.[11,12] Presence of cutaneous infarcts without presence of any blisters or ulcers are unusual, as reported by Magaña et al[11]in only 3 out of 12 patients with Lucio Phenomenon.

Lesions usually begin as erythematous painful patches over extremities; which evolve into purpuric lesions, become necrotic and further heal in about 2–4 weeks with superficial, atrophic scars with minimal constitutional symptoms and tends to disappear in 6–8 weeks after initiation of treatment or it may develop reaction, mainly of ENL type while on treatment.[7,13] All the above features were seen in our patient and a good response was seen after starting MBMDT.

Histopathology shows infiltration of AFB (both solid and fragmented) within macrophages and endothelial cells of vessel walls involving small and medium-sized vessels; were seen in our patient.[11] Medina reaction using lepromin test could not be elicited due to nonavailability.

Lucio phenomenon ususally overshadows the diagnosis of leprosy as its features mimics Connective Tissue Disorders, Vasculitis or Cryoglobulinemias.[14,15] It is seen in untreated cases of LL Disese where the bacillary load increases to invade the endothelium of the blood vessels thus giving it a vasculitic picture clinically as well as histopathologically.

Conclusion: Lucio Phenomenon being a rare presentation is scarcely reported from Indian subcontinent. It is important for the clinician to differentiate Leprosy from other presentations of Cutaneous Vasculitis as the former is eminently curable with antibiotics and prudent use of immunosuppressive agents. We should always keep infectious etiologies in the differential diagnosis of vasculitis as the treatment for the two is dramatically different and inappropriate immunosuppr ession alone can be disastrous in the context of infection. When in doubt, a skin biopsy often helps to get the final diagnosis.

Conflict of Interests: none

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