ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 9, Issue – 5, September – 2024 , Page No. : 144 – 147

De Novo Histoid Leprosy - A Rare Variant of Lepromatous Leprosy

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Citation this Article: Dr. Nikunj Rameshbhai Bhalsod, Dr. Divya Shetty, Dr. Anita Sharan, "De Novo Histoid Leprosy - A Rare Variant of Lepromatous Leprosy", IJMSIR - September - 2024, Vol – 9, Issue - 5, P. No. 144 – 147.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Histoid leprosy (HL) is a rare variant of lepromatous leprosy (LL) which is a highly transmissible disease. Here, we describe a de novo case of histoid leprosy. This variant typically arises in multibacillary patients who have received irregular or insufficient treatment for lepromatous leprosy. Its distinctive clinical presentation makes diagnosis challenging, potentially delaying treatment and contributing to the persistence of the disease as an endemic condition.

So biopsy and histopathological examination are essential for the accurate diagnosis of histoid leprosy.

Keywords: Leprosy, Histoid Leprosy, Mycobacterium leprae, Communicable disease.

Introduction

Leprosy is a chronic granulomatous infectious disease caused by Mycobacterium leprae, an acid-fast, rod-shaped bacterium that has afflicted humans for centuries. It primarily affects the skin and peripheral nerves in most cases. [1] The term histoid leprosy was first described by Wade in 1963. He observed it in patients undergoing long-term dapsone monotherapy and linked its occurrence to drug resistance. Histologically involves bacillary-rich lepromas composed of spindle-shaped cells, with the notable absence of globus formation, which is typically seen in ordinary lepromas and it exhibits a fibromatoid tendency in a chronic form. [2,4] However, de novo cases have also been reported.

The clinical appearance of histoid leprosy (HL) includes smooth, shiny, and firm dome-shaped papules, nodules, and plaques emerging from apparently normal-looking skin. [2]

We reported a de novo case of histoid leprosy as a rare variant of lepromatous leprosy in the post-global leprosy elimination era.

Case Report

A 38-year-old male patient presented with a skin-colored lesion over the upper limb and trunk region since 3 months associated with itching and burning sensation. The patient had a history of epistaxis 1 year back. There was no history of fever, weakness, or fluid draining from the lesions.

On examination, there were diffuse erythema and infiltration plaques seen over the face with conjunctivitis. Madarosis was present bilaterally. Multiple well-defined erythematous to copper-colored papules and nodules

were present over the bilateral pinna, face, bilateral upper limb, and trunk region. Sensory nerve examination revealed decreased fine and cold touch sensation over bilateral feet. Motor nerve examination was normal. (Fig. 1, 2A, B)

Figure 1: Multiple erythematous papules and nodules over the face.

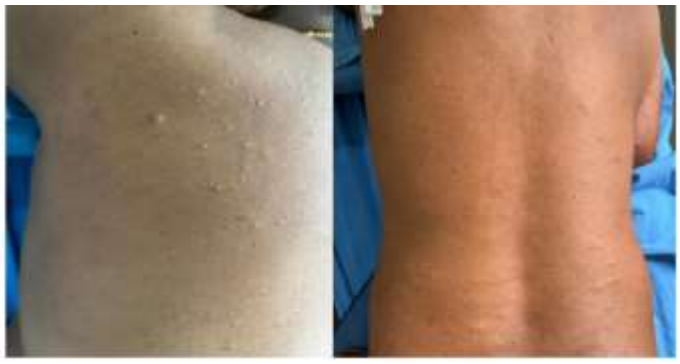

Figure 2 A, B: Multiple skin-colored papules and nodular lesions over the back region

We received a 4 mm punch biopsy specimen from the lesion over the back for histopathological examination.

Standard tissue processing protocol was followed.

Hematoxylin and Eosin (H&E) slides were evaluated and a special stain Fite Faraco was done for this case.

Microscopically, the epidermis was atrophic. the dermo-epidermal junction showed a Grenz zone.

The dermis showed patchy spindle cell proliferation arranged in vague fascicles and admixed with foamy macrophages (Lepra cells).

Trapping of appendageal structures and neural tissue was identified with mild chronic inflammatory cell infiltrate. Similar cells were also seen in perivascular locations.

There was no evidence of granuloma formation, well-defined nodules, fungal organisms, atypia, or malignancy in the sections studied. (Fig. no 3, 4, 5)

Fite Faraco stain was positive for lepra bacilli and the bacillary index was 6 (>1000 bacilli/hpf) Predominently solid stained bacilli were noted. The lepra bacilli were also seen within the appendageal structures. (Fig. no 4, 5)

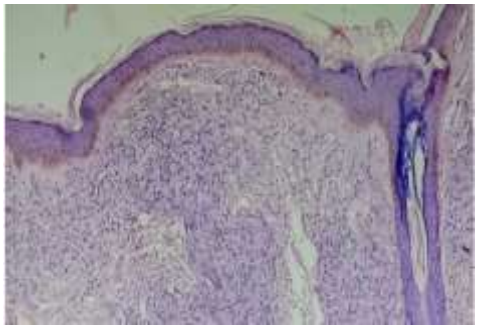

Figure 3: Atropic epidermis and grenz zone (H&E 100X)

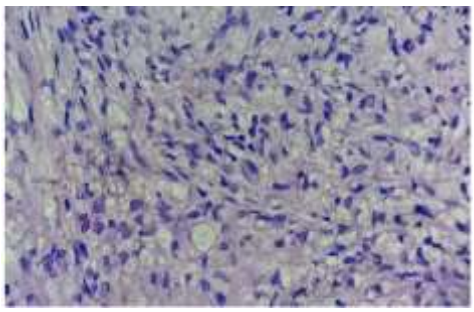

Figure 4: Spindle cell proliferation with foamy macrophages (H&E 400X)

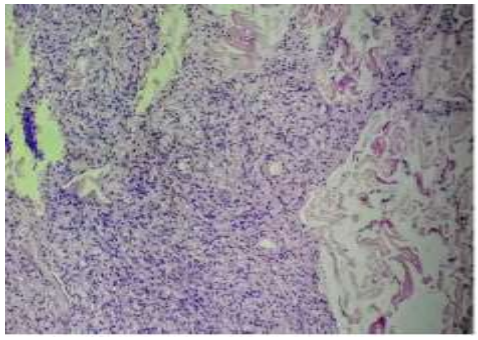

Figure 5: Perivascular inflammatory infiltrate (H&E 100X)

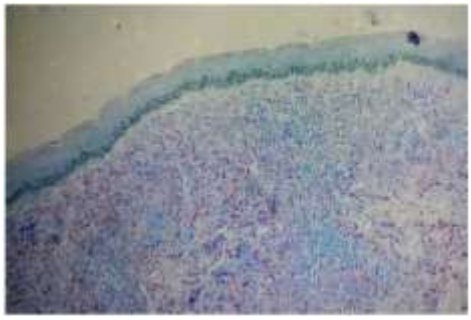

Figure 6: Positive Fite Faraco stain (100X)

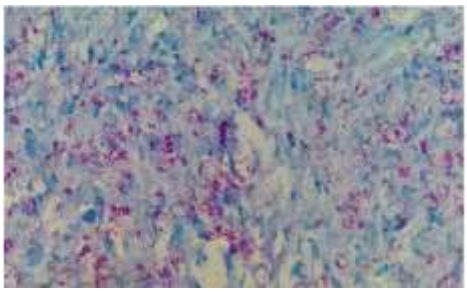

Figure 7: Rod-shaped Lepra bacilli with high bacillary index (400X)

Discussion

The clinical-histopathological spectrum of leprosy, based on the Ridley-Jopling classification, reflects the interaction between Mycobacterium leprae and the host's cell-mediated immunity. This classification includes five main presentations: polar tuberculoid (TT), borderline tuberculoid (BT), borderline borderline (BB), borderline lepromatous (BL), and polar lepromatous (LL). Indeterminate leprosy (I) and pure neuritic leprosy are not included in this spectrum classification.[1]

Histoid leprosy is a rare variant of lepromatous leprosy affecting 1.1-3.6% among total leprosy patients with male preponderance and average age at diagnosis ranging between 21 to 40 years.[1,4]

The pathogenesis of histoid leprosy remains unclear, but it is known that both cell-mediated and humoral immune responses against Mycobacterium leprae are more pronounced in histoid leprosy compared to classical lepromatous leprosy (LL). This heightened response includes increased expression of cluster of differentiation 36 (CD36) by keratinocytes, along with elevated levels of CD4 T lymphocytes, B lymphocytes, and immunoglobulins. However, despite this enhanced immune response, macrophages appear to lack the functionality to effectively kill M. leprae, which leads to a high bacterial load in histoid lesions.[1]

The histopathology of histoid leprosy includes three distinct patterns: pure fusocellular, fusocellular with an epithelioid component, and fusocellular with vacuolated cells. [3]

Classical histopathological findings of histoid leprosy include epidermal atrophy due to dermal expansion by the underlying leproma and the presence of an acellular band (Grenz zone) just beneath the epidermis. The leproma itself is composed of fusiform histiocytes arranged in a whorled, crisscross, or storiform pattern, containing numerous acid-fast bacilli. [4]

Unlike in lepromatous leprosy, the bacilli in histoid leprosy do not form globi. Histoid leprosy is often referred to as a "great masquerader" due to its variable clinical presentation. The high bacillary load in these patients makes them a significant reservoir for leprosy transmission if diagnosis is delayed. [5]

Conclusion

Histoid leprosy is a rare variant of lepromatous leprosy (LL) that should be considered a significant differential diagnosis in endemic regions for patients presenting with skin nodules. A histopathological examination is essential for a definitive diagnosis. Although histoid leprosy represents a small percentage of all leprosy cases, early diagnosis and timely treatment are critical to reduce the impact of these high-bacillary reservoirs on disease transmission and the global burden of leprosy.

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