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SHORT COMMUNICATION

Oculodermal Melancytosis, the Nevus of Ota Histopathology Highlights

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Abstract

Oculodermal Melancytosis/ nevus of Ota a rare congenital disorder has intrigued, because of its cardinal clinical connotation in addition to histopathology highlights, emphasizing sparing of the epidermis, the blunt of disease is borne by the dermis evidence as prominent sebaceous lobules of the facial skin in addition to many slender, dendritic, pigmented melanocytes arranged horizontally between / entrapped amongst the collagen bundles in the mid-dermis. Melanophages are either few or none, reiterating the classification once mooted for management purposes. However, eye examination is mandatory.

Introduction

Oculodermal melancytosis, the Nevus of Ota has always been a fascinating overture ever since its first description [1]. It is a rare congenital disorder, and as well celebrated clinical manifestations apparent primarily as grayish white or blue coloration of the skin like of which is also affecting the sclera of the eye, occasionally oral mucosa [2]. It is largely asymptomatic in the beginning, but may result in malignant degeneration, should it be associated with a cellular blue nevus [3]. Besides, glaucoma [4] may be outcome of long standing disease. It is, therefore, worthwhile to form work up of these cases oculodermal melancytosis, emphasizing periodic checkups to obviate the emergence of possible complications.

Case Report

A 24-year-old-women of Indian origin had reported with an asymptomatic gray/slate blue color pigmentation inflicting right side of face and the eye ever since birth, the extent of pigmentation has been increasing and had corresponded to that of age. However, it was perceived has a huge cosmetic concern, impelling are parents to seek the advice of specialist in the field.

Skin surface examination was conspicuous, and had variable size gray/slate white macules the margin of which was merging imperceptibly into the adjoining skin, occupying the malar area just below the right eyes. Macules or / spots were also located on the sclera of the eye (Figure 1). The distribution of lesions were unilateral conforming to ophthalmic and maxillary branches of the trigeminal nerve.

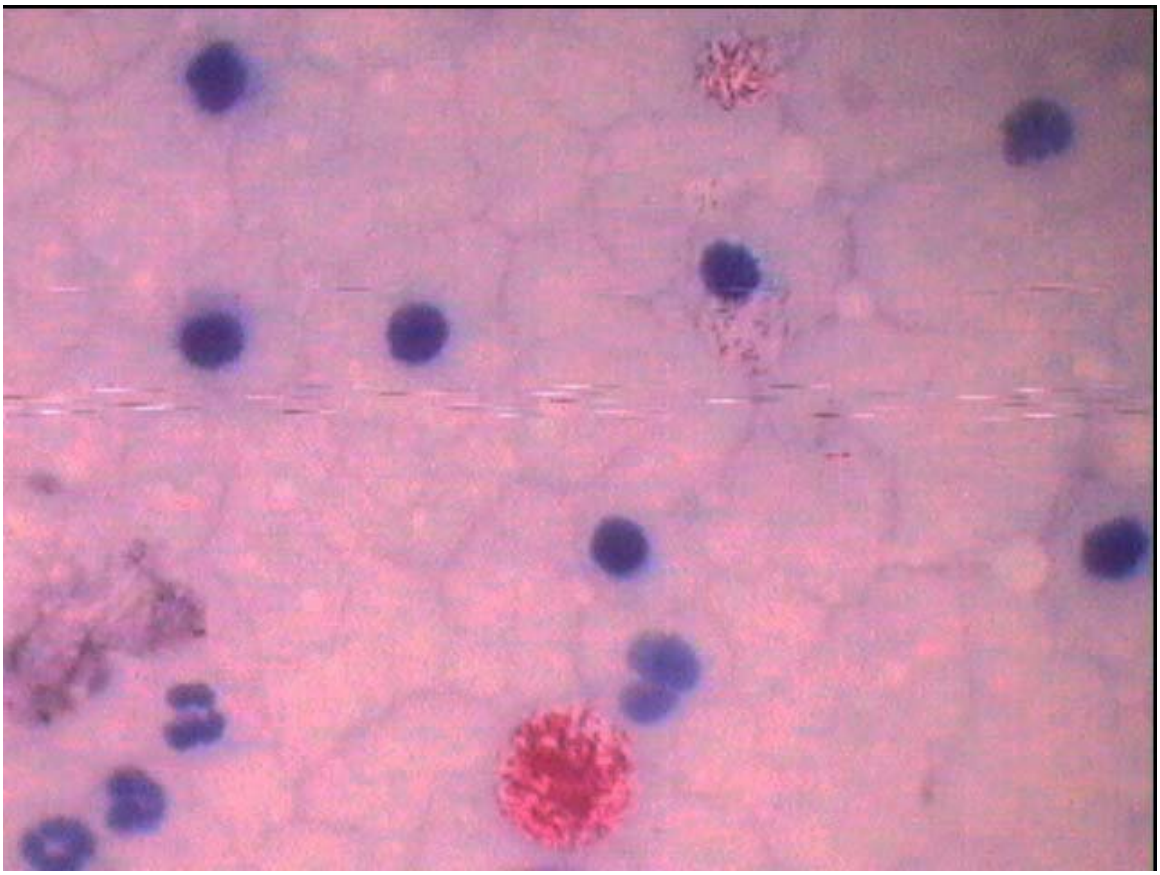


Figure 1: M. Laprae in slit skin smear.

causative agent Mycobacterium leprae. It encompasses borderline lepromatous, midborderline, and lepromatous leprosy [3].

Mode of Transmission

- Air-borne transmission: through inhalation of the bacilli
- Indirect transmission: Clothes and sleeping mates
- Direct prolong and intimate skin-to-skin contact

Pathogenesis of leprosy

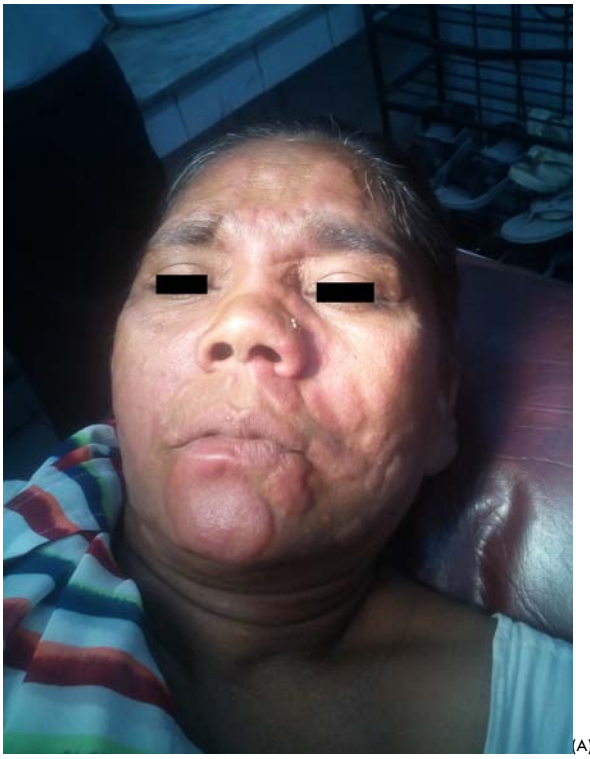
M. leprae genes help to define the minimal gene set necessary for in vivo survival of this mycobacterial pathogen as well as genes potentially required for infection and pathogenesis [4] seen in leprosy.

Furthermore, M. leprae has a predilection to invade regenerating and / or degenerating branches of peripheral nerve undergoing inadvertent injury or trauma, seem to be dictated by multiple signaling pathways, playing key regulatory roles during the development of peripheral nervous system (PNS) and also in neuro regeneration process following nerve degeneration.

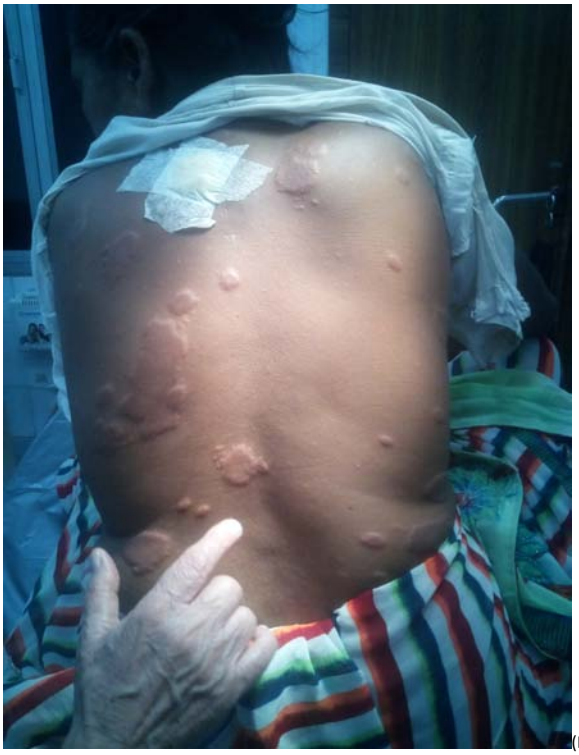
Schwann cells, the glial cells of the PNS, by interacting with neuronal (axonal) ligands, mainly neuregulins via Receptor Tyrosine Kinase (RTK) complex, ErbB2/ ErbB3, initiate intracellular signaling pathways to drive proliferation and differentiation of schwann cells, both during development and the process of regeneration and re-myelination after nerve injury. One of the major signaling kinases, extracellular signal-regulated kinase-1/2 (ERK1/2), that is also a downstream signaling pathway of neuregulin-ErbB2/ErbB3 activation, has been identified as a key regulator of Schwann cell proliferation, differentiation, demyelination and nerve regeneration.

Recent studies have provided evidence that the bacterium that causes human leprosy, Mycobacterium leprae that has a unique capacity to invade Schwann cells of the adult PNS, utilizes the neuregulin-ErbB2/ErbB3 associated signaling network to the bacterial advantage. M. leprae directly bind to ErbB2

on myelinated Schwann cells and activate the RTK by a novel route that bypasses the classical neuregulin/growth factor-induced ErbB2-ErbB3 heterodimerization, and subsequently induce downstream the canonical Erk1/2 signaling, leading to myelin breakdown and subsequent axonal damage.



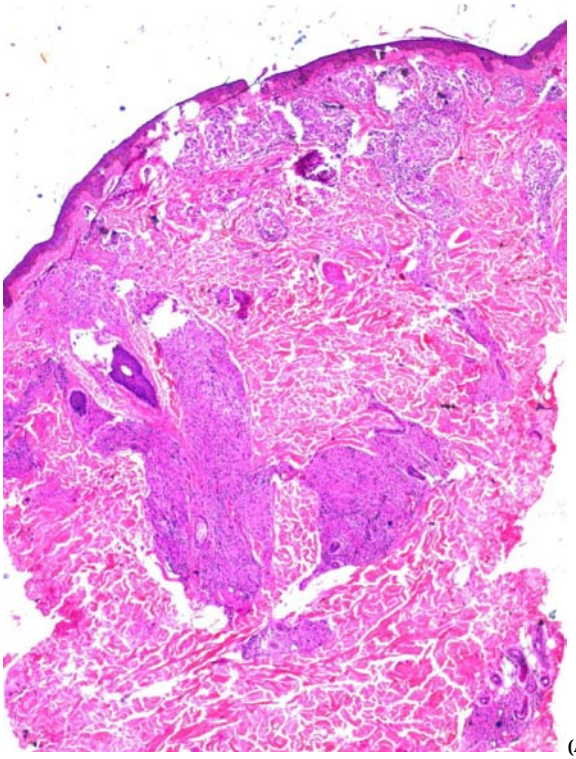
(A)



(B)

Figure 2 (A,B): Borderline borderline (BB) Leprosy, depicting an indurated plaque with serrated / irregular margin affecting the face and back of the truck in a woman, numerous but countable.

This initial injury provides a survival advantage for M. leprae as it induces de-differentiation and generates myelin-free cells, which are highly susceptible to M. leprae invasion and promote bacterial survival. Once invaded M.



(A)

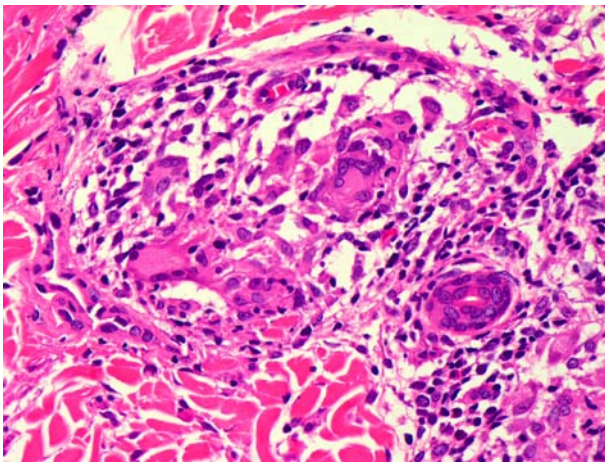


Figure 3 (A,B): The presence ill and well defined granuloma formed by epitheliod and lymphocytes in the dermis AFB are demonstrable in the granuloma of the dermis.

leprae activate Erk1/2 via a non-canonical pathway and subsequently increase the cell proliferation and maintain the infected cells in de-differentiated state, thereby preventing remyelination. Therefore, by subverting major RTKs and signaling pathways in adult schwann cells M. leprae appear to propagate the bacterial niche and maintain survival within the PNS [5].

Diagnosis of leprosy is largely clinical [6,7] (Figure 2 (a,b)) histopathology (Figure 3 (a,b) supplements not supplant [8-10] the diagnosis. Classification is an effective means of understanding and communicating a difficult disease. Leprosy is a disease with various manifestations and, therefore, difficult to understand without comprehensive classification. Accordingly, leprosy posses a continuous spectrum envisaging five [11] or seven [12] group.

Recapitulation of groups of leprosy based on the immunological and histopathological form the basis in addition to the clinical and bacteriological findings. The classification divides leprosy into five groups

- Tuberculoid tuberculoid (TT)
- Borderline tuberculoid (BT)
- Borderline borderline/ Midborderline (BB)
- Borderline lepromatous (BL)
- Lepromatous lepromatous (LL)

Seven group classification in addition has indeterminate and polyneuritic Besides, histoid leprosy emanating in consequence to emergence of dapsone resistant strain [13]. Five and seven groups are now being practiced field work, research in institution

Leprosy and Gender

There seems an intractable effect of leprosy vis-a-vis gender, which attracted the focused attention, and are considered worthwhile, for they may add to comprehensive understanding of this so-called factor. It is, therefore, worthwhile, to form a perception keeping in view co-laterals namely [14]

- Biological,
- Socio-cultural / economic and
- Service-related factors

Interestingly, biological factors in the countries such as; Indonesia Nigeria, Nepal and Brazil were found to be similar irrespective of the male/female ratio; more men than women were registered with multi-bacillary (MB) leprosy, socio-cultural factors explaining why women were under reporting. Yet, accessible, well reputed services augmented female participation and helped to diminish stigma, which in three out of the four societies seemed greater for women than for men. These positive effects could still be higher if the services would enhance community and patient education with active participation of patients and ex-patients themselves. Earlier, Similar gender leaning was observed in the across Indian sub-continent [15,16].

Multi-Drug Therapy [17] (MDT) comprising diaminodiphenyl sulfone (DDs) dapsone, rifampicin and clofazimine in recommended doses in the main stay of treatment for both pauci-multi bacillary leprosy and its elimination[18].

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