TROPICAL GROVE

Single lesion lepromatous leprosy in a teenager: an unexpected scenario

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Introduction: Hansen's disease (HD) is a chronic granulomatous disease principally affecting the skin and peripheral nervous system caused by *Mycobacterium leprae*. The incubation period varies from months to more than 30 years. The tuberculoid form of HD usually presents with a single hypoesthetic patch and skin biopsy shows epithelioid granulomas with absence of bacilli on Fite-Faraco stain. In contradistinction, lepromatous leprosy usually presents with numerous papules, plaques and nodules with induration of the ears and nose. Biopsy shows foamy granulomas with presence of acid-fast bacilli on Fite-Faraco stain.

Case summary: We present a case of a 13-year old female who presented with a 3-year history of a single hypoesthetic patch on the left knee. The initial clinical diagnosis was tuberculoid leprosy. However, histopathology revealed a Grenz zone, and a nodular granulomatous infiltrate consisting of epitheloid and foamy histiocytes with scattered lymphocytes. Fite-Faraco stain showed a bacillary index (BI) of 3+. Slit-skin smear revealed a BI of 4+. She was then started on multidrug therapy.

Conclusion: This case highlights the importance of slit-skin smear and biopsy as routine procedures in all new cases of suspected HD. These procedures will help differentiate multibacillary from paucibacillary forms of the disease which will influence decisions for treatment and prognostication. This case emphasizes that lepromatous leprosy may present with single lesions and may be misdiagnosed as paucibacillary leprosy if skin-slit smear and biopsy have not been done. This case further suggests that there are factors yet undetermined which play significant roles in determining the host response to *M. leprae* which are believed to influence morphology, configuration, number and distribution of skin lesions.

Keywords: Leprosy, Hansen's disease, Borderline Lepromatous, Tuberculoid, Leprosy, single lesion

INTRODUCTION

Hansen's disease is a chronic granulomatous disease principally affecting the skin and peripheral nervous system caused by Mycobacterium leprae. The incubation period varies from months to more than 30 years. Mycobacterium leprae is a non-cultivitable, gram-positive, obligate intracellular, acid fast bacillus. Ridley and his associates constructed a sixmember spectrum, ranging from high to low resistance starting from TT (polar tuberculoid, BT (borderline tuberculoid, BB (borderline), BL (borderline lepromatous) and LL (polar lepromatous). Tuberculoid leprosy usually presents with a sharply marginated hypoesthetic

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Source of funding: none Conflict of interest: none Corresponding author: Carmella Jane C. Luis Email: carmellajaneluis@gmail.com erythematous plaque, often annular secondary to peripheral propagation of bacilli. The central clearing demonstrates the tendency for spontaneous resolution. Biopsy would show epithelioid granulomas with multinucleated giant cells and absence of bacilli in Fite-Faraco stain. In lepromatous leprosy, the diminished immunity would permit unrestricted bacillary replication and widely disseminated, multiorgan disease. It would present with multiple sharply marginated erythematous papules or nodules, sometimes forming plaques. There is also exaggeration of the skin folds and nodule formation producing "leonine faces." Biopsy would show nodular infiltrate consisting of foamy or undifferentiated macrophages, with loss of appendages and presence of acid-fast bacilli in Fite-Faraco stain.

CASE REPORT

A 13-year old female presented with a 3-year history of a solitary erythematous patch on the left knee (Figure 1). Lesion gradually increased in size extending to the left thigh, now accompanied by hypoesthesia. No other lesions were



Figure 1. Solitary well-defined irregularly-shaped erythematous hypoesthetic plaque with elevated borders, left knee

noted over the entire body, with no perceived thickening of peripheral nerves. Father was previously treated for Hansen's Disease, lepromatous type with multidrug therapy for 2 years and was released from treatment. Physical examination revealed a solitary well-defined irregularlyshaped erythematous plaque on the left knee extending to the thigh. On caloric testing, there was no sensation felt upon application of cold compress and a 50% reduction in sensation upon application of a hot pack and pinprick test. There was complete absence of sensation on light touch.

Histopathologic findings were mild thinning of the epidermis and presence of a subepidermal grenz zone (Figure 2A). The dermis reveals a nodular granulomatous

inflammatory infiltrate of epitheloid histiocytes, scattered lymphocytes and some multinucleated giant cells. The infiltrate surrounds adnexal structures and nerves and extends to the deep dermis (Figure 2B). Modified Acid-fast stain revealed a BI of 3+ and was consistent with a slit-skin smear of 4+ (Figure 3).

Patient was started on Multidrug therapy consisting of Rifampicin, Clofazimine and Dapsone and is currently on her 6th blister pack with remarkable improvement.

DISCUSSION

A solitary lesion with high bacterial index is a rare occurrence and is considered as an unusual presentation. The clinical presentation of our patient was consistent with tuberculoid leprosy, however, based on histopathology and slit-skin smear patient was classified as borderline lepromatous type. This discrepancy between clinical and histological diagnosis is rare with crucial therapeutic implications. Moreover, any case of HD which does not conform to the current WHO clinical classification and which does not correlate with the expected histopathological findings remains a diagnostic challenge.

The exact pathogenesis of such unusual presentation is still unknown. The most common mode of transmission of leprosy is through respiratory droplets, however, there are reports of leprosy developing because of inoculation at the sites of trauma^{1,2}. Histological discrepancies between skin and nerve was reported by Kumar et al who observed a multibacillary nerve histology with paucibacillary clinical

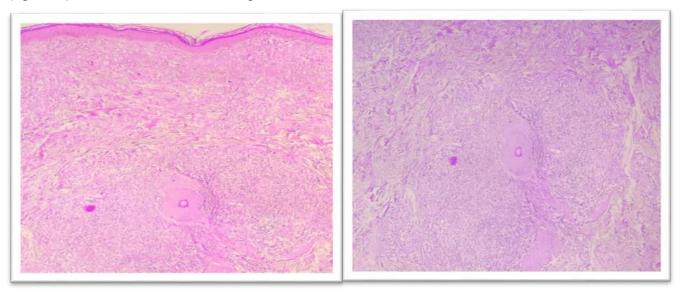


Figure 2. Histopathology shows a basket woven stratum corneum, thinning of epidermis and a subepidermal grenz zone (A) (H&E x100). The dermis reveals a granulomatous inflammatory infiltrate of epithelioid histiocytes, scattered lymphocytes and multinucleated giant cells (B) (H&E x400).

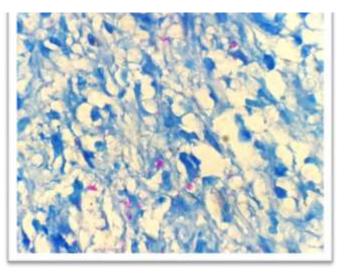


Figure 3. Modified Acid Fast stain shows numerous acid fast bacilli (Bacillary index = +3) (Fite Faraco stain x1000).

picture in 3 out of 27 patients. In such scenario, a biopsy with Fite Faraco stain, AFS and nerve biopsy are warranted to prevent the under treatment of multibacillary cases^{3,4}

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CONCLUSION

This case illustrates and emphasizes the importance of slit-skin smear and biopsy as routine in all new suspected cases of HD to help differentiate between multibacillary and paucibacillary forms. This case highlights that lepromatous leprosy may present with single lesions and misdiagnosed as paucibacillary leprosy if skin-slit smear and biopsy have not been performed. This case further suggests that there are factors yet undetermined which play significant roles in determining the host response *to M. leprae* which are believed to influence morphology, configuration, number and distribution of skin lesions.