

CASE REPORT

Lepromatous leprosy: Presenting as erythema nodosum leprosum

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INTRODUCTION

Leprosy is a chronic granulomatous disease principally affecting the skin and peripheral nervous system. It exhibits a wide spectrum of presentation. Erythema nodosum leprosum (ENL) is an inflammatory complication of leprosy that results in painful skin lesions on the arms, legs and face. Despite the fact that ENL mostly occurs later during the course of treatment; it may be the first manifestation of leprosy in untreated cases of very long duration.¹

CASE REPORT

A 25 year-old housemaid Indonesian woman, has come to Kuwait for the first time since about 2 months. Three weeks ago, she presented to Jahra Hospital with fever and skin lesions involving both lower limbs and was diagnosed by a surgeon as having cellulitis and treated with different types of antibiotics without any improvement. Two weeks later, she developed tender erythematous edematous nodules and plaques symmetrically distributed over the face and the extensor surfaces of upper extremities, with few lesions on neck and trunk. Lesions on the face involved the mandible, cheeks, eye lids and perorbital skin, giving the

patient a characteristic leonine appearance (Fig 1). There was partial hair loss of the outer third of the eyebrows (madarosis) (Fig 2). Prominent ichthyosis was detected on both legs (Fig 3). Peripheral nerves; ulnar and median nerves, were thickened and tender, however, sensory and motor examination was normal.



Fig. 1 Infiltrated plaques on the face

Beside fever, the patient was also suffering from malaise, anorexia, conjunctivitis, iritis, muscle pain and arthralgia. History of previous attack of

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Fig. 2 Prominent plaques and partial madarosis



Fig. 3 Prominent ichthyosis on both legs

epistaxis was found to be positive. However, the patient denied any history of similar skin lesions, or family history of Leprosy.

Laboratory investigations revealed elevated ESR and moderate leucocytosis. Urine and other hematological investigations were within normal level. A biopsy was taken from one of the nodules on the left forearm. Histological examination showed diffuse inflammatory infiltrate formed mainly from foamy macrophages (Fig 4), mild

lymphocytic infiltrate, and leukocytoclastic vasculitis; features consistent with ENL. Fite stain revealed a large number of acid-fast leproae bacilli (Fig 5). Slit-skin smear for acid-fast bacilli from the nodules revealed a BI of 4+ and MI of 2 percent.

Treatment was initiated including; prednisolone 40 mg daily plus clofazimine 50 mg, and aspirin. However, the patient has traveled back to her country.

DISCUSSION

Leprosy is a slowly progressive infectious disease caused by *Mycobacterium leprae* (*M.leprae*) which is an acid fast bacillus, first identified in 1873 by the Norwegian physician, Gerhard Henrik Armauer Hansen. It is highly infective with low pathogenicity and virulence, and has a long incubation period.²

Though seldom lethal, the disease causes appreciable morbidity principally through its effects on the skin, peripheral nerves, and nasal mucosa.³ *M. leprae* prefer the cooler parts of the body sparing warm areas like; groin, perineum, scalp, axilla, and the narrow zone of lumbosacral region which are considered to be the immune zones in leprosy.⁴

Leprosy is primarily endemic in subtropical regions of the world, which include Brazil, India, Madagascar, Mozambique, Myanmar, and Nepal.⁵ However, due to international travel, leprosy can be found anywhere in the world.

In 1966, Ridley and Jopling created a classification of leprosy, based on the immunologic response of

the host to *M. leprae*, into a five-group system: TT (polar tuberculoid), BT (borderline tuberculoid), BB (borderline), BL (borderline lepromatous), and LL (polar lepromatous).⁶ In 1982, the WHO recommended the classification of all patients be based on Ridley-Joplin classification and the estimated bacterial load in skin-slit smears. The WHO classifies patients as paucibacillary (PB) if there are five or fewer skin lesions or multibacillary (MB) if there are six or more skin lesions, or if slit skin smear is positive. These classification systems are utilized in determining treatment regimens and predicting clinical outcome in patients.⁷

Lepromatous leprosy has the greatest number of bacilli and is known to cause widespread involvement of the skin attributed to hematogenous dissemination of the organisms. Skin lesions include ill-defined, erythematous or hypopigmented macules, papules, nodules and plaques symmetrically distributed over the face, trunk, and extremities. Later findings include facial infiltration (leonine facies), ocular effects, neurologic involvement, and hypogonadism.⁸

About 50 % of lepromatous leprosy and 25 % of borderline lepromatous leprosy patients experience an ENL reaction, which is an antigen antibody immune complex reaction (Type III humoral hypersensitivity). Increased cell-mediated immunity may play a role in the precipitation of an attack. The tumor necrosis factor alpha is elevated in ENL.⁹ Amongst the precipitating factors are physical and mental stress, multiple drug therapy, vaccines, pregnancy, surgical procedures, injuries, intercurrent infections, and other antibacterial treatments.¹⁰

ENL, as seen in our patient, is characterized by the sudden appearance of crops of erythematous tender nodules or plaques, and rarely necrotic lesions mainly on the extensor surfaces of the extremities and face. Other signs and symptoms are fever and malaise, iritis, epistaxis, muscle and bone pain, nerve pain, joint pain, lymphadenitis, epididymo-orchitis, and proteinuria.^{9, 11}

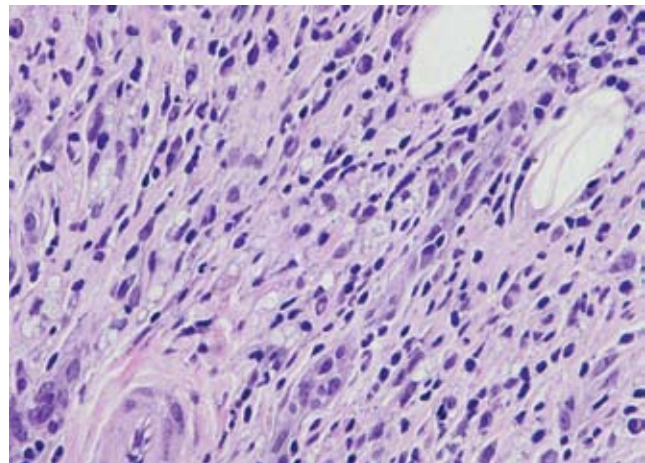


Fig. 4 Histopathology showing large number of foamy histiocytes

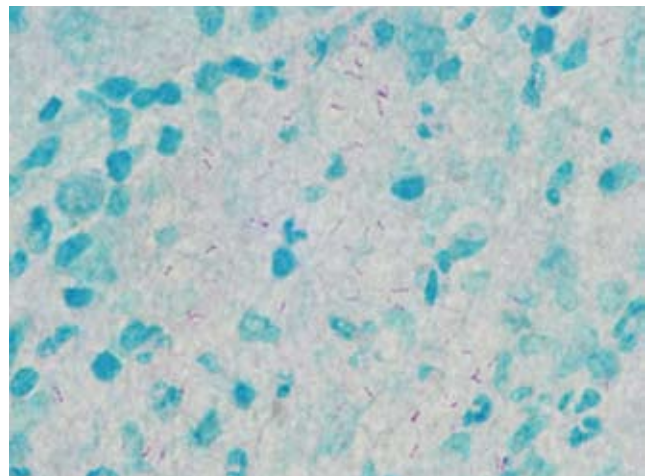


Fig. 5 Fite stain showing numerous leprae bacilli

CONCLUSION

This report highlights the role of infection and physical stress as precipitating factors for ENL

reaction. It also support the theory that leprosy can be found anywhere in the world due to international travelling.

However, it raised the inquiry about the possibility of leprosy to be presented for the first time in a reaction form, as the patient claimed. On the other hand, the positive history of a previous attack of epistaxis may means that the patient had been misdiagnosed for a long time.

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