A Woman with Erythematous Plaques

A 28-year-old domestic helper from Indonesia presented with a 1-year history of persistent, red, swollen and non-pruritic rash on her body and face. She had no significant contact history. Her medical history and drug history were unremarkable. She did not respond to a trial of intramuscular steroid injection from her general practitioner.

Physical examination revealed multiple erythematous swollen indurated plaques on her forehead, cheeks, neck, body and limbs (Fig. 1). There was no fever, ulceration, thickened nerves, joint pain or regional lymphadenopathy.

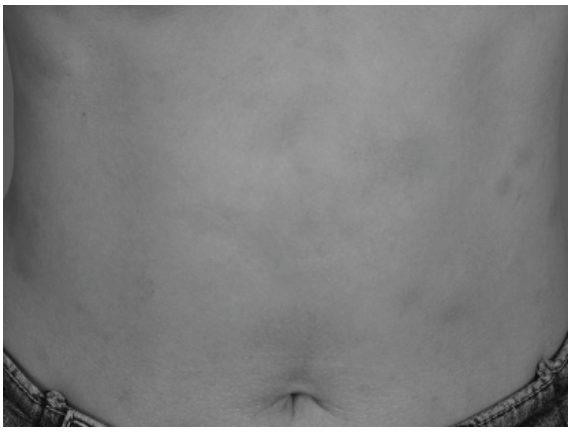

Fig. 1. Multiple erythematous swollen indurated plaques on the abdomen.

A skin biopsy from the abdomen showed numerous ovoid and sausage-shaped granuloma within the superficial and deep dermis. Some granulomas were near the nerve fibres. There was mild lymphocytic infiltrate admixed within. These granulomas consisted of foamy histiocytes with abundant pale-staining cytoplasm. Some of these foamy histiocytes contained amphophilic aggregates within their cytoplasm (globi). On the Wade-Fite stain, numerous acid-fast bacilli were seen singly and in aggregates within the cytoplasm of the histiocytes (Figs. 2 and 3).

Slit skin smear showed a bacteriological index of 2+, 5+ and 4+ on the right ear lobe, left forehead and left arm respectively.

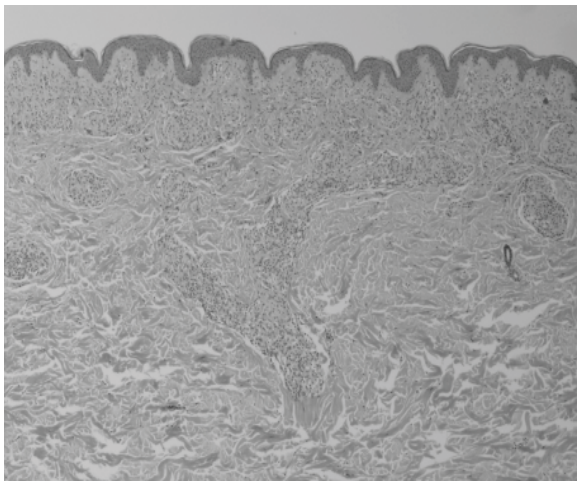

Fig. 2. Numerous round and sausage-shaped granulomas within the superficial and deep dermis with mild lymphocytic infiltrate. Granulomas consisted of foamy histiocytes within the vicinity of nerve fibres (haematoxylin and eosin x 40).

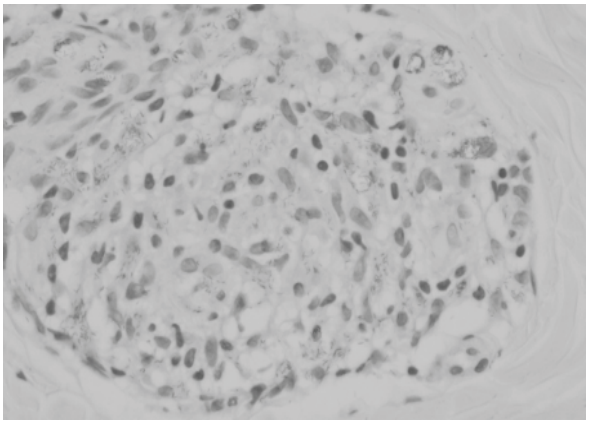

Fig. 3. Numerous acid-fast bacilli seen singly and in aggregates within the cytoplasm of the histiocytes (Wade-Fite stain x 400).

What is the diagnosis?

A. Urticarial vasculitis
B. Multibacillary borderline lepromatous leprosy
C. Cutaneous lupus erythematosus
D. Sarcoidosis
E. Granuloma annulare

Discussion

Leprosy is a chronic granulomatous infection caused by the obligate intracellular organism *Mycobacterium leprae*, an acid-fast, rod-shaped bacillus. Principally affecting the skin and peripheral nerves, it can cause irreversible impairment in nerve function leading to chronic disabilities, deformity and stigmatisation if not treated effectively.[1-4]

In the early 1980s, an estimated 15 million people worldwide were affected by leprosy. Its prevalence rate has fallen by 90%, from 21.1 cases per 10,000 population in 1985 to less than 1 per 10,000 in 2000—a goal accomplished by the World Health Organization (WHO). This significant decrease in the prevalence is attributed to the introduction of multidrug therapy (MDT).[1]

The WHO classifies patients as paucibacillary if there are 5 or less skin lesions and multibacillary if there are 6 or more skin lesions or if skin smear is positive. The Ridley-Jopling system uses clinical, histopathological features and the bacteriological index. The biopsy findings can be classified into: indeterminate, tuberculoid, borderline tuberculoid, mid-borderline, borderline lepromatous and lepromatous.[1-4]

Majority of people are not susceptible to leprosy and after typical exposure will not develop the disease. Host genetic factors are thought to influence susceptibility to infection as well as disease progression.[3] Variants of genes in the NOD2-mediated signalling pathway (which regulates the innate immune response) are found to be associated with susceptibility to infection with *Mycobacterium leprae*.[3,5]

The diagnosis of leprosy can be made with the presence of either anaesthetic or hypoaesthetic hypopigmented to erythematous skin lesions, and/or enlarged nerve and/or acid-fast bacilli on skin smear or histology.[2,6] In advanced multibacillary cases, patients may demonstrate loss of eyebrows and lashes, nasal septal perforation with collapsed nose (saddle nose deformity), and voice hoarseness (due to laryngeal involvement).[1,4,5]

In this case, the differential diagnoses also included urticarial vasculitis, sarcoidosis, granuloma annulare and cutaneous lupus erythematosus. However, urticarial vasculitis normally presents with painful erythematous wheals which may resolve with ecchymoses or post-inflammatory hyperpigmentation. Her rash was not photosensitive in which cutaneous lupus erythematosus would have normally presented. Sarcoidosis and granuloma annulare have similar presentations and are usually diagnosed histologically. In addition, she had no response to the intramuscular steroid injection which would normally be effective in all the above conditions.

The WHO MDT regimens for adult patient with multibacillary disease consist of monthly rifampin (600 mg) and clofazimine (300 mg) plus daily dapsone (100 mg) and clofazimine (50 mg). WHO initially recommended that treatment be continued for 24 months but current recommendation is 12 months. In the United States, the treatment regimen is similar apart from daily rifampicin and the duration of treatment is 24 months.[1,4,6]

Our patient reported complete resolution of her symptoms and had remained well after 24 months of MDT regimen of rifampicin, clofazimine and dapsone.

Conclusion

In summary, a diagnosis of leprosy ought to be considered in any patient with erythematous plaques not responding to conventional treatment. In an era of globalisation of labour and air travel, the physician may face challenges of diagnosing diseases not endemic to the indigenous population. An early diagnosis of leprosy with resulting appropriate treatment will help to prevent the long-term sequelae of irreversible nerve impairment and deformity.

Answer: B

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