Infantile lepromatous leprosy with vulvar localization

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Reports of leprosy during the first year of life are uncommon. In the early diagnosis, the spectrum for most of the children is reported to be largely confined to tuberculoid (TT), borderline tuberculoid (BT), mid-borderline (BB), and indeterminate forms of leprosy. We report a one-year-old female infant presenting numerous erythematous papules on her labium majus and pale flat macules on her chin and right cheek for about four months. A biopsy revealed changes typical for lepromatous leprosy (LL). Infantile leprosy and the route of transmission are discussed here.

Key words: leprosy, infancy, vulva.

The incubation period for leprosy is very long, at 2-5 years on average. For lepromatous leprosy (LL), it is twice as long as with tuberculoid leprosy[1]. There have been reports showing the occurrence of leprosy under the age of three[2]. LL seems to be rare during the first year of life. The minimum incubation period reported is as short as a few weeks, and this is based on the occurrence very occasionally of leprosy among young infants[3]. In most children, when diagnosed early, the spectrum of leprosy is reported to be incomplete and largely confined to tuberculoid (TT), borderline tuberculoid (BT), mid-borderline (BB), and indeterminate forms[2,4]. When LL and indeterminate leprosy develop in children, it seems especially that the lesions are confined to the head and the extremities, i.e. the colder parts of the body and also areas that are more prone to scratches and injuries[5]. The scalp, intertriginous areas and genitalia are relatively rich vascular and warmer zones for leprosy. Vulvar localization of LL in a one-year-old female has not been reported previously.

Case Report

A one-year-old female infant was brought to our outpatient clinic with lesions located on her chin, right cheek and labium majus. These lesions had been noticed about four months before. The pubic lesions started earlier than the facial lesions. She was a healthy, normally developed infant, born following an uncomplicated full-term pregnancy. There was no family history of tuberculosis and no history of immunodeficiency, oral candida infection, or recurrent infections. The mother of the infant and the siblings had no history of leprosy. However, her father was suffering from active LL and had been on multidrug treatment for the last six months. The laboratory studies revealed normal complete blood count, electrolytes, and renal and hepatic function tests. Dermatological examination revealed numerous copper-red papules on her labium majus and mons pubis (Fig. 1) and pale flat macules on her chin and right cheek. No peripheral nerve thickening was observed. The skin smears taken from the nose and the lesions were positive for acid-fast bacilli. Histopathological examination of the biopsy specimens taken from the skin lesions revealed evidence of LL. The patient was treated with multidrug treatment for one year (dapsone 25 mg once daily, clofazimine 50 mg twice a week and rifampicin suspension 300 mg once monthly). The treatment resulted in complete resolution of the lesions.

Discussion

Leprosy is known to occur at all ages, ranging from early infancy to old age. It is extremely rare in infants. In some areas, as many as 10% of cases develop in children younger than 15 years[6]. Usually, depending on the endemicity

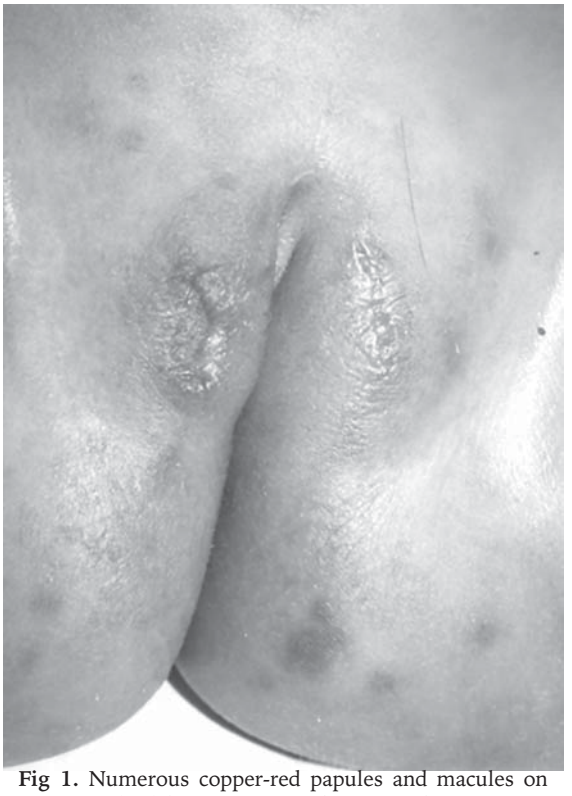

Fig 1. Numerous copper-red papules and macules on the labium majus and mons pubis.

of leprosy and socioeconomic circumstances, leprosy manifests itself after the age of six years. An important reason for this is that the incubation period appears to be between 2-5 years. The minimum incubation period reported thus far is a few weeks among young infants and the maximum is 30 years[3]. Most children, when diagnosed early, show indeterminate leprosy or leprosy with borderline features, mostly TT[2,5]. Since paucibacillary leprosy needs only a few bacteria to be clinically symptomatic, the clinical manifestations of TT develop earlier in life than those of LL. It is a paradox that children who have poor cell-mediated immunity rarely present with multibacillary disease. Brubaker et al.[2] reported 91 cases of leprosy in infants on the basis of information obtained from a review of the literature from the United States Armed Forces Institute of Pathology files and from a correspondence survey. Biopsy confirmation was available in 19 infants. None of them had LL. In 32 patients, the diagnosis of leprosy was considered certain even though histopathological confirmation was not performed. Information was derived from third-party sources in the remaining. Among them, only six infants were considered to have LL. Since then, four additional infants with leprosy have been reported (2 BT, 2 indeterminate leprosy) [1,7,8]. In the present case, although the clinical presentation of the lesions, especially the vulvar localization, did not suggest leprosy, the skin smears taken from the nose and the lesions were positive for acid-fast bacilli (5-6 bacilli (3+), in each field). In addition, histopathological findings confirmed the diagnosis of LL.

The mode of infection is still a point of discussion. Most leprologists no longer consider the skin of great importance as the port of exit or entry of *Mycobacterium leprae*[9]. They consider leprosy to be an airborne disease having a transmission pattern similar to that of tuberculosis, in which infectious patients or carriers discharge bacteria from the nasal mucosa[10]. As port of entry, the respiratory tract has been suggested, with the nose playing a central role[11]. There is also epidemiologic evidence to suggest that leprosy may be transmissible from mothers to offsprings via the placenta[12]. Here, the mother did not have leprosy or subclinical disease at parturition. The first lesion of the disease is reported by several investigators to occur on the injury-prone and more exposed areas of the body[13]. There are also numerous observations in Africans of a first patch on the forehead or cheek of a baby carried on the naked back of its lepromatous mother, and of the first lesions seen on the bare buttocks of toddlers from sitting on contaminated soil[14]. Girdhar[14] considers that the skin is at least one of the most important routes of transmission of the disease, particularly for those at the lepromatous end. In the present case, the lesions first developed on the labium majus and subsequently on the right cheek. When LL develops in children, it seems especially to be confined to the head and the extremities, i.e. the colder parts of the body. The vulva is a relatively warm zone for developing leprosy lesions. To the best of our knowledge, vulvar involvement has not been reported previously. Here, the possible mode of transmission is intriguing. The father is a case of active LL without lesions on the hands. In addition, he did not change the diaper of his child. Thus, it is difficult to explain the occurrence of vulvar lesions via skin-to-skin transmission. It is

possible that our patient acquired the infection postnatally via intrafamilial household contact, and a rapid multiplication of the organisms resulted in the disease.

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