CASE REPORT

Genital involvement and type I reaction in childhood leprosy

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Summary We describe the case of a 4-year-old boy, with a positive family history of multibacillary leprosy (borderline–borderline) in his 12-year-old sister. The patient was diagnosed to have borderline lepromatous (BL) leprosy, BI of 4+ and had two erythematous, infiltrated plaques over the scrotum. He developed type reaction, 3 months following initiation of multibacillary multidrug therapy (MB-MDT) and responded favourably to systemic corticosteroids.

Introduction

Leprosy is a well documented entity in children. Though overall prevalence of leprosy in India has declined from 5.3/10,000 in the year 2000 to 2.44/10,000 in the year 2004, it still constitutes a sizeable health problem in the paediatric age group, with an incidence of 13.8%.1 In children, it presents predominantly as paucibacillary disease, and there are only a few reports of genital involvement, the youngest being a 12-year-old boy.2,3 We report the case of a 4-year-old boy with borderline lepromatous (BL) leprosy, in type 1 reaction and the presence of scrotal lesions.

Case report

A 4-year-old male child, a resident of Uttar Pradesh, was seen in the leprosy clinic while undergoing routine family screening, as his 12-year-old sister was diagnosed to have borderline-borderline (BB) leprosy, in type reaction and had been on multibacillary multidrug therapy (MB-MDT) for 8 months. Examination of the boy revealed multiple asymptomatic, hypopigmented lesions over the body. The patient had had one episode of...
epistaxis 1 year previously. There was no history of peripheral anaesthesia, painless ulceration or muscle weakness.

The patient weighed 15 kg and a BCG scar was absent. On cutaneous examination, there were multiple, hypopigmented macules, varying in size from 0.5 to 3 cm, with ill to well defined margins, present bilaterally but asymmetrically over the face, limbs and trunk. There was loss of hair over the lesions, with impairment of sensations that could not be graded by the child. Two erythematous, non-tender, infiltrated plaques of size 0.5 × 1.5 cm, with well-to ill-defined margins were also seen on the scrotal skin. Scrotal contents were palpable normally and were non-tender. A cremasteric reflex was present. Bilateral ulnar and common peroneal nerves were uniformly thickened, more on right side, firm and non-tender. Systemic examination did not reveal any abnormality.

Haematological investigations including complete haemogram with ESR (Hb 11.8 g%, ESR 8 mm 1st hour), biochemical investigations, hormonal evaluation (LH, FSH) and urine examination were within normal limits. Chest X-ray did not reveal any abnormality. Testicular ultrasound did not show any evidence of testicular atrophy. After obtaining written consent from the parents, slit skin smears and skin biopsy were performed. Slit skin smear examination from eyebrows, ear lobules, skin lesion on right elbow and the scrotal lesion showed an average bacteriological index (BI) of 4 + . Histopathology from the skin lesion showed epidermal atrophy and the presence of periappendageal loose granulomatous infiltrate comprising histiocytes, lymphocytes and a few plasma cells. The nerve bundles showed reactive proliferation of the perineurium resembling onion skin appearance, features suggestive of borderline lepromatous (BL) leprosy. Biopsy from the scrotal lesion showed features of lepromatous lepromatous (LL) leprosy in the form of diffuse granulomas comprising histiocytes, lymphocytes and foamy macrophages containing acid-fast bacilli in mid and lower dermis.

The patient was started on MB-MDT in accordance with his body weight. After 3 months of treatment, he developed sudden erythema and oedema of the old lesions, including those of the face and scrotum, and new lesions appeared, some of which were punched out (Figures 1 and 2). There were no constitutional symptoms and neuritis was absent. A diagnosis of type reaction was made; the patient was started on non-steroidal anti-inflammatory drugs and the dose of clofazimine was increased from 50 mg twice a week to 50 mg daily. On follow-up after 15 days, there was no improvement and oedema in the lesions had increased. The patient was started on prednisolone 15 mg daily, with marked resolution of oedema at 4 weeks. Over 6–8 weeks, prednisolone was tapered to 10 mg daily with complete resolution of erythema and oedema.

Discussion

In developing countries, leprosy constitutes a sizeable health problem and it is widely accepted that children especially under 5 years are more susceptible to developing leprosy than adults. Many studies conducted on childhood leprosy shows a preponderance of leprosy in the age group 6–14 years and only 5.8–6% of cases below 5 years of age, indicating that disease usually takes longer to manifest due to the long incubation period. However, in highly susceptible individuals, it is reasonable to assume that small inocula could rapidly multiply to produce high bacterial burden and clinical disease manifests in a brief period.
The most common type of presentation in this age group (<5 years) is of paucibacillary leprosy, with borderline tuberculoid being commonest. The majority of cases have solitary lesions, situated on exposed parts. The rate of smear positivity is less than 2% of cases (mainly in BL/LL cases), and it has been reported that smear positivity increases with age. In the present report, a 4-year-old boy had BL leprosy with multiple asymmetrical lesions distributed on both exposed and unexposed parts, with BI of 4+.

There are several case reports indicating that BCG vaccination attenuates the severity and incidence of multibacillary leprosy by modulating immunity. In the present case, the unvaccinated status of the child might have contributed to the development of BL leprosy. It has been shown that the risk of a person developing leprosy is 4 times higher when there is leprosy contact in the neighbourhood; the risk is increased to 9 times if the contact is within the immediate household, and even higher when the contact is multibacillary. This patient had an elder sister with BB leprosy.

In children, reactional episodes and disabilities are less frequently seen and usually occur in older children, due to their relatively well developed immunological status. There are no reports of reactions (type 1 or type 2) in BL/LL cases in children below 5 years of age. This patient developed type I reaction 3 months after initiation of therapy, with intense oedema over the lesions, including those on the scrotum. This emphasizes the need for regular follow-up and prompt use of steroids if necessary, in case of reactions, to prevent deformities and testicular atrophy.

Figure 1. Infiltrated plaques of leprosy in type I reaction over face and upper limbs.
Although the involvement of male genitalia, particularly the gonads, is well known in leprosy, scrotal skin involvement has been described as an unusual site because of its relative warmth and because it is an infrequent site of injury. However, there are now reports of involvement of scrotal skin in all types of leprosy due to the liberal supply of nerves. Genital lesions have been mostly described in adults, the youngest being a 12-year-old boy. The present case report describes a 4-year-old boy with genital lesions over the scrotum, an unusual site.

This report of a 4-year-old BCG unvaccinated child, with BL leprosy type I reaction and scrotal lesions, has many unusual clinical features. Therefore, careful family screening of young children in high endemic areas following the detection of an index case, and complete examination inclusive of genitalia to initiate timely therapy, is warranted in order to prevent disease progression and complications. This case also stresses the need for parental education for early detection and treatment of leprosy, so as to halt disease transmission amongst family members and the community.
References