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Original Article

Clinical Presentation and Histopathology of Childhood Leprosy

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Abstract

Background: Leprosy is one of the oldest diseases of mankind. It is well-documented in children with an incidence of 13.3%, as immune system is not fully developed. 75% of cases regress spontaneously without treatment.

Aim: To study incidence, duration, spectrum, reactions, deformity, slit skin smear (SSS) and skin, and nerve biopsy in childhood leprosy.

Materials and Methods: Leprosy cases from 0 to 14 years age who attended Department of Dermatology and Leprosy of Government General Hospital, Chennai, were collected for 2 years (study period).

Results: During the study period of 2 years, total numbers of childhood leprosy cases were 46. Most number of cases was seen in 10-14 years age group - 36 cases (78.3%). Most common spectrum is borderline tuberculoid 27 cases (58.7%). Percentage of cases in household contact was 19.6%. Type 1 reaction was seen in 3 children. Type 2 reaction was not seen. The deformity was seen in 5 children. SSS was positive in only 4, all of them were borderline leprosy cases.

Conclusion: Leprosy still occurs in children in sizable and constant proportion, even though the prevalence rate has reduced below one per thousand and we are in the run for total eradication of leprosy.

Key words: Borderline leprosy, Childhood leprosy, *Mycobacterium leprae*

INTRODUCTION

Leprosy is a chronic disease caused by *Mycobacterium leprae*, infectious in some cases, and affecting the peripheral nervous system, skin, and certain other tissues.[1] It has been a major public health problem in many developing countries for centuries. Children are believed to be the most vulnerable group to *M. leprae* infection and clinical manifestation is often seen in adolescence or young adulthood following the long incubation period. Leprosy in children has epidemiological significance and is considered as index of the prevalence of disease. It forms an important link in the study of natural evolution of disease. India represents about 76% of global burden. The overall prevalence of leprosy in India has declined from 5.27/10000 in the year 2000 to 1.34 in 2005, still, it constitutes a sizable health problem in pediatric age group with incidence of 13.3%. Childhood leprosy forms an important link in the study of natural evolution of disease. The spectrum of disease is usually incomplete in children. It is rare under 2 years of age.[2] It is often unrecognized, and 75% cases regress spontaneously without any treatment. The youngest age reported for occurrence is 3 weeks in Martinique. The youngest case of tuberculoid leprosy confirmed by histopathology was infant of 2.5 months old.[3] In children it is equally prevalent in both sexes. Childhood leprosy usually responds rapidly to treatment. Reactions, relapses are not uncommon.[4] Ocular leprosy, deformities, infectivity, poor tolerance to drugs, and special variants like Histoid leprosy are rare in children. The worldwide prevalence of disease has decreased dramatically, since the inception of elimination

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plan, but the disease deduction rate has remained almost constant over last 10 years with the high rate of infection (17%) in children.

Aim

To study incidence, duration, spectrum, reactions, deformity, slit skin smear (SSS) and skin, and nerve biopsy in childhood leprosy.

MATERIALS AND METHODS

This prospective observational study was conducted in the Department of Dermatology, Government General Hospital, Chennai. Childhood (0-14 years) leprosy cases were screened, complete history of presenting complaints, duration of illness, history of contact, treatment, Bacillus Calmette–Guérin vaccination, were taken from the informant usually parents. A complete general and dermatological examination regarding the morphology, number, size, site, color, anesthesia, margins, surface, satellite lesions and general clearing of skin lesions and involvement of peripheral truncal nerves, and cutaneous nerves were done. Reactions and deformities were also noted. Slit smear examination was done from a minimum of three sites (If there was a single lesion, i.e., from the lesion and both ear lobes and from a minimum of four sites if there was more than one lesion, i.e., from both ear lobes and two skin lesions). A SSS examination was performed using standard techniques. Stained by Ziehl–Neelson stain and graded by Ridley's scale for bacteriological index. A skin biopsy was obtained using local anesthesia, following consent and stained with hematoxylin and eosin for histopathological examination. Nerve biopsy was done and stained with hematoxylin and eosin and Fite-Faraco staining in the case of pure neuritic leprosy.

RESULTS

A total number of childhood leprosy cases is 46. Of these, 36 (78.3%) were in 10-14 years age and 7 (15.2%) were in 5-9 years. The age of onset, in most cases, 29 cases (63%) was 10-14 years. Borderline tuberculoid is seen in 27 cases (58.7%). Only 4 (8.7%) cases of borderline lepromatous leprosy were seen and no lepromatous leprosy. Pure neuritic leprosy is seen in 5 (10.9%) children. 2 presented with foot drop, 1 with claw hand and trophic ulcer and 1 with ulnar nerve abscess and clawing of right little finger. Single skin lesion was the most common presentation in 22 cases (47.8%). Ulnar (27cases, unilateral in 22 and bilateral in 5) is most common truncal nerve and radial cutaneous (13 cases, unilateral in 7 and bilateral in 6) is most common peripheral nerve involved. Type 1 reaction was seen in 3 cases. Type 2 reaction is not seen. Deformities were seen in 5 children. Claw hand, foot drop, and trophic ulcer were the deformities encountered. Ocular and Histoid leprosy is not seen.

DISCUSSION

The incidence of leprosy is higher in 10-14 years of age and 30-60 years of age group. A total number of childhood leprosy cases (0-14 years) seen was 46 during the study period of 2 years. Of the 46 patients, 36 (78.3%) were in 10-14 years age group, 7 (15.2%) were in 5-9 year age group, and 3 (6.5%) were in 0-4 year age group. Age distribution reflects a clear preponderance of older children because of relatively long incubation period. Incubation period may be as short as few months as long as 20 years or more. On an average it is between 2 and 5 years.[5] The youngest patient seen in the study was a 4-year-old girl who had borderline tuberculoid leprosy with age of onset at 3 years. Two more children of 4 years old, one with intermediate leprosy and another with tuberculoid leprosy with age of onset of 3 years 6 months and 3 years 9 months, respectively, were also seen in the study. The age of onset, (obtained by subtracting the duration of disease from age of patient) in most cases, 29 cases (63%) was 10-14 years among the children in the study group (0-14 years). There was a preponderance of boys (31 cases over 15 cases, with a male-female ratio of 2.07:1). This observation is similar to most of earlier studies, and it could be because of environmental and socio-cultural factors such as greater exposure in boys.

Duration of disease at the time of detection in most of the cases was found to be less than or equal to 1 year. Overall 9 children gave a history of contact with a leprosy patient within the household, father being the index cases in 4 cases and mother in 2 cases. Borderline tuberculoid is most common type seen in children.[6,7] In our study, borderline tuberculoid is seen in 27 cases (58.7%). Only 4 cases of borderline lepromatous leprosy were seen and no lepromatous leprosy was seen indicating clearly the spectrum is incomplete. It is a paradox that children with poor cell-mediated immunity rarely presents with multibacillary disease. Claw hand is most common deformity followed by trophic ulcer, foot drop, and wrist drop.[7] Pure neuritic leprosy was seen in 5 children. 2 presented with foot drop, other with numbness below the right knee, 1 with claw hand and trophic ulcer, and 1 with ulnar nerve abscess and clawing of right little finger.

The clinical presentation in most cases was hypopigmented skin lesions with or without sensory impairment. Single skin lesion is more common followed by 2 to 3 and more than 4 is rare.[2] In our study, single skin lesion was the most common presentation in 22 cases (47.8%). The sites for

the development of single lesion are predominantly seen on the exposed parts.[8-12] Similarly, exposed parts such as forearm, face, knee, and leg were the most common sites involved in 15 cases (68%) than covered parts such as back, arms, buttocks, and thighs in 7 cases (32%). Some of earlier studies observed an increased incidence of single skin lesion over gluteal region. In this study among 22 patients presented with a single skin lesion, 6 had lesions over forearm (common site) followed by cheeks in 5 cases The most common morphology of the lesions seen were macules, patches, plaques, and usually hypopigmented followed by erythematous and copper colored lesions. Size of lesion varied from <1 cm - 45*15 cm involving most of lower limb, the largest lesion in the study. Ichthyosis and traumatic fissure were seen in few cases. Of the peripheral truncal nerves, ulnar nerve was the most common nerve involved followed by lateral popliteal, posterior tibial, and median nerve. Radial cutaneous nerve was the most common cutaneous nerve involved followed by greater auricular, sural, supraorbital, and supraclavicular nerves. Involvement of lateral, intermediate and medial cutaneous nerves of thigh was seen in one case.

Type 1 reaction was seen in 3 cases of whom 2 had borderline lepromatous leprosy and 1 had borderline tuberculoid leprosy. Type 2 reaction was not seen. Relapse was seen in a case of 10-year-old boy who presented with increase in size of skin lesion 2 years after completion of paucibacillary treatment. Deformities were seen in 5 children. Claw hand, foot drop, and trophic ulcer were the deformities encountered. 14-year-old male child, the case of pure neuritic leprosy had claw hand both sides and trophic ulcer right foot. 14-year-old male child, case of borderline tuberculoid leprosy had left side claw hand and left foot drop. 11-year-old male child, a case of pure neuritic leprosy, had clawing right little finger. 12-year-old female child (pure neuritic leprosy) had left foot drop. 6-year-old female child (pure neuritic leprosy) had left foot drop. Ocular leprosy among children is rare and nonblinding.[4] Ocular involvement was not seen in any of the cases. Special variants like Histoid leprosy were also not seen.

The frequently encountered differential diagnosis was polymorphic light eruption, pityriasis alba, tinea versicolor, early vitiligo, resolving morphea, and post-inflammatory hypopigmentation. A case of ganglion, mimicking nerve swelling, along the course of lateral popliteal nerve was also seen.

SSS was positive in only 4 cases; all of them were borderline lepromatous cases. Histopathological examination of skin and nerve biopsy was very useful in establishing the diagnosis.

CONCLUSION

The most common age of occurrence is 10-14 years. Incidence was more among male than female. Single skin lesion, occurring over exposed parts is the common presenting feature. Ulnar nerve is most commonly involved truncal nerve, and radial cutaneous nerve is most commonly involved cutaneous nerve. Borderline tuberculoid is the most common spectrum. Leprosy toward lepromatous pole is rare. Reactions, relapse, and deformities are rare. Leprosy still occurs in children in sizable and constant proportion, even though the prevalence rate has reduced below one per thousand and we are in the run for total eradication of leprosy.

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