

Clinical pattern of deformities and disabilities in leprosy patients in rural Bangalore – A two year study at tertiary level hospital

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Abstract

Introduction: Aim of the study was to know the various patterns of deformities and disabilities in leprosy and their relation to various demographic elements and the occurrence of grade 2 deformities in newly diagnosed cases.

Materials and Method: A total of 50 cases of leprosy belonging to all age groups and both sexes were selected and included in the study after taking their consent. Initially a detailed history and examination was done with reference to clinical features of leprosy. Deformities were noted. In all cases necessary investigations were done.

Result: Majority of patients belong to age group of 21 to 30 years with male preponderance and low socioeconomic status. Borderline tuberculoid being the most common presentation. Grade 2 deformity was more when compared to grade 1 disability.

Conclusion: In conclusion it can be said leprosy still continues to be a disease which is commonly seen in low socio-economic group of people, probably because of the poor knowledge of the disease among illiterates, leprosy continues to be a domestic, national and global burden.

Keywords: Leprosy, Deformities, Disability, Hansen's disease, Low socio-economic status, Illiterates.

Introduction

Leprosy or Hansen's disease is a common infectious disease of mankind. It has been mentioned as early as 600 B.C in the Indian literature.⁽¹⁾ It is a chronic granulomatous⁽²⁾ disease caused by *Mycobacterium leprae* affecting the peripheral nervous system, the skin and certain other tissues. World Health Organization declared it as a major health problem as it is known to be associated with crippling deformities. Due to its crippling deformities, it is responsible for much of the social stigma and social ostracization of the patients even with the successful treatment. The deformities resulting from leprosy cause misery to patient and result in extensive loss of manpower and economy loss to society.⁽³⁾ Causative bacillus was discovered by Sir Gerhard Henrick Armauer Hansen in 1873.

M. Leprae is an intracellular acid-fast, alcohol fast, gram positive, aerobic, rod-shaped obligatory bacterium and is surrounded by the waxy cell membrane coating characteristic of mycobacterium species.⁽⁴⁾ Due to extensive loss of genes necessary for independent growth, *M. Leprae* is an obligate pathogen and uncultivable in the laboratory, a factor that leads to difficulty in definitively identifying the organism under a strict interpretation of Koch's postulates.⁽⁵⁾

Leprosy is unique among bacterial infections in that it parasitize the peripheral nerves and damage them. The disease is feared for the deformities and disability it produces in its host. Disability not only occurs due to the reactions which usually occur after initiation of treatment, but also a large proportion of it result from insidious progression of the disease. A considerable portion of disability load is the result of failure to incorporate activities relating to its prevention

by using simple techniques and patient motivation into leprosy management.⁽⁶⁾ Both magnitude and severity of disability in new leprosy patients measure indirectly the disease transmission in the community and rapidity of case detection.⁽⁷⁾

Under leprosy control program more attention has been given to prevention of disability. Among communicable diseases, leprosy remains a leading cause of peripheral neuropathy and disability in the world, despite extensive efforts to reduce the disease burden. In India, efforts to control leprosy began in 1955 when the National Leprosy Control Program was launched. The program had been chalked out by the center and has been operated through the states by establishment of Leprosy Control Units, Survey Education & Treatment centers, Urban Leprosy Control Units. It was redesignated as National Leprosy Eradication Program in 1982 with the aim of eradicating leprosy by 2000 A.D.⁽⁸⁾

The WHO has estimated that about 1- 1.5 million people were disabled due to leprosy in 1995. The disability rates declined from 10.3% to 3.7% in 1995. The Government of India declared that leprosy has been eliminated from the country in December 2005, with a prevalence rate of 0.92 per 10000 population.⁽⁹⁾

Disability assessment is very important not only to evaluate the effectiveness of the control program but also for the patient, whose most important worry is the stigmatizing deformities leprosy patients suffer. The earliest detection of sensory loss might reduce these secondary deformities. Therefore timely diagnosis of grade-1 disability, is required for disability limitation and mitigation.⁽¹⁰⁾

Materials and Method

The study was conducted patients with Hansen's disease who attended the outpatient department of Dermatology, Venereology and Leprology, MVJ Medical College and Research Hospital, Hoskote, Bangalore. The study was conducted over 24 months between November 2014 and November 2016.

Inclusion criteria:

1. All patients who presented to us and were diagnosed with leprosy, irrespective of treatment status.
2. Patients who were diagnosed for the first time, who are on active treatment and those who were RFT but in the surveillance period are all included.

Exclusion criteria:

1. Patients with disabilities of some other known cause.

All patients fitting the criteria and willing to participate in the study were selected. Consent was taken from the patients after being informed about the aim of the study. Case Profroma was prepared which was used to record the complaints, history, clinical features and results of clinical tests and investigations.

1. Data regarding sociodemographic variables like age, sex, education, occupation were recorded.
2. Detailed history was noted.
3. Patient's whole body was examined for skin lesions.
4. Peripheral nerves examined for size, consistency, nodularity, abscess and tenderness.
5. Skin of hands and feet were examined for presence / absence of sweating, fissures, ulcers, callosities, scars.
6. Face, hands and feet were examined for any of these Visible Deformities.
7. Nerve function assessment for motor and sensory loss.
8. Examination of eyes.
9. Slit skin smear.
10. Skin biopsy for histopathological examination was done to confirm the diagnosis.

Results

Fifty patients of Hansen's disease who presented to the outpatient Department of Dermatology, Venereology and Leprology, MVJ Medical College and Research Hospital, Hoskote, Bangalore were studied.

Age distribution: The age of the patients ranged from 11 to 90 years. The majority were in the 81-90 years group followed by 21-30 years age group.

Sex distribution: Out of the 50 patients, 39 were males (78%) and 11 were females (22%). Male to female ratio was 3.5: 1.

Socioeconomic status: 56% patients belonged to lower class, 42% belonged to middle class and 2% belonged to upper class.

Spectrum of leprosy: Of the 50 cases, proportion of BT cases was 34%, which is higher compared to other forms of leprosy. PNL was 26%, BL was 18%, LL was 18%, BB was 2% and TT was 2%.

Disability grade: Among the 50 patients, 37 patients (74%) had grade 2 deformity and 13 patients (26%) had grade 1 deformity.

Type of deformity

Claw hand: In this study, 19 patients (38%) had claw hand which was higher compared to other visible deformities.

Trophic ulcers: Of the 50 patients, 19(38%) had trophic ulcer.

Anesthesia (Grade 1 deformity): In this study, the proportion of cases with anesthesia was 24% (12 patients).

Lagophthalmos: 3 patients (6%) presented lagophthalmos.

Foot drop: It was seen in only 1 patient.

Others: In this study, 16% had auto amputation of digits, 4% had corneal opacity, 4% had ape thumb, 2% had madarosis, 2% had leonine facies, 2% had banana fingers, 2% had premature senility and 2% had Buddha ears.

Sensory function deficits are more commonly seen in patients in lepromatous end of the spectrum. In BT Hansen's disease sensory functions are affected only in patients with patches on hands and feet.

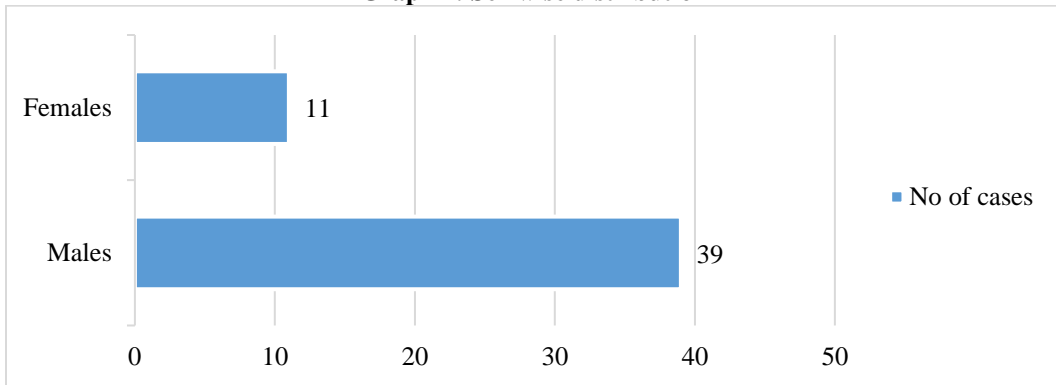
Frequency of motor deficits like claw hand and lagophthalmos is common in BT leprosy followed by pure neuritic leprosy but the severity of nerve damage is high compared to lepromatous leprosy.

In our study, 24% patients presented in reaction out of the total 50 cases. Out of 12 patients, 7 (14%) patients presented with Erythema nodosum leprosum, 5 (10%) patients presented with type 1 reaction.

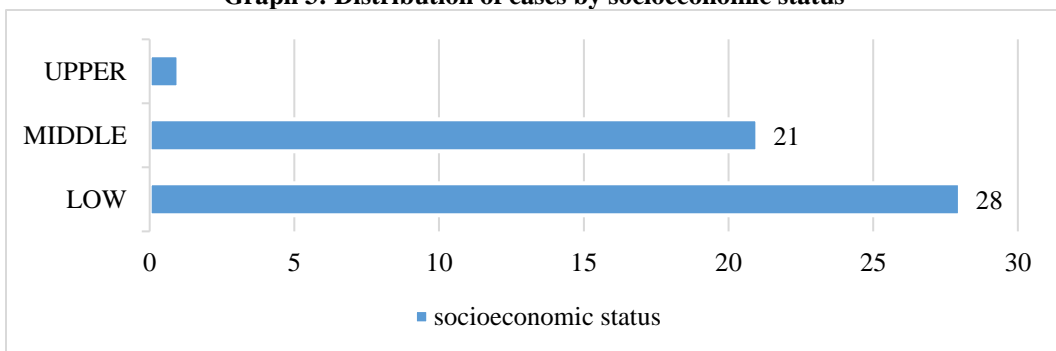
Graph 1: Age wise distribution



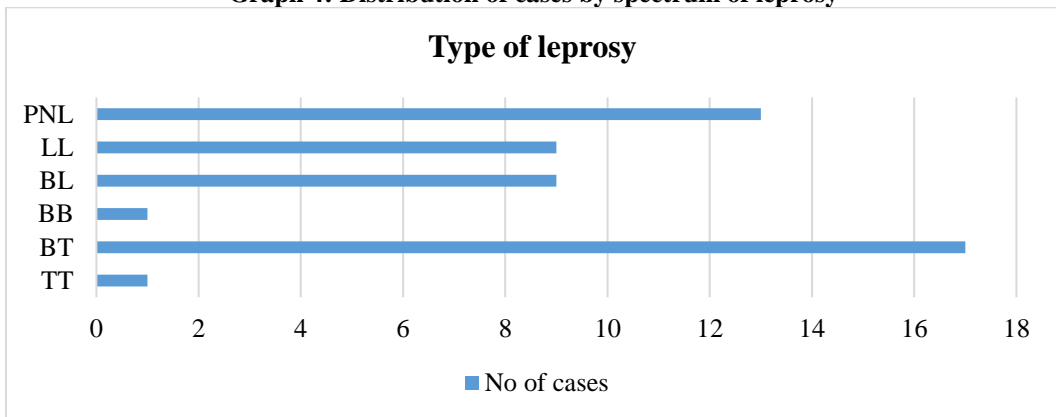
Graph 2: Sex wise distribution



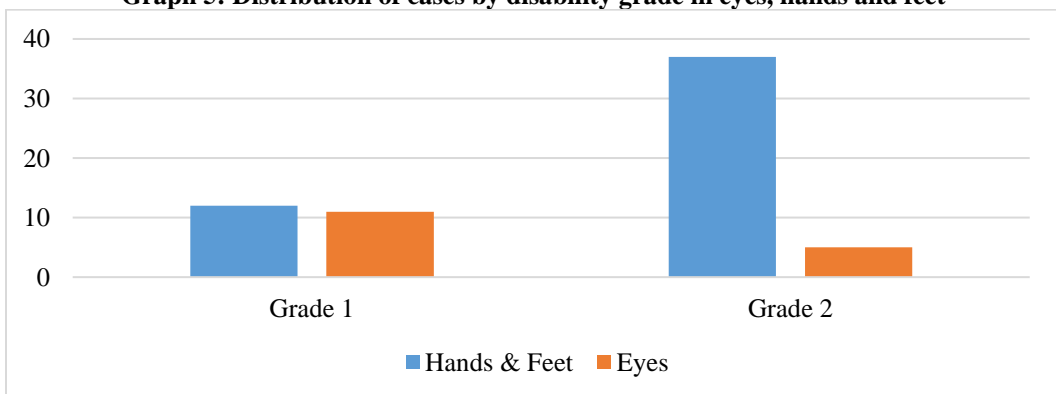
Graph 3: Distribution of cases by socioeconomic status



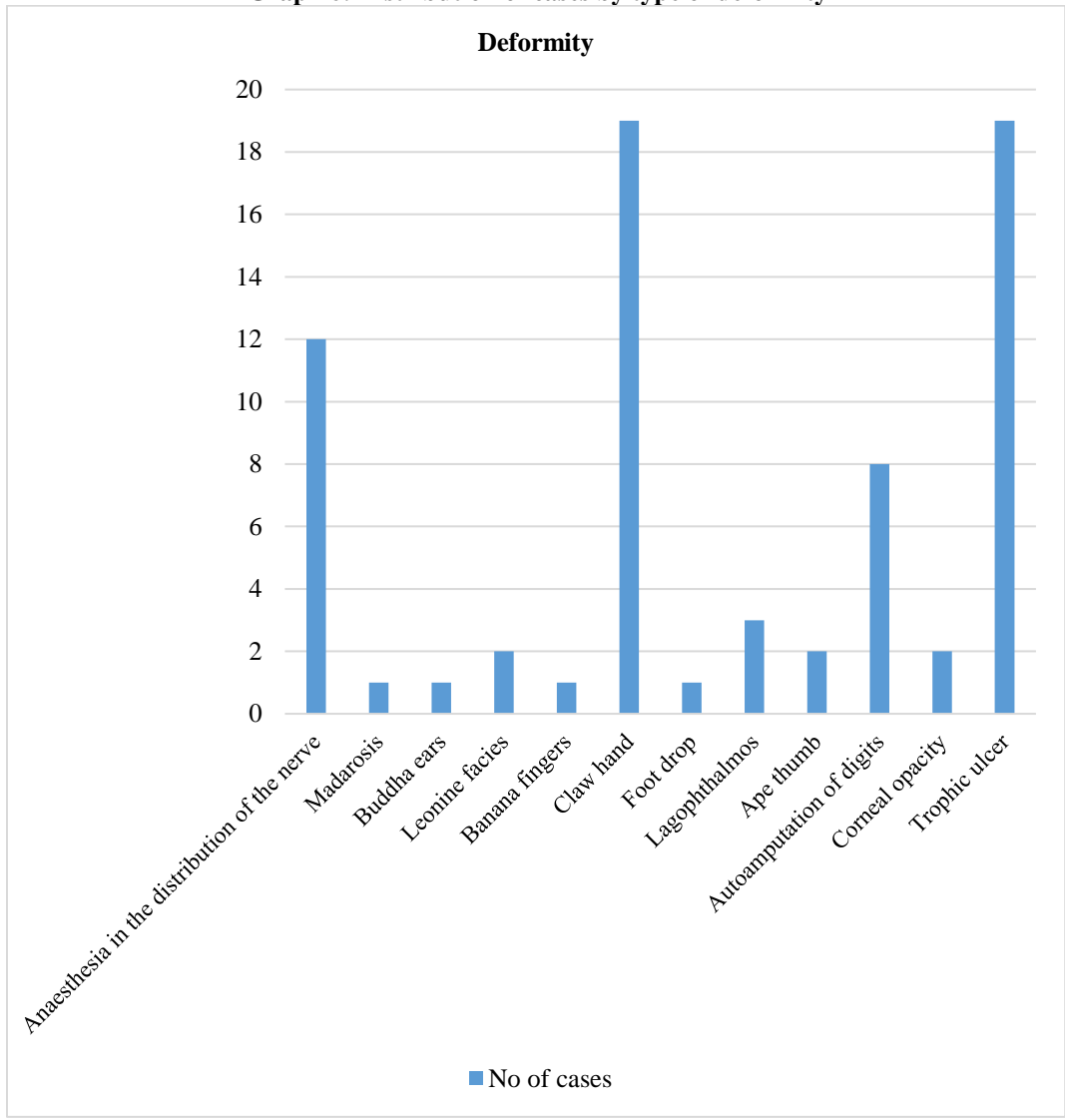
Graph 4: Distribution of cases by spectrum of leprosy



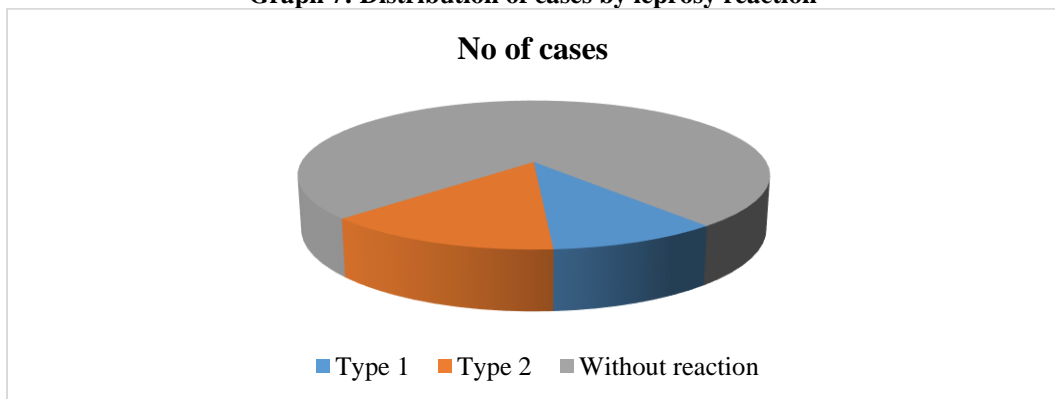
Graph 5: Distribution of cases by disability grade in eyes, hands and feet

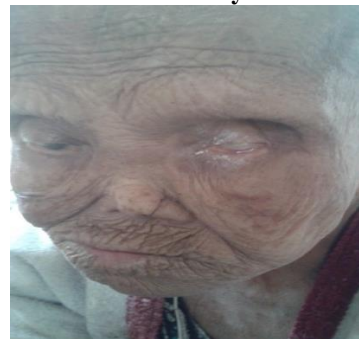


Graph 6: Distribution of cases by type of deformity



Graph 7: Distribution of cases by leprosy reaction



Photograph 1: Total claw hand with contracture**Photograph 2: Trophic ulcer over the base of metatarsophalangeal joint****Photograph 3: Bilateral madarosis****Photograph 4: Bilateral lagophthalmos****Photograph 5: Resorption of fingers****Photograph 6: Corneal opacity with sunken nose deformity**

Discussion

Disablement of leprosy affected persons is a process, comprising chain of events occurring over a period of time. Three major stages are recognized in the process of disablement:

Impairment:⁽¹⁾ This refers to changes or abnormalities (including losses) in body parts and functions (including mind) as a consequence of a disease/disorder (changes in the structure and functioning of certain parts of the body).

Impairments may be:

- Primary- Directly due to disease.
- Secondary- As a consequence of primary ones.

Primary impairments in leprosy include- Thickening and nodulation of skin & subcutaneous tissue, eye changes, loss of sensibility, motor paralysis in hands, feet & eyes.

Secondary impairments include ulceration, contractures, loss of vision.

Disability:⁽¹⁾ (activity limitation): It is defined as inability to do certain activities, which are normally possible. The affected person finds it difficult / unable to perform some activities at home or workplace because of impairments.

Handicap⁽¹⁾ (participation restriction): Persistently disabled persons experience many disadvantages that limit or prevent them from fulfilling their normal role in

society. Leprosy affected persons may lose their jobs, earning capacity, social rejection.

Deformity: Deformity is defined as any loss or abnormality of psychological, physiological or anatomical structure or function. Deformities may be either visible impairments or consequences of invisible impairments. A primary impairment is a direct consequence of the causative disorder (e.g. madarosis, collapse of nasal bridge) and secondary impairment is not directly from the original disorder (e.g. trophic ulcer).⁽¹²⁾

Risk factors for deformities in leprosy⁽¹³⁾

- Type of Leprosy – The more extensive and bacilliferous types like borderline and lepromatous leprosy carry a high risk, unlike the more circumscribed and low bacilliferous types like indeterminate and tuberculoid leprosy in which the risk of deformity is absent or very low.
- Number of nerve trunks involved – patients with involvement of three or more nerves have a high risk of developing a significant disability.
- Attacks of reaction and neuritis increases the risk.
- Duration of active disease – The longer the disease remains active, the greater is the risk of developing disability.

Epidemiological studies in South India show that males, adults and heavy manual workers develop deformities more often than females, children and those engaged in light work respectively.⁽¹²⁾

Types of deformities⁽¹³⁾

Depending on their causation, deformities in leprosy patients are described as

1. Specific deformities – arise from local infiltration with *Mycobacterium leprae*
 - a. Face – Madarosis, premature senility, Buddha ears, deformities of pinna, sunken nose deformity, nasal septal and palatal perforation.
 - b. Hand – Banana fingers, shortened fingers, reaction hand.
2. Paralytic deformities – result from damage to motor nerves
 - a. Hand – claw fingers, wrist drop, wasting of muscles, guttering, and ape thumb
 - b. Foot – claw toes, foot drop
 - c. Face – lagophthalmos, facial palsy
3. Anesthetic deformities – occur as a late sequelae of neglected injuries in parts rendered insensitive.
 - a. Face – exposure keratitis
 - b. Hand & foot – ulceration, scar contractures, shortening of digits, skeletal disorganization of foot, fixed deformity of foot.

Specific and paralytic deformities are examples of primary impairments caused directly by the disease. Whereas anesthetic deformities are secondary impairments since they follow sensory loss. When anesthesia is considered, feet are more affected but

when grade II deformity is considered, hand deformities were found almost twice as often as that of feet.⁽¹⁴⁾

Mechanisms causing disability in leprosy: Disability in leprosy result from the nerve damage and the direct infiltration of skin & mucosa by the bacilli.

In Lepromatous leprosy fine nerve twigs of sensory & autonomic fibers in skin are affected while in tuberculoid leprosy major peripheral nerves are affected at sites of predilection.⁽¹⁵⁾ The three physiological functions of nerves – sensory, motor and autonomic are affected.⁽¹⁶⁾

In tuberculoid leprosy, nerve damage begins early and progresses rapidly by infiltration of the nerve by the granuloma and reactional edema. Also the nerves are involved at sites where they pass through unyielding canals of fibrous tissue such that any increase in intraneural substance like inflammatory cells results in raised local pressure compromising its blood supply. Ulceration, scar formation & secondary infection create a vicious cycle of events that increase the damage caused.

In Lepromatous leprosy peripheral nerve damage progresses slowly. Sensory nerve endings of cooler parts of the body like dorsum of hands & feet, ears, nose are also affected. Sensory changes & motor paralysis in distribution of peripheral nerve trunks occur after several years when extensive fibrosis replaces the nerve axons. Involvement of ears, eyes, nose & other organs result in specific deformities and disabilities like loss of vision.

Patients with borderline leprosy have the most extensive nerve involvement & their liability to reactions predisposes them to most severe nerve damage.

The eight causes of deformity are –

1. Anaesthesia
2. Dryness of skin
3. Muscular paralysis
4. Misuse
5. Ulceration
6. Scar formation
7. Secondary infection
8. Invasion of tissues by *M. Leprae*

At the beginning of this new millennium, we have been able to reach the last mile in the race to eliminate leprosy in most part of the world including India.

According to official reports received from 138 countries from all WHO regions, the global registered prevalence of leprosy at the end of 2015 was 1,76,176 cases (0.18 cases per 10 000 people). The number of new cases reported globally in 2015 was 2,11,973 (0.21 new cases per 10 000 people). In 2014 the number of new cases reported was 213 899, and in 2013 the number of new cases reported was 2,15,656.⁽¹⁷⁾

The nature of leprosy problem is thus shifting from simply providing antileprosy treatment to the affected persons to dealing with the consequences of leprosy especially prevention of disability due to leprosy.

Occurrence of disability in leprosy indicates some lacunae in leprosy control.

In this study, fifty patients of leprosy who were diagnosed for the first time, who were on active treatment and those who were release from treatment but in the surveillance period were screened and included.

Age Distribution: A study conducted by Kar et al⁽¹⁸⁾ reported that 60% were aged above 40years and 40% were in 21 to 40 years age group. Reddy et al⁽¹⁹⁾ reported in his study 30.76% were above 40years. Anil Kumar et al⁽²²⁾ in his study reported 65% of the patients in their study group belongs to age above 59 years.

In our study, peak age group affected was 21 to 30 years (20%) followed by 41 to 50 years (18%) and 61 to 70 years (18%).

So, the data in current study is in accordance with data recorded in previous studies.

Sex Distribution: In a hospital based retrospective study conducted by Peters ES⁽¹⁸⁾ between 1988 to 1997, it was reported that 66% of patients were males and 33% were females with a ratio of 2:1. In WHO expert committee report⁽¹¹⁶⁾ of 2003 on leprosy situation in India, 63.2% were males and 33.8% were females.

In the present study, 78% were males and 22% were females. Male to female ratio was 3.5: 1.

Although leprosy affects both sexes, in most parts of the world males are affected more frequently than females often in the ratio of 2:1. The increased incidence among males can be attributed to the increased chances of exposure to disease in men as in our set up but not in rural India. There the norm is still of males going out and working. Whereas women are homemakers still. Most of the health workers who screen patients for leprosy in rural side are males. So women are hesitant to approach them, and hence they will not be diagnosed.

Socioeconomic Status: In an epidemiological study conducted by Anil Kumar⁽²⁰⁾ et al, most of their patients belong to low socioeconomic group. The possible reason for this increased incidence of disease in lower class group may be related to overcrowding, increased environmental exposure to disease and lifestyle.⁽²¹⁾

In this study, 56% patients belonged to lower class, 42% belonged to middle class and 2% belonged to upper class.⁽²⁴⁾

Spectrum of leprosy: In this study, proportion of BT cases was 34%, which is higher compared to other forms of leprosy. Pure neuritic leprosy was 26%, BL was 18%, LL was 18%, BB was 2% and TT was 2%.

An epidemiological study in leprosy conducted by Anil Kumar⁽²⁰⁾ reported 30.5% patients belonged to BT spectrum. As explained in Hasting's text book of leprosy, BT form is the most common type of leprosy.⁽²¹⁾ PNL was the second most common type in this study i.e. 26%. The incidence of PNL in a study by Anil Kumar 114 was 20%. In most Indian studies PNL occurs with a higher frequency in South India where it

constitutes up to 18%. So, increased incidence of PNL in this study appears to reflect the existing epidemiological trend in South India.

Disability Grade: In a study conducted by Schipper,⁽²²⁾ 19% had grade-1 deformity and 21% had grade-2 deformity.

In this study, among 50 patients, 37 patients (74%) had grade 2 deformity and 13 patients (26%) had grade 1 deformity.

The risk factors for development of deformities are as follows

1. BT cases with more than five lesions
2. PNL cases with more than two or more nerve trunk involvement
3. LL cases

In this study proportion of BT cases and PNL cases were higher compared to other forms of leprosy. This explains the increased incidence of deformities in this study.

Type of deformity

Claw hand: In a study conducted by Srinivasan H⁽²³⁾ reported 21.5% of the patients had claw hand deformity. A study conducted by Sow SO⁽²⁴⁾ reported 33% had claw hand deformity in their study.

In this study proportion of claw hand was 38% which was higher compared to other visible deformities.

The probable reason for this increased incidence of claw hand in this study is explained as follows:

Paralytic deformities of hand occur because of destruction of motor fibers in the major nerve trunks supplying the intrinsic and extrinsic muscles of hand. In leprosy the ulnar nerve is damaged most often followed by median nerve, hence ulnar claw hand is the most common deformity followed by total claw hand.⁽²⁵⁾

So, possible reason for increased incidence of claw hand deformity in this study may be due to increased frequency of ulnar nerve involvement in leprosy.

Trophic ulcers: A pilot project conducted on leprosy by Jagannathan,⁽²⁶⁾ 25% had trophic ulcers of the foot. A study conducted by Sow SO,⁽²⁷⁾ 11% had trophic ulcers of foot. In this study proportion of plantar ulcer was 38% which was higher compared to other studies.

The reason for this increased incidence of plantar ulcers in this study was due to the fact that, a large proportion of cases had anesthesia of feet and foot ulcers are among the most common foot problem seen in leprosy patients.

Anesthesia: In this study, the proportion of cases with anesthesia (Grade 1 deformity) was 24%. The reason for this is increased proportion of BT and PNL cases, where in involvement of nerves is early with extensive destruction.

Lagophthalmos: In a study conducted by Kar et al,⁽¹⁸⁾ 8% of the patients had lagophthalmos and Srinivasan H⁽²³⁾ reported lagophthalmos in 10% of his study

subjects. In a study conducted by Courtright et al,⁽³⁰⁾ lagophthalmos was observed in 3.3% of cases.

In this study number of patients with lagophthalmos was 6%.

The reason for decreased incidence of lagophthalmos in this study appears to be due to decreased number of patients with facial lesions and less number of LL cases.

Foot drop: A pilot project done by Jagannathan,⁽²⁶⁾ foot drop was observed in 7.76% of the patients. In a study conducted by Sow SO113, 11% had foot drop.

In the present study, 2% of the patients had foot drop. In the lower limb the common peroneal nerve in the popliteal region and the posterior tibial nerve lower down in the leg are affected very often. About 2% of leprosy patients develop foot drop because of damage to common peroneal nerve in the popliteal region.⁽²⁵⁾ The low occurrence of cases of foot drop in our study could be a normal variation in the clinical presentation.

Other deformities: In this study, 16% had auto amputation of digits, 4% had corneal opacity, 4% had ape thumb, 2% had madarosis, 2% had leonine facies, 2% had banana fingers, 2% had premature senility and 2% had Buddha ears.

Sensory function deficits are more commonly seen in patients in lepromatous end of the spectrum. In BT Hansen's disease sensory functions are affected only in patients with patches on hands and feet.

Frequency of motor deficits like claw hand and lagophthalmos is common in BT leprosy followed by pure neuritic leprosy. This is because, even the number of nerves involved in BT leprosy is lesser, but the severity of nerve damage is high compared to lepromatous leprosy. Also type 1 reaction occurring in patients without the initiation of treatment is common in BT leprosy.

In our study, 12 patients presented in reaction. Out of 12 patients, 7 (14%) patients presented with Erythema nodosum leprosum, 5 (10%) patients presented with type 1 reaction.

Proportion of patients with grade 2 disability was higher than with grade 1 disability. At National level to assess program effectiveness only grade 2 disability record is maintained but for prevention of disability, grade-1 assessment is more important. Because, before visible deformity (grade-2) occurs, nerve function impairment definitely occurs (sensory, motor or both) i.e. those patients with G2D must have passed through the stage of grade-1. Therefore, while examining any leprosy case, after examination of skin lesions, thorough neurological examination of peripheral nerves is essential. For this, assessment of sensory NFI, motor NFI are needed along with nerve palpation for thickening, tenderness and reactions for detection of grade-1 disability.

The most important factor to prevent disability in leprosy patients is early detection and adequate treatment of neural impairment. Registration delay is a

recognized risk factor for disability in leprosy, but is the result of complex interactions between physical, social, economic and psychological factors.

Among the studied 50 patients, disability was more among those who were manual labourers (36%) followed by farmers (32%) than patients engaged in other occupations like business persons and housewives etc. Leprosy patients, as a result of nerve involvement suffer from loss of sensation in hands and/or feet and often with motor weakness. This increases their vulnerability towards injuries, burns, thorn prick, etc. specially who were engaged in manual occupation or had to walk for long distances.

In leprosy, eyes, hands and feet are the commonly affected areas of impairment even in advanced stages. In this study, hands and feet were more commonly involved when compared to eyes. Sensory NFI was the commonest nature of disability found in feet (Trophic ulcer) and motor NFI in hands (Claw hand). In eyes, lagophthalmos and corneal opacity were the commonly found deformities. These findings emphasize the importance of routine assessment of NFI of all leprosy patients to search for grade-1 disability. After diagnosis if we can properly educate these patients for self-care like not to walk barefoot, daily inspection of hands & feet for any blisters, red spots, physiotherapy, eye care, change in occupation, etc. No doubt surgery plays a major role with respect to motor function and appearance, provided it is performed in suitable patients. However, it does not influence sensory loss and therefore patients should be instructed in care of hands and feet.

This study is mainly for information on disability and deformity in leprosy. It's not meant to focus detection of new cases.

Conclusion

In conclusion it can be said leprosy still continues to be a disease which is commonly seen in low socio-economic group of people, probably because of the poor knowledge of the disease among illiterates, leprosy continues to be a domestic, national and global burden.

The words of Latapi, the renowned Mexican Leprologists, quoted by Frenken in his book, 'Leprosy cannot be completely rooted out with physicians, control offices, leprosaria and propaganda; it will disappear when the economic and cultural factors change, because leprosy is the thermometer of civilization.'

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