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A clinical study of the cranial nerve involvement in leprosy. .

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Abstract

Background: Leprosy commonly affects the cranial nerves predominantly the 5th (trigeminal nerve) and the 7th (facial nerve). Lepra reactions are risk factors for cranial nerve involvement.

Objective: To study the frequency and pattern of cranial nerve involvement in leprosy and to find its relation with facial patch.

Patients and methods: The present clinical study was undertaken on 100 consecutive leprosy patients to find out the involvement of cranial nerves in leprosy and to study the relationship between cranial nerve involvement and leprosy patch/patches on facial skin.

Results: Cranial nerve involvement was detected in 22 patients on clinical grounds; 7 Borderline Tuberculoid (BT), 6 Lepromatous Leprosy (LL), 6 Borderline Lepromatous (BL), 1 Pure Neuritic (PN), 1 Tuberculoid Tuberculoid (TT) and 1 Borderline Borderline (BB). The most commonly involved cranial nerves were the facial and trigeminal, seen in 9% each; followed by the olfactory in 6% and the auditory in 3%. Most cases with facial and trigeminal nerve involvement were of BT leprosy types while the majority with olfactory and auditory nerve involvement was of the lepromatous leprosy type (BL, LL). The association between lagophthalmos of recent origin, type 1 lepra reaction and significant facial patch was statistically significant.

Introduction

Leprosy is the most common cause of treatable peripheral neuropathy in India and probably also in the world because of the large number of affected individuals perhaps closely matched by diabetic neuropathy [14]. A cardinal sign is sensory loss which always precedes paralysis in all types of leprosy [17]. Leprous neuropathy is characterized by involvement of superficial nerve trunks in areas such as ulnar, radial cutaneous, median, common peroneal, supra orbital and greater auricular which are cool and liable to trauma [10]. Any cranial nerve can be affected predominantly the 5th and the 7th [15]. The zygomatic branch of facial nerve which supplies the orbicularis oculi muscle is the most frequently affected [16]. Appearance of type 1 reaction puts a patient at risk of nerve damage and secondary deformities [6]. Early recognition and medical treatment of early nerve damage with steroids may result in full restoration of nerve function. The presence of significant facial patch around eyes or over malar region together with type 1 lepra reaction is a severe risk factor for the development of lagophthalmos and paralysis of other facial muscles [6]. Hypaesthesia and anaesthesia are most often observed in maxillary division of the 5th nerve [16]. In most cases of facial nerve involvement in leprosy, there is sensory impairment or hypopigmented and hypoanesthetic patch as well in the territory of trigeminal nerve especially over the maxillary branch. It becomes conceivable that leprosy infection entering the malar skin through sensory fibres progresses in such a way that it involves the peripheral motor branches of facial nerve in the area [5]. Sensorineural hearing loss is more in lepromatous leprosy with ENL (Erythema Nodosum Leprosum) reaction [2]. Involvement of nasal mucosa is greatest in lepromatous leprosy. The present study was undertaken to assess the frequency and pattern of involvement of cranial nerves, to characterize the type of leprosy that may be associated with damage of cranial nerves and to unravel some characteristics that were not explored in the past.

Material and methods

One hundred consecutive leprosy patients diagnosed on the basis of skin lesions, nerve involvement, slit skin smear examination and histopathological examination enrolled / attending the urban leprosy centre were screened for cranial nerve involvement irrespective of the type, duration or treatment status of the disease. Detailed history and complete clinical examination of each patient was performed with respect to age, sex, duration of disease, number, morphology and distribution of skin lesions including facial patch if any, nerve involvement with particular emphasis on cranial nerve involvement, duration of cranial nerve involvement, type of leprosy and treatment status. Ophthalmological and ENT consultations were conducted wherever required. Patients with complaints like nasal stuffiness, deviated nasal septum, intranasal adhesions and scarring in nasal mucosa (i), cataract (ii), history of head injury (iii, iv, vi, vii), acute/chronic ear discharge/drug intake such as aminoglycosides, salicylates, antiepileptics, tranquillizers, diuretics or family history of hearing loss (viii), history of diabetes, hypertension, renal impairment, anaemia etc were excluded because related cranial nerve couldn't be assessed accurately in them.

Individual cranial nerves were tested clinically for sensory, motor and special functions. No specialized laboratory or electrophysiological tests were conducted

(except audiometry for confirmation of sensorineural hearing loss). The recorded data was tabulated and analysed using chi square test.

Results and observations

The study material included 76 males and 24 females in the age ranging from 10 to 70 years. Twenty-two patients had cranial nerve involvement; 9 (18%) of 50 PB and 13 (26%) of 50 MB cases. Most of the patients with cranial nerve involvement were in age group 21-40 (50%) with a mean age of 42 years and male female ratio of 3.4:1.

The majority of cases were of BT (32%) followed by LL (25%), BL (20%), TT (9%), PN (9%) and BB (5%). Thirty percent of BL patients had cranial nerve involvement followed by 24% LL and 22% BT.

Cranial nerve involved	Type of leprosy						Total
	PN	TT	BT	BB	BL	LL	
Facial	-	-	4	1	2	2	9
Trigeminal	1	1	4	-	2	1	9
Olfactory	-	-	1	-	2	3	6
Auditory	-	-	1	-	1	1	3
Total	1	1	10	1	7	7	27

Table 1: Cranial nerve involvement across the spectrum.

Facial and trigeminal nerves were the most commonly involved cranial nerves. On analysis of spectrum of leprosy, 4 out of 9 cases of facial nerve involvement were seen in BT, 2 each in BL and LL and 1 in BB. Trigeminal nerve involvement was seen in 4 BT patients, 2 each of BL and LL patients and 1 each of PN and TT patients. Involvement of the olfactory nerve was seen in 6 patients, 3 of them had LL, 2 BL and 1 BT. Auditory nerve affection was seen in 3 patients only, 1 each of BT, BL and LL (Table 1).

No of cranial nerves involved	Type of leprosy						Total
	PN	TT	BT	BB	BL	LL	
1	1	1	5	1	5	5	18
2	-	-	1	-	1	1	3
>2	-	-	1	-	-	-	1
Total	1	1	7	1	6	6	22

Table 2: Number of cranial nerves involved and type of leprosy.

One cranial nerve involvement was seen in 18 out of 22 patients, two in 3 (13.64%) patients and three in 1 (4.54%) patient. Overall, 27 cranial nerves were affected in 22 patients (Table 2).

Muscle		MB	PB	Total	%age
Frontalis	Unilateral	1	1	2	22.22
	Bilateral	1	1	2	22.22
	No involvement	3	2	5	55.66
Orbicularis oris	Unilateral	1	2	3	33.33
	Bilateral	2	0	2	22.22
	No involvement	2	2	4	44.44

Table 3: Affected muscles other than orbicularis oculi muscle in 9 lagophthalmos patients (5 MB, 4 PB).

Weakness of the orbicularis oculi muscle (lagophthalmos) was seen in all the 9 (100%) patients with facial nerve involvement. Weakness of the frontalis muscle was found in 4 (44.44%) patients and that of orbicularis oris muscle in 5 (55.55%) patients. Involvement of buccinator and platysma muscles or loss of taste sensation was not detected in any patient (Table 3, Fig 1).

Five patients had some form of unilateral facial palsy and 4 had bilateral affection. In LL and BL cases there was more symmetrical pattern of paralysis (50%) than in BT/BT+ cases (25%) which was not significant ($p > 0.50$).

	No of Patients with recent lagophthalmos	Total
Significant patches	4	12
Other/No patches	3	88
Total	7	100

Table 4: Relationship between recent lagophthalmos and facial patches.

The facial patches were classified as significant in 12 patients and other patches in 25 patients. Significant patch is pale and flat or red and raised lesion (in type 1 reaction), located around the eye or in the malar region at least 3 cm in diameter (Table 4).

Lagophthalmos of recent origin was present in 4 (33.33) of 12 patients with significant facial patches (Fig 2). Patients with facial patch had a statistically significant higher frequency of involvement of facial nerve ($p < 0.02$).

	No of patients with recent lagophthalmos	Total
With type 1 reaction	4	24
Without type 1 reaction	3	76
Total	7	100

Table 5: Relationship between recent lagophthalmos and type 1 reaction

Type 1 reaction was present or apparent during treatment in 24 patients. Lagophthalmos of recent origin (of <1 year duration) was found in 7 patients out of total of 9 patients with lagophthalmos and in 4 patients with type 1 reaction. The association between lagophthalmos of recent origin and type 1 lepra reaction was statistically significant ($p < 0.02$) (Table 5)

	Recent lagophthalmos	No of patches
Significant patch with type 1 reaction	4	8
Significant patch without type 1 reaction	0	4
Total	4	12

Table 6: Relationship between recent lagophthalmos, significant patches and type 1 reaction.

Lagophthalmos was present in 4 (50%) of 8 patients who had both significant facial patches and type 1 reaction. The lagophthalmos was without exception at the same side as patch and in case of large patches covering the whole face, it was often bilateral. Altogether, 4 (57.14%) of 7 patients with recent lagophthalmos had significant red and raised patches in type 1 reaction (Table 6).

Trigeminal nerve affection was seen as loss of corneal reflex in all 9 (100%) patients, hypaesthesia in maxillary area in 4 (44.44%) patients, hypaesthesia in ophthalmic area in 2 patients and hypaesthesia/anesthesia in 2 patients in mandibular area. Five cases had sensory loss in the form of hypaesthesia of face and anesthesia in 1 patient. Motor weakness was not seen in any case.

Impairment of olfaction was found in 6 (6%) cases as asymptomatic hyposmia (1 BT, 2 BL, 3 LL). Changes in nasal mucosa were not seen in any case.

Sensorineural hearing loss was detected in 3 (1 BT, 1 BL, 1 LL) cases only and hearing impairment was discovered on tuning fork testing and confirmed by audiometry. Hearing impairment was bilateral in 1 (BL) and unilateral in 2 other cases. Conductive hearing loss was not detected in any patient. Evaluation of vestibular system by clinical testing did not reveal any abnormality.



Fig 1: Left paresis of frontalis, orbicularis oculi & orbicularis oris with deviation of angle of mouth towards right side.



Fig 2: Significant facial patch in a patient with recent lagophthalmos.

Discussion

The hallmark of leprosy is invasion and inflammation of nerves which are present in all stages of different varieties of the disease. Although its overall prevalence is decreasing, leprosy continues to be a major cause of peripheral neuropathy worldwide. Although leprosy usually affects the superficial nerves, yet any nerve in the body including the cranial nerves can be affected. It is important to note that changes secondary to cranial nerve impairment can be as trivial as loss of smell which seems to be of minor importance to the patient to as severe as disfigurement and disabilities such as blindness and deafness.

Twenty- two (22%) of the 100 consecutive patients in present study had cranial nerve involvement. The 5th and 7th nerves were most frequently affected. Thappa et al (2004) and Kumar et al (2006) reported incidences of 22% and 18% with facial nerve involvement of 10% and 9.8% respectively. Incidence of cranial nerve involvement was more in multibacillary leprosy (26%) than paucibacillary leprosy (18%) which was not statistically significant ($p>0.50$). This was attributed to probably longer duration of disease in multibacillary patients.

The majority of our patients with facial nerve involvement had BT leprosy. Various studies [11,15,16,19] found majority of their patients with facial palsy in BT. Lagophthalmos was seen in all our 9 patients (100%) with facial nerve involvement. This could be because of the superficial location of the zygomatic branch and large, red, facial patches in malar region or around eye in 4 of our patients with lagophthalmos.

Out of 9 patients, 3 (1 PB, 2 MB) had developed bilateral facial palsy. Lubbers et al (1994) stated that in LL and BL leprosy cases, there is symmetrical pattern of paralysis and in BT/BT+ cases, pattern is asymmetrical. This observation was also made in our study, but was not significant ($p>0.50$) possibly due to lower number of cases. We did not find weakness of buccinator or platysma in our study comparable to other studies [3,4,19].

It is well known that the appearance of type 1 reaction puts a patient at risk of nerve damage and secondary deformities [6]. The relationship between lagophthalmos of recent origin and type 1 reaction was statistically significant ($p>0.02$) in the present study. Facial nerve involvement in leprosy occurs only when there are coexisting skin lesions on face and there is centripetal involvement of branches of facial nerve [1]. The relationship between lagophthalmos and facial patch was statistically significant ($p<0.02$). Both these observations are comparable to other studies [6,19].

The association of facial nerve involvement, facial patch and type 1 reaction was also highlighted by Hogeweg et al (1991). A majority of their patients (85%) with recent lagophthalmos had significant patch over the malar region or around the eye on the same side as nerve damage together with clinical signs of type 1 reaction. In our study 4 out of 7 patients (57.14%) with recent lagophthalmos had significant patch over malar region or periorbital area together with type 1 reaction. Leprosy infection entering the malar skin through sensory fibres progress in such a way as to involve the peripheral motor branches of facial nerve in the area [5].

Thappa et al (2004) and Kumar et al (2006) found trigeminal nerve involvement in 7% and 8% of their patients respectively. Reichart et al (1982) found trigeminal nerve affection in the form of hypaesthesia and anesthesia of face and no motor weakness was detected in any of their patients. The predominant manifestations were loss of corneal reflex in all our 9 patients, similar to that observed by Konuncu et al (1994) and Ramadan et al (2001).

Reichart et al (1982) found the majority of patients with trigeminal nerve affection in BT leprosy. The pattern of hypaesthesia and anesthesia in our study were comparable to that of facial nerve lesions since frontal and maxillary divisions were also often affected. As in facial paralysis, most cases of hypaesthesia and anesthesia were seen in BT.

Olfactory nerve involvement was seen as hyposmia in 6 patients (6%), majority (3 cases) belonged to LL. Kaur et al (1979) found anosmia in 4% patients all with LL and Thappa et al (2004) observed olfactory loss in 9% cases. This could be attributed to longer duration of disease in lepromatous leprosy.

Auditory nerve involvement (cochlear part) was seen in 3 (3%) of our patients. Most other studies [2,9,13,15,18] observed higher frequency of cochlear nerve impairment whereas Thappa et al (2004) and Kumar et al (2006) detected 3% and 2% cochlear nerve involvement respectively. However, it should be noted that audiometric testing was not performed in our patients that could be limitation in assessing the exact involvement of auditory nerve. Koyuncu et al (1994) observed 11.1% of their patients with vestibular dysfunction which is contrary to other studies [2,8,13] including ours.

Among 22 patients with cranial nerve involvement, only those with facial nerve involvement knew the duration of involvement of nerves. The duration of facial nerve involvement ranged from 3 months to 12 years with mean duration of 1.91 years. Seven out of 9 patients had lagophthalmos of less than one year duration which was comparable to Hogeweg et al (1991) who studied only those patients with history of recent lagophthalmos of less than one year duration. Other patients were not aware of duration of their disabilities.

Conclusion:

One should thoroughly examine the cranial nerve functions in every case of leprosy as this commonly affects cranial nerves. All leprosy patients, especially those at increased risk (significant facial patch in periorbital region, lepra reactions) should be monitored from the outset of the disease in order to detect nerve damage early and to prevent permanent loss of function.

References

1. Antia NH, Divekar SC, Dastur DK. The facial Nerve in leprosy. Clinical and operative aspects. Int J Lepr. 1966; 34: 103-17
2. Awasthi S, Singh G, Dutta R, et al. Auriculovestibular nerve involvement in

leprosy. Indian J Leprosy. 1990; 6

3. Bruce M, Richard, Peter R Corry. Cervical branch of facial nerve in leprosy. Int J Lepr other Mycobact Dis. 1997; 65:170-177

4. Crawford CL and Hobbs MJ. Why is the cervical branch of facial nerve not enlarged in leprosy? J Anthes. 1994; 184: 188

5. Dastur D.K. Pathology and pathogenesis of predilective sites of nerve damage in leprosy neuritis. Nerves in the arm and the face. Neurosurg Rev. 1983; 6: 139-152

6. Hogeweg M, Udaya Kiran K, Sujai Suneetha. Significance of facial patch & type 1 reaction for development of facial nerve damage in leprosy. A retrospective study among 1226 paucibacillary patients. Lepra Review 1991; 62: 143-149

7. Kaur S, Malik SK, Kumar B, et al. Respiratory system involvement in leprosy. Int J Lepr, 1979; 47: 48

8. Kokhar DK, Gupta DV, Chauhan S, et al. Study of brain stem auditory evoked potentials and visual evoked potentials in leprosy. Int J Lepr other Mycobact Dis, 1997; 65: 157-165

9. Koyuncu M, Celik O, Ozturk A, et al. Audiovestibular system, fifth and seventh cranial nerve involvement in leprosy. Indian J Leprosy, 1994; 66: 421-428

10. Kumar Sudhir, Alexander Mathew, Gnanamuthu Chandan. Cranial nerve involvement in patients with leprosy neuropathy. Neurology India, 2006; 54: 283-285

11. Lemieux L, Cherian TA, Richard B. The stapedial reflex as a topographical marker of proximal involvement of the facial nerve in leprosy. A pilot study. Lepr Rev, 1999; 70: 324-332

12. Lubbers WJ, Schipper A, Hogeweg M, et al. Paralysis of facial muscles in leprosy patients with lagophthalmos. Int J Lepr other Mycobact Dis, 1994; 62: 220-224

13. Mann SBS, Kumar B, Yande, et al. Eighth nerve evaluation in leprosy. Indian J Leprosy. 1987; 59: 20-25

14. Paul JT, North-Wilhelm K, Higdon GA, et al. Multiple cranial neuropathies associated with leprosy. South Med J, 1994; 87: 937-940

15. Ramadan W, Mourad B, Fadel W, et al. Clinical, electrophysiological and immunopathological study of peripheral nerves in Hansen's disease Lepr Rev, 2001; 72: 35-49

16. Reichart PA, Srisuwan S, Metah D. Lesions of the facial trigeminal nerve in leprosy. Int J Oral Surg, 1982; 11: 14-20

17. Schuring AG and Istre, Hansen's disease and hearing. Arch otolaryngol, 1969; 89: 478-481

18. Singh TR, Aggarwal SK, Bajaj AK, et-al. Evaluation of auriculovestibular status in leprosy. Indian J Leprosy. 1984; 56: 24-29

19. Thapa DM, Gopinath DV, Jaishanker TJ. A clinical study of the involvement of cranial nerves in leprosy. Indian J Leprosy, 2004; 76: 1-9

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