Eighth Cranial Nerve Affection in Leprosy¹

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Leprosy is a systemic disease which has a marked predilection for skin and nerves. The enormous preponderance of bacilli in the peripheral nerves compared with other tissues and viscera favors a selective affinity of these bacilli for the peripheral nerves (7). Khanolkar (6) stressed that even in the skin lesions, the basic changes are in or around the cutaneous nerve fibers. Fite (5)effectively stressed the neural involvement with the statement, "To the histopathologist all leprosy is neural leprosy." It is established that in all types of the disease, there is invasion of the peripheral nerves by lepra bacilli but the probability of damage resulting from this invasion is earliest and greatest in tuberculoid leprosy.

According to Brand (2), the peripheral nerves which are frequently affected and the pattern of nerve damage throughout the body is remarkable constant. On the motor side, leprosy implicates particularly the ulnar nerve, the median nerve, the common peroneal nerve, the posterior tibial nerve and very occasionally the radial nerve. Sensory derangement follows very much the same pattern but in addition to the mentioned nerves, there is involvement of the great auricular nerve (which is also motor to the pinna and parotid gland) and the long cutaneous nerves that run down from the upper arm to supply the forearm and hand and those that run down the thigh to supply the leg below the knee. There is, as well, irregular patchy anesthesia that occurs anywhere in the body and results from involvement of the fine cutaneous nerve fibers.

There is apparently very little stress in

the literature on cranial nerve affection except for the trigeminal and facial nerves. It is even stressed by Cochrane (³), that the eighth nerve, as far as is known, is never affected in leprosy. His descriptions of the ear lesions were only restricted to the pinnae which occasionally showed gross ulceration and deformity.

More recently, however, El Garem and Wahba (⁴) in a clinical study of Egyptian material, reported the involvement of other cranial nerves. They found implication of the eighth nerve together with the fifth (and query the seventh) in one patient. This patient had also sluggish pupillary reactions most probably due to central leprotic lesions rather than to involvement of the optic and oculomotor nerves which form the reflex arc for the pupillary light reactions. In another patient, the same authors found clinical evidence of unilateral partial eighth nerve deafness and, in a third patient, they could detect involvement of the spinal accessory nerve of one side.

It is evident, therefore, that only few cases of eighth nerve involvement have been reported in the literature. However, it is an observation that in the past few years, a good number of leprotic patients have been referred to the Ear, Nose and Throat Department with a complaint of hearing loss. They have always been attributed to one of the numerous well-known causes of loss of hearing. The possibility of a relationship of these cases to the leprotic process was not suspected till Abdel Latif in his thesis (1) reported the occurrence of eighth nerve hearing loss in as much as 25 per cent of a series of 60 leprotic patients and related this condition to leprosy. It seemed appropriate to submit this finding to a more detailed investigation which is the aim of this work.

MATERIALS AND METHODS

One hundred and two randomly selected patients were the subjects of this study.

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Ten cases were inpatients in the dermatology department of Alexandria University Hospital (9 males and 1 female). The remaining 92 cases were inpatients in Amreya Leprosarium where only males are admitted.

In all cases, the diagnosis of leprosy was established by clinical examination and confirmed by appropriate laboratory tests. They were further studied as regards their residence, occupation, duration of their illness, its clinical type, the presence of any associated condition and the results of the latest bacteriologic examination.

The patients were also examined with reference to the ear, nose and throat with particular stress on the condition of the drum, middle ear and Eustachian tube. A search was also made for septic foci in the nose, sinuses, teeth and throat. Tuning fork tests (Rinne, Weber and Schwabach tests), speech tests (for conversation speech and whispered voice) and pure tone audiometry were done for all of them. Patients who proved to have perceptive hearing loss received further clinical and laboratory study. Their present, past and family history were inquired into and detailed otolaryngologic and general examination was made aiming at finding out any probable cause for their loss of hearing. Leading questions were always asked for the subjective sensations of diplacusis, hyperacusis, distortion or fullness in the ear. They also received special audiometric study, namely conventional pure tone audiometry in completely silent surroundings with the use of masking whenever indicated. The pattern of the audiogram obtained was noted and the percentage of hearing loss was calculated according to the method recommended by Western Electric Company (10) in which the percentage of hearing loss is equal to the mean of hearing losses at 512, 1024 and 2048 c.p.s. multiplied by 0.8. The phenomenon of recruitment was tested for using the binaural loudness balance method in patients with unilateral perceptive hearing loss and the Denes and Nauton method (10), for the other patients with bilateral hearing loss.

The loudness adaptation phenomenon

was tested simply by the pure tone audiometer as regards the sound intensity.

To assess the function of the vestibular component of the inner ear, cold and hot caloric testing was done by the use of Cawthorne's caloric apparatus and the Hallpike procedure.

RESULTS

Studied cases. One hundred and two patients (101 males and one female). Their ages ranged between 11 and 85 years.

Occupation. Most patients were farmers or laborers with a low income and poor hygienic conditions of living.

Residence. This group of patients represented nearly all localities of Egypt but a relatively larger number came from Upper Egypt, especially Kena and Sohag.

Duration of the disease. A marked variation in the duration of the disease was noticed among the group. The duration periods ranged between four months and 40 years.

Clinical types.

Lepromatous:	41	patients
Tuberculoid:	33	patients
Borderline (intermediate):	28	patients

acteriologic exan	nination (na	asa	sm	iears).
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(intermediate): Positive in 18 patients Hearing loss cases: 16 patients.

Males: 15 Females: 1

Ages: Ranging between 17 and 66 years Type of hearing loss: Perceptive: 15 cases Mixed: 1 case

In addition there was one case of conduction hearing loss due to right chronic suppurative otitis media and as the cause is evident, it was excluded.

Side affected:

Both ears:	14	cases
One ear:	2	cases

Onset and duration of hearing loss. Slowly progressing loss of hearing in all cases. In bilaterally affected ears, it started simultaneously in both ears. The duration of hearing loss varied between one and 18 years, without any relation to the time of cnset or duration of the disease.

Other probable etiologic factors.

Prolonged streptomycin therapy: 3 cases

Senility (above 65): 3 cases

Diabetes mellitus: 1 case

Septic foci in sinuses, teeth and tonsils: 4 cases.

The antileprotic drugs received by these patients were sulfone preparations, which are not known to have any side effects on hearing or on the eighth nerve.

The past and family history were irrelevant as far as loss of hearing is concerned.

Audiologic signs and symptoms. Loss of hearing: in one case, there was no complaint of hearing trouble and the condition was detected by examination; in three cases there was a complaint of difficulty in distinguishing certain words of speech and the remaining 12 cases gave a complaint of difficulty in hearing.

Tinnitus was the main symptom in nine cases (bilateral in 3, unilateral in 6). Tinnitus was continuous and of the high pitch type.

No patient gave a complaint of vertigo, unsteadiness, or other signs of vestibular involvement.

Eustachian tube function was obstructed in two cases. In one of these, obstruction was bilateral and the patient showed bilateral mixed loss of hearing.

Ear drum. Bilateral chronic suppurative otitis media in one case.

Diplacusis: no case

Hyperacusis: no case

Fullness in the ear: two cases

Associated speech changes: four cases had loud talking with slurring during speech.

Hearing tests:

Tuning fork tests.

- Rinne's test: positive in 15 and negative in one.
- Weber's test: localized to the better side in nine and no lateralization in seven cases.
- Schwabach's test: shortened in all cases.

Speech tests. Conversational speech and whispered voice were heard at lesser distances than 40 and 20 feet respectively in all cases.

Pure tone audiometry. All audiograms in 16 cases showed different degrees of perceptive deafness. In one case only the air conduction hearing was much more affected than the bone conduction hearing giving the picture of mixed loss of hearing. The 16 audiograms which were obtained could be classified according to the type and grade of hearing loss into:

- Gradual hearing loss: 4 cases (bilateral and symmetric).
- Low tone loss: 2 cases (one unilateral in the left ear and one bilateral and symmetric).

Flat hearing loss: 10 cases.

No other special grades were found among the patients.

Percentage of hearing loss. Calculated according to the Western Electric Company method (10), this was found to range between mild hearing loss (5%) and severe hearing loss (55%). In nine cases of the 14 bilaterally affected patients, the percentage of hearing loss was nearly equal in both ears but in the other five cases, great variations were detected.

Recruitment. This was absent in all the deaf patients, even in those patients who had received long courses of streptomycin injections, denoting an underlying perceptive nerve hearing loss.

Loudness adaptation phenomenon. Present in all members of the group showing loss of hearing with varying degrees of marked adaptation.

Vestibular system. Caloric testing by the Cawthorne's caloric apparatus was done for the group with loss of hearing and revealed a reduced or absent reaction to both cold and hot water, denoting an affected or nonfunctioning vestibular apparatus.

Correlation of the auditory and dermatologic findings. In the 16 patients with perceptive and mixed loss of hearing, leprosy had been present for a period varying between one and 40 years. They showed the clinical picture of lepromatous leprosy in six cases. Bacteriologic examination of nasal smears from these patients yielded positive results in seven cases and negative finding in nine cases. The ages, duration of leprosy, duration of hearing loss clinical type, bacteriologic data and the percentage

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of perceptive hearing loss are given in Table 1.

Although pathologic examination of the eighth nerve in the patients with loss of hearing would be highly desirable, this has not been possible since no deaths occurred among patients during this study.

DISCUSSION

The present series of 102 leprotic patients who were studied in a nonselected manner represented the main features of the disease with its onset in young ages without apprently influencing the life span (8), its high endemic nature in Upper Egypt particularly in Kena and Sohag and its greater prevalence among the low social class individuals.

The series included 41 patients with lepromatous leprosy, 33 patients with the tuberculoid type and 28 cases of borderline leprosy. The patients displayed marked variation in the duration of the disease which ranged from four months to 40 years.

Bacteriologic examination of nasal smears yielded positive results in 38 cases of the lepromatous type, 11 cases of the tuberculoid type and 18 patients with borderline leprosy.

Careful study revealed that 16 patients

Age No. (yrs.)	ge Clinical s.) Sex type		Duration of leprosy (yrs.)	Duration of hearing loss	Per cent of	Result of		
		Clinical type			Right ear	Left ear	bacteriologic examination	
1	17	male	Leprom.	2	1 yr. (Lt. ear)	15	15	Positive
2	66	male	Tuberc.	1.5	6 mos. (Bilat.)	17	16	Negative
3	57	male	Leprom.	3	1.5 yr. (Bilat.)	10	10	Positive
4	25	female	Tuberc.	1.5	5 mos. (Bilat.)	24	24	Negative
5	-50	male	Leprom.	4	3 yr. (Bilat.)	60	65	Positive
6	65	male	Tuberc.	30	6 mos. (Bilat.)	15	17	Negative
7	57	male	Borderline	10	4 yr. (Lt. Ear)	Normal	29.2	Positive
8	42	male	Tuberc.	30	12 yr. (Lt. Ear)	20	Normal	Negative
9	52	male	Leprom.	8	5 yr. (Bilat).	12.8	20.8	Positive
10	35	male	Borderline	8	5 yr. (Bilat.)	32.8	32.8	Negative
11	35	male	Tuberc.	15	5 yr. (Bilat).	25	25	Negative
12	16	male	Leprom.	1	1 yr. (Bilat.)	37	14	Positive
13	50	male	Leprom.	20	1 yr. (Bilat.)	17	20	Positive
14	66	male	Tuberc.	40	3 yr. (Bilat.)	27	27	Negative
15	38	male	Borderline	25	18 yr. (Bilat.)	23	52	Negative
16	46	male	Tuberc.	12	3 yr. (Bilat.)	45.8	40	Negative

TABLE 1. Correlation of the auditory and cutaneous findings.

had loss of hearing. The group of patients with loss of hearing represented the various clinical types, they were of different ages and had been suffering from leprosy for varying periods, meaning that no rule could be put concerning the time of occurrence of hearing loss or any age predilection.

Loss of hearing was diagnosed as percepcive by the tuning fork test and pure tone audiometry. The absence of recruitment or abnormal loudness adaptation and the lack of subjective sensations of diplacusis, hyperacusis or hearing distortion excluded the possibility of inner ear disease as a cause for this perceptive loss of hearing. It was also not a brain stem lesion as this gives severe or total bilateral organic loss of hearing besides other paralytic manifestations in the body (°). Hence, the possibility of eighth cranial nerve disease must be considered.

A careful search has been made by all possible means for any etiologic factor that could explain this relatively high percentage of perceptive deafness in leprotic patients. There were three elderly patients (above 65 years), an age with a high incidence of senile loss of hearing (9), but the detection of hearing loss in 13 patients of younger ages gives no weight to the possible effect of senility as a cause for the high incidence of loss of hearing in our group of patients with hearing losses. One patient, aged 32 years, has diabetes mellitus. However, the young age of this patient and the recent onset of diabetes mellitus rules out the possibility of this latter disease as a significant etiologic factor for his loss of hearing. Four out of the 16 patients with loss of hearing had septic foci in the nose and throat, but this percentage is of little significance as it is nearly similar to that present in the general population; besides the effect of septic foci on the eighth nerve is doubtful.

The degree of hearing loss was mild in some cases and severe in others without any relation to the age of patients or duration of the disease, a finding which might indicate that eighth nerve affection might be a matter of predisposition and the degree of affection, or its rate of progress, depend on this susceptibility. Affection of

both the hearing power and the vestibular function suggests that there is a universal affection of the eighth cranial nerve which is responsible for the perceptive nerve loss of hearing and the failing vestibular function. It has been mentioned that the clinical and audiologic findings suggest no affection of the inner ear, a finding which conforms with the experience of Cochrane (³). This presumably means that the leprotic process affects the eighth nerve but not the sensory receptors in the inner ear. Such a tendency can find its explantion in the conclusions of Lumsden (7) that the bulk of bacillary spread and multiplication occurs along the Schwann cells enveloping the individual nerve fibers and not axonally. It is known that Schwann cells are present, surrounding the eighth nerve, but not around the receptors. The pathology of leprotic nerve affection consists of cellular infiltration and edema which causes relative ischemia of the nerve leading to its gradual destruction with irreversible damage $(^{2})$. This is possibly the sequence of events in the pathogenesis of eighth nerve affection.

CONCLUSIONS

Hearing may be impaired due to eighth nerve affection in leprosy without any relation to the age of the patients or the duration of the disease.

As much as 15.7 per cent of leprotic patients are expected to suffer from perceptive loss of hearing, a percentage which is rather high and shows the great tendency of the disease to implicate the eighth cranial nerve. This is in contrast to what has been previously reported in the literature.

The main pathogenesis of this perceptive loss of hearing is selective bilateral eighth nerve affection, without any detectable lesions in the middle ear, inner ear, or the central connections of hearing.

Involvement of the eighth nerve alone without any changes in its terminal fibers in the inner ear, or its central connections in the brain stem, is perhaps due to the fact that leprosy affects the Schwann cells enveloping the individual nerve fibers and not the naked axons or brain tracts.

When the eighth nerve is affected, it

probably suffers from relative ischemia which leads to its gradual destruction, this explains the gradual onset and the progressive course in all affected cases.

SUMMARY

A study of 102 leprotic patients from the dermatologic and otolaryngologic point of view revealed a relatively high incidence of eighth cranial nerve affection. The studied group presented all ages, different clinical types and stages of the disease and represented an appropriate material for the study of this subject.

Loss of hearing was proved to be perceptive in type and apparently due to eighth cranial nerve affection and not to any other pathologic changes.

RESUMEN

Un estudio de 102 enfermos de lepra desde los puntos de vista dermatológico y otolaringológico reveló una incidencia relativamente alta de compromiso del octavo nervio craneano. El grupo estudiado comprendía todas las edades, tipos clínicos distintos y etapas de la enfermedad y representaba un material apropiado para el estudio de este tema.

Se comprobó que la pérdida de la audición era de tipo perceptivo y que se debía aparentemente al compromiso del octavo nervio craneano y no a ningún otro cambio patológico.

RÉSUMÉ

L'étude de 102 malades atteints de lèpre, du point de vue dermatologique et otolaryngologique, a révélé une incidence relativement élevée d'atteinte du huitième nerf crânien. Le groupe étudié comprenait tous les âges, des types cliniques différents, des stades différents de l'affection, et constituait un matériel approprié pour l'étude de ce sujet.

La perte d'audition s'est révélée être d'un type perceptif; elle était apparamment due à une affection du huitième nerf crânien, et non à une quelconque autre modification pathologique.

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