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Original Article

Study of patients presenting with symptoms of peripheral neuropathy and thickened greater auricular nerve

Neopane A[1], Upadhyaya B[2], Dungana S[3], Karki DB[4]

[1]Assistant Professor, [2]Registrar, [3]House Officer [4]HOD, Department of Medicine, KMCTH

Abstract

Objective: To analyze symptoms and make a clinical diagnosis of leprosy in patients presenting with symptoms of peripheral neuropathy and found to have thickened greater auricular nerve. **Design:** Cross- sectional study **Setting**: Kathmandu Medical College Teaching Hospital, Sinamangal, Kathmandu. Patients attending the medical out patient department of this hospital were taken in the study. **Materials and methods**: 40 patients presenting with symptoms of peripheral neuropathy and are found to have thickened greater auricular nerve were included. **Results**: Thickened greater auricular nerve and sensory symptoms showed male predominance (97.5%). Mean age of involvement was 28.65 years**.** The symptom most frequently complained of was chest pain (75%), followed by pins and needle sensation 67.5%, burning of the upper extremities, nape of the neck and chest 57.5%, palpitation 45%, disturbed sleep mostly said to be due to burning 35%, sweating 20%, dizziness17.5%, shortness of breath 7.5%, and numbness of the limbs in 2.5%. None of the patients had somatic neuropathy. Autonomic neuropathy was present in 42.5 %.

Conclusions:

1. In endemic areas patients with thickened peripheral nerve and sensory symptoms should be diagnosed clinically as primary neuritic leprosy.
2. In absence of objective loss of somatic sensation autonomic neuropathy may be the only early indicator of neuritis.
3. Close follow up of these patients is necessary.

Key words: peripheral neuropathy, primary neuritic leprosy, autonomic neuropathy, follow up

In general practice we sometimes encounter patients, mostly male presenting with complaints of vague burning sensation over the nape of the neck, anterior chest, pricking type of precordial pain, excessive sweating, dizziness, palpitation, pins and needle over the tip of the fingers without any objective sensory loss. These patients are usually seen by physicians, and even psychiatrists and sometimes referred to the cardiologists. This study involves all such cases who have been symptomatic since at least three months and have taken analgesics, anxiolytics and had all cardiac investigations done including exercise stress test.

One thing common to these patients coming to us with the above symptoms was thickened greater auricular nerve. Leprosy is the commonest cause of thickened peripheral nerve/nerves and peripheral neuropathy in endemic areas[1].

Primary neuritic form of leprosy is a well established though rare form of leprosy[2]. Reports show from studies done in South India that 6% of early leprosy lesion belongs to this type. It shows male predominance[3] with peak incidence in 20-35 years[4]. It is characterized by neuritic manifestations caused by the asymmetrical involvement of usually one or at times several peripheral nerve trunks. Symptoms of sensory impairment, paraesthesia, nerve enlargement with or without tenderness and paresis may occur in varying combination, at times without any objective sensory impairment[5]. Somatosensory and autonomic neuropathy may be the only manifestation of leprosy[6]. It is difficult to diagnose in the absence of hypoaesthetic skin lesion supposed to be pathognomonic of leprosy. Nerve biopsy if can be done is diagnostic but not easily feasible and cases may be missed at the early stage. Disease may progress to higher spectrum and patients may eventually present with sensory and motor deficit and with hypoaesthetic skin lesions that have histological

Correspondence
Dr. Arpana Neopane, Department of Medicine, Kathmandu Medical College Teaching Hospital, Sinamangal. E-mail neopane @vsnl.com GPO 1288

characteristics ranging from indeterminate to tuberculoid to borderline lepromatous leprosy[5]. Lone involvement of peripheral nerve though rare, is reported

This study tries to draw the clinician's attention to the primary neuritic form of leprosy including the clinical presentation of these patients and the need for close follow up which can itself be an effective strategy for diagnosis when diagnostic facilities are not available[5].

Aims and Objectives

To analyze symptoms of patients presenting with unilateral or bilateral greater auricular nerve thickening; to make a clinical diagnosis of primary neuritic leprosy in them and emphasize the need for long term follow up of these patients to confirm the diagnosis clinically.

Design

A cross-sectional descriptive study of patients coming to the out patient department of Kathmandu Medical College Teaching Hospital, a referral hospital, with symptoms of peripheral neuropathy.

Material and method

This study includes patients coming to the medical out patient department of Kathmandu Medical College teaching hospital with symptoms of peripheral neuropathy and are found to have thickened greater auricular nerve (nerve said to be thickened if both visible and palpable). Patients with systemic diseases like diabetes, hypertension, thyroid, renal and hepatic dysfunction were not included. Also patients on drugs like INH, Hydralazine, Phenytoin were excluded from the study. Patients with unilateral or bilateral nerve involvement were included. Patients with multiple peripheral nerve thickening were sent to the leprosy clinic in Patan Hospital, held every week on Wednesdays and were not included in our study. A total of 40 patients were included.

A thorough history of all patients was taken, symptoms were elaborated and noted. Neurological examinations were done. Sensory examination was done in detail including all dermatomes of the body. Temperature sensation was tested with both warm water and ice-cold water. Autonomic function tests were also done using Ewing's battery of cardiac autonomic function tests[7] that includes three parasympathetic and two sympathetic tests. All results are expressed in tabulated form.

Results

Table1 Age distribution

Age in years	Total no.	Percentage of total	Remark
15-30	23	57.5%	Maximum(57.5%) in the 15-30 yrs age group Mean age = 28.65yrs
31-45	17	42.5%	
>46	0	0	

Table2 Sex distribution

Sex	Number	Percentage (%)	Remarks
Male	39	97.5	Male predominance
Female	1	2.5	

Table 3 Symptoms distribution

Symptoms	Present (n=)	Present %	Remarks
Chest pain burning or pricking type	30	75	Symptoms of sensory neuropathy and autonomic neuropathy are both present.
Pins and needle sensation limited to one or both side of upper half of body	27	67.5	
Burning sensation over the nape, chest, upper extremities	23	57.5	
Palpitation	18	45	
Disturbed sleep	14	35	
Sweating	8	20	
Dizziness	7	17.5	
Shortness of breath	3	7.5	
Numbness of the upper limb	1	2.5	

Table 4 Examination of greater auricular nerve

Greater Auricular Nerve	No of patients	Remark
Visible	38 (95%)	Non had tender nerve
Palpable	40 (100)%	
Both/bilateral	38 (95%)/4(10%)	

Table 5 Examination of temperature sensation

Impaired temperature	No of Patients	Remark
Round the neck	0	None of the patients showed loss /impaired sensation
Anterior chest	0	
Other areas and dermatomes	0	

Table 6 Results of autonomic function tests

Tests (abbreviation)	Normal	Abnormal	No of patients With 1+ve tests	No of patients with >than 1+ve tests	Total	Remark
Valsalva ratio (VSR)	>1.21	<1.20	9(22.5%)	8 (20%)	17 (42.5%)	Objective evidence of definite autonomic dysfunction was present in 8(20%)with>1 tests +ve[7]
Expiration/inspiration heart rate variation ratio (HIXEN)	Variation> 15 beats/min	Variation <10 beats/min				
Ratio of $30^{th}/15^{th}$ beat (ECG) on standing (HRRS)	>1.04	< 1.0				
BP response to sustained hand grip (inflated BP cuff) (SHG)	Rise in systolic BP >16mmHg	Less than 10mmHg				
Postural fall in SBP (FSBP)	<10mmHg	>30mmHg			9(22.5%) pts wit one test positive had early autonomic neuropathy as per Ewing's severity classification[7].	

Table 7 Elaboration of autonomic function tests done

Autonomic tests positive in various combinations	No of patients	Symptomatic Y/N (yes/no)	Remark
VSR+HRRS	1	Y	All symptomatic patients complaining of palpitation/sweating/dizziness had >1 tests positive
HEXN+FSBP+SHG	1	Y	
HRRS+VSR+SHG	3	Y	
HEXIN+HRRS+FSBP	1	Y	
VSR+FSB	2	Y	
HEXIN or HRRS or VSR(only one test positive)	3+3+3=9	N	
Total	17(42.5%)		

Out of the 40 patients studied, mean age of affection was 28.65yrs. The age group most involved was the 15-30 years. Males were affected in 97.5% and females only in 2.5%. The symptom most frequently complained of was chest pain (75%), followed by pins and needle sensation 67.5%, burning of the upper extremities, nape of the neck and chest 57.5%, palpitation 45%, disturbed sleep mostly said to be due to burning 35%, sweating 20%, dizziness17.5%, shortness of breath 7.5% and numbness of the limbs in 2.5%. None of the patients had objective evidence of somatic neuropathy. But autonomic dysfunction was seen in 17 patients (42.5%) after evaluating the five autonomic function tests described by Ewing et al[7]. Among these more than one tests were positive in 8 patients (20%) and only one in 9(22.5%). All five cardiovascular autonomic function tests were positive in none of the patients. Parasympathetic autonomic nerves were more involved than sympathetic. None of the patients had lone sympathetic nerve involvement.

Discussion:

This study tries to show that primary neuritic leprosy may not be so uncommon. They may present with common symptoms of peripheral neuropathy and a single nerve thickening[8]. In the absence of objective sensory loss, clinical suspicion may be the only early method of considering the diagnosis. As in diabetic neuropathy, autonomic nerves may however be affected early in this variety of leprosy[9]. One study has even showed autonomic dysfunction to be present in leprosy contacts in endemic region like Nepal and have suggested it to be an early indicator of infection by M. leprae[10]. Patient's symptoms may help to suspect it and simple bedside tests mentioned above may help to confirm autonomic neuropathy as we have done in our study. The definite diagnosis of this form of leprosy is difficult as it manifests without skin lesions. Involved nerve biopsy[10] is the diagnosis of choice which may not always be feasible. In this context clinical suspicion and long term follow up is necessary.

Various studies have shown that after a variable period of time ranging from 4months to 4 years, hypoaesthetic cutaneous lesion may develop and confirm the diagnosis[5,11]. We recommend that these patients should be closely followed up and diagnosed early so that treatment can be instituted.

Conclusions:

In endemic areas like Nepal, patients with symptoms of peripheral neuropathy and thickened peripheral nerve should be clinically diagnosed to have primary neuritic leprosy which is apparently not as uncommon as believed. Autonomic neuropathy may be present in these patients and can be the indicator of early disease. Close follow up of these patients for appearance of skin lesion at the vicinity of involved nerve is helpful in confirming diagnosis and starting treatment thus preventing progression to higher spectrum of the disease with associated morbidity.

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