Bangladesh Journal of Medicine (BJM)
ISSN : 1023 - 1986
eISSN : 2408 - 8366

CASE REPORT

DIAGNOSTIC ENIGMA IN A RARE CASE OF PRIMARY NEURITIC LEPROSY

SHARIF AHMED[1], AMINUR RAHMAN[2], TOFAEL AHMED[1], MD ALAMGIR HOSSAIN[3] , MAHBUBUL HAKIM MISHU[4] AJAY KUMAR AGARWALLA[1], ABUL HASNAT MD.RUSSEL[3], SHAHJADA MOHAMMAD DASTEGIR KHAN [5,] BIPLAB PAUL[5], PALLAB KANTI SAHA[5], MD DAHARUL ISLAM[6]

Abstract:

Leprosy is one of the most prevalent diseases affecting peripheral nerves. Pure neuritic leprosy accounts for 4-10% of all leprosy cases, with clinical symptoms restricted to the peripheral nerves and no skin abnormalities. Leprosy is diagnosed based on the skin and nerves' typical clinical and histological involvement. The absence of typical dermatological characteristics significantly reduces clinical diagnosis accuracy, necessitating histological confirmation. We presented the case of a 26-year-old male with an 8-year history of growing numbness in his right leg, recurrent ulcers in his right foot, and hand abnormalities. The patient was subsequently identified with pure neural leprosy (PNL), a kind of leprosy that affects peripheral nerves but lacks conventional skin lesions. As a result, in leprosy-endemic countries such as Bangladesh, this kind of leprosy should be extensively explored, especially in patients with no skin abnormalities.

Keywords: *Diagnostic enigma, primary neuritic leprosy, mononeuritis multiplex*

Received: 12.08.2024 *Accepted : 22.08.2024*
DOI: https://doi.org/10.3329/bjm.v35i3.75847

Citation: *Ahmed S, Rahman A, Tofael Ahmed T, Hossain MA, Mishu MH, Agarwalla AK. Diagnostic Enigma in a Rare Case of Primary Neuritic Leprosy. Bangladesh J Medicine 2024; 35: 214-217.*

Introduction:

Mycobacterium leprae, often known as Hansen's bacillus, is the causative agent of leprosy, a chronic, communicable granulomatous illness. It has a lengthy incubation period (ranging from nine months to twenty years), with an average of four years for tuberculoid leprosy and eight years for lepromatous leprosy [2]. It is spread from person to person. Because it seems to thrive better at temperatures near 30°C rather than 37°C, the mycobacterium prefers peripheral tissue. As a result, it impacts various tissues like bone and certain viscera, as well as the skin, peripheral nerves, and upper respiratory mucosa. [3]. The rates of new case detection in Brazil, Indonesia, and India were 16 856, 31 044, and 126 913, respectively. Currently, these three are the most prevalent nations, making up around 81% of all new cases globally. Wade in 1952 first proposed the disease "neural leprosy," in which only peripheral nerves were involved. The technical committee of the International Leprosy Congress in 1953, held in Madrid, accepted "neuritic leprosy" as one subtype among the major groups of leprosy. According to clinical case studies, 4–8% of cases of leprosy are thought to be restricted to the peripheral nerves. [6] When both cutaneous and neurological involvements are present at the same time, leprosy diagnosis is simple. Pure neuritic leprosy is when a patient solely has nerve involvement and no visible original skin lesions.[7] Due to the lack of skin lesions,

1. Assistant Registrar, Department of Neurology, Sir Salimullah Medical College Mitford Hospital, Dhaka- 1100, Bangladesh.
2. Professor, Department of Neurology, Sir Salimullah Medical College, Dhaka- 1100, Bangladesh.
3. Registrar, Department of Neurology, Sir Salimullah Medical College Mitford Hospital, Dhaka- 1100, Bangladesh.
4. Resident , Department of Neurology, Sir Salimullah Medical College Mitford Hospital, Dhaka 1100, Bangladesh.
5. Assistant Professor, Department of Neurology, Sir Salimullah Medical College, Dhaka- 1100, Bangladesh.
6. Professor, Department of Medicine, Sir Salimullah Medical College, Dhaka- 1100, Bangladesh.

Address of Correspondence: Prof . Dr. Aminur Rahman, Professor and Head, Dept. of Neurology, Sir Salimullah Medical College, Dhaka-1100, Bangladesh, E-mail: draminur@yahoo.com, https://orcid.org/0000-0001-9261-7734

patients with the "pure neuritic" type of leprosy are commonly misdiagnosed. [8]

We described a case of unusual presentation of mononeuritis multiplex in leprosy, identified by nerve conduction study results that were in line with the histology of nerve biopsy and sensorimotor axonal polyneuropathy, which were diagnosed of PNL.

Case report:

A 26-year-old no-diabetic male presented with a history of progressive numbness in his right leg for 8 years, and recurrent painless ulcers on the right leg and foot for 7 years. Since then, he had been suffering from recurrent ulcers in multiple sites of the right lower leg and foot, like the tip of the great toe, around the head of metatarsals, and along the shin for which he went through both conservative and surgical management several times. He had difficulty walking due to foot drop for the last year. He also reported a deformity of the fingers and loss of sensation in the 4th and 5th digits of the right hand for 8 months, resulting in difficulty holding objects and performing fine motor tasks (Figure 1). No history of fever, cough, arthritis, skin pigmentation, or suspicious skin lesions for leprosy was noted. Examination revealed an ulcer on the right sole (4x3 cm) with irregular margins, a sloughy floor, and serosanguinous discharge (Figure: 2). The right foot showed muscle wasting, reduced tone, and weakness in plantar flexion, dorsiflexion, and eversion (Figure:3). Sensory examination indicated decreased pain, touch, and temperature sensation in the lateral aspect of the right leg and dorsum of the foot, along with impaired joint position and vibration sense on the right side. The left upper & lower limbs exhibited normal motor and sensory function. No signs of systemic illness were observed, with normal vital signs and the absence of fever, cough, arthritis, anemia, jaundice, cyanosis, and clubbing.

Laboratory investigations (Table 1) revealed hypochromic microcytic anemia. Blood sugar, liver& renal function tests were within normal limits. Serology showed VDRL & HIV nonreactive, ANA, cANCA, pANCA negative, X-ray chest was normal and CSF study was normal. Specific investigations were in Table:2 . Slit Skin Smear showed bacterial index (BI)-0 and morphological Index (MI)-1.no AFB found on microscopy, which consisted with multi-bacillary leprosy. A nerve conduction velocity study revealed that CMAP and SNAP amplitudes of the right Ulnar nerves are absent. CMAP amplitudes of right Tibial and Peroneal nerves are absent. SNAP amplitudes of right sural and superficial peroneal nerves are absent. NCS findings of other nerves are normal. These electrophysiological findings are consistent with Sensorimotor Axonal Polyneuropathy affecting some of the studied nerves. Clinical suspicion was Mononeuritis Multiplex. Primary Neuritic Leprosy? vasculitic, ?autoimmune origin.

For confirmation of peripheral neuropathy, a sural nerve biopsy was performed. Histopathological examination showed thick nerve bundle infiltration of chronic inflammatory cell with focal granuloma like area and multinucleated cells and few scattered mast call Special stain (Fite – Faraco)-revealed no acid-fast bacilli and no evidence of malignancy . The diagnosis of primary neuritic leprosy was made. (Figures 2, 3, 4). We started the treatment with Modified WHO-recommended multidrug therapy (MDT) regimens in leprosy

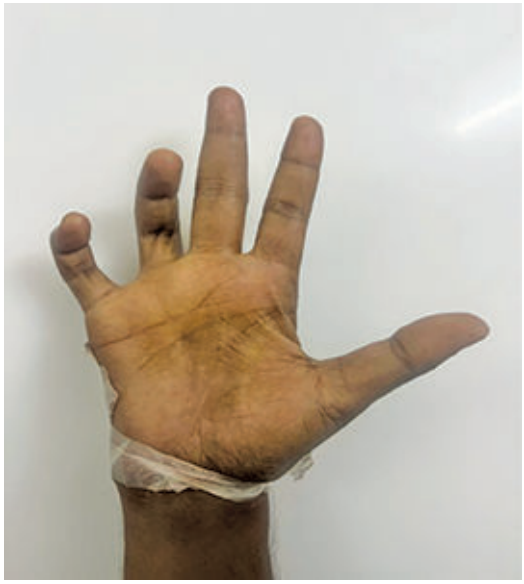

Figure 1: *Ulnar claw hand*

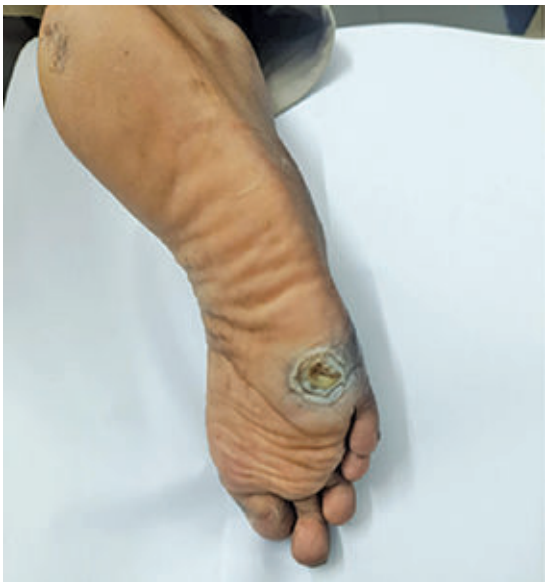

Figure 2: *Trophic ulcer*

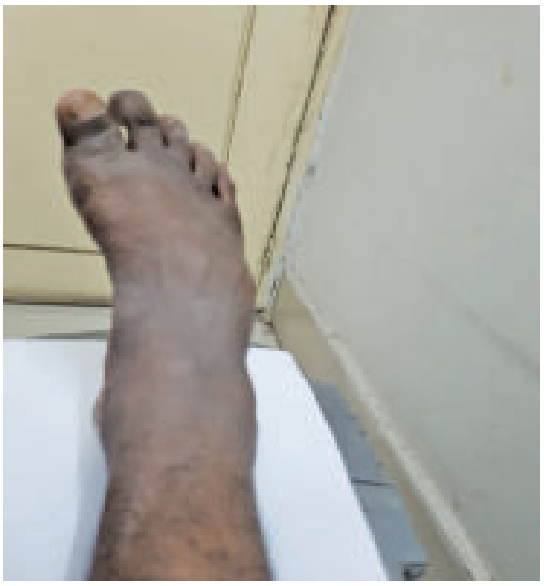

Figure 3: *3Foot drop*

Table-I
Investigations

CBC with PBF	Hb- 11.0 g/Dl, WBC-6.50, Platelet- 354 k/uL Hypochromic microcytic anemia
Serum creatinine	1.09 mg/dL
RBS	7.13 mmol/L
X ray chest	normal
HbA1C	5.9%
Anti HIV 1,2	Negative
ANA	Negative
cANCA, pANCA	Negative
CSF study	Cells 1-2 Llymphocytes and Protein-36gm/dl

Table-II
Specific investigations

Slit Skin Smear	Bacterial Index (BI)-0 and Morphological Index (MI)-1No AFB found on microscopy Impression : multi-bacillary leprosy
Nerve conduction study	Findings: CMAP and SNAP amplitudes of right Ulnar nerves are absent. CMAP amplitudes of right Tibial and Peroneal nerves are absent. SNAP amplitudes of right Sural and Superficial Peroneal nerves are absent. NCS findings of other nerves are normal.Impression: These electrophysiological findings are consistent with Sensorimotor Axonal Polyneuropathy affecting some of the studied nerves (Mononeuritis Multiplex). .
Sural nerve biopsy	Histopathological examination showed thick nerve bundle infiltration of chronic inflammatory cell with focal granuloma like area and multinucleated cells and few scattered mast call Special stain (Fite – Faraco)-revealed no acid-fast bacilli.

Discussion:

Because skin lesions, which are the hallmarks of leprosy, are never present, the clinical identification of pure neural leprosy (PNL) continues to be a challenge for public health care.[9]

Furthermore, even in cases when a nerve biopsy is feasible, identifying the leprosy bacillus remains a challenge. This study used many criteria to try to determine a valid PNL diagnosis in patients referred to our Leprosy Outpatient Clinic. Polymerase chain reaction (PCR) was used to identify M. leprae from the nerve samples taken from the 67 patients whose clinical, neurological, and electrophysiological test results supported peripheral neuropathy. [10]

Mononeuritis multiplex was the most common clinical and electrophysiological pattern of nerve damage, with sensory (89% of all cases) outnumbering motor (81%), and mainly axonal neuropathy, which was consistent with the current study.[11] Axonal neuropathy was the most common electrophysiological result, whereas histological nerve examination revealed epithelioid granuloma in 14% of patients, acid-fast bacilli in 16%, and nonspecific inflammatory infiltration and/or fibrosis in 39%. [12] In 46 instances of PNL, peripheral nerves were thickened in 32.6%, followed by trophic ulcers in 21.7%, muscle wasting, claw hand, and foot drop in 15.2%, 10.8%, and 8.7%, respectively, which was consistent with our case study.[13]

The patient in this case had symptoms compatible with PNL, such as nerve thickening, recurring painless ulcers, and mononeuritis multiplex, but lacked the usual anesthetic skin lesions associated with leprosy.

This case also demonstrates the diagnostic enigma associated with PNL, stressing the need to raise clinical suspicion and consider leprosy in patients presenting with unexplained neuropathy, especially in endemic areas. The patient's nerve conduction study results were compatible with sensorimotor axonal polyneuropathy, which is a hallmark of mononeuritis multiplex in PNL.

The diagnosis was confirmed by the abundance of AFBs in Schwann cells with macrophage and plasma cell infiltrates seen in the sural nerve biopsy, which was compatible with PNL. Because skin lesions, which are the hallmarks of leprosy, are never present, the clinical identification of PNL continues to be a challenge for public health care. [11] Multibacillary (MB) patients correspond to those who have two or more damaged nerves. Rifampicin, clofazimine, and dapsone are used in multidrug therapy, which continues for 12 months[14, 15]. In our PNL patients, we started treating them with multidrug therapy (MDT) regimens that are recommended by the WHO guidelines.

Conclusion:

Pure neural leprosy, however uncommon, should be examined in individuals with neuropathy and nerve thickening even when skin lesions are absent. Early diagnosis and implementation of WHO-recommended multidrug treatment (MDT) are critical for preventing additional nerve damage and impairment.

Consent:

Informed consent was obtained from the patient for the publication of this case report.

Declaration:

The authors declare no conflict of interest.

Acknowledgments:

The authors were grateful to the staffs of the Department of Neurology in Sir Salimullah Medical College Mitford Hospital, Bangladesh.

References:

1. Chimenos-Küstner E, Pascual-Cruz M, Piñol-Dansis C,Viñals-Iglesias H, Rodríguez de Rivera-Campillo ME, López-López J. Lepromatous leprosy: A review and case report. Med Oral Patol Oral Cir Bucal 2006;11:E474-9.
2. Jariyakulwong N, Julanon N, Saengboonmee C. Lepromatous leprosy with a suspected 30-year incubation period: A case report of a practically eradicated area. J Taibah Univ Med Sci. 2022 Feb 1;17(4):602-605. doi: 10.1016/j.jtumed.2021.12.005. PMID: 35983441; PMCID: PMC9356350. https://doi.org/10.1016/j.jtumed.2021.12.005
3. Rao PN. Global leprosy strategy 2016-2020: Issues and concerns. Indian J Dermatol Venereol Leprol. (2017) 83:4-6. doi: 10.4103/0378-6323.195075. https://doi.org/10.4103/0378-6323.195075
4. Scollard, D. M., Adams, L. B., Gillis, T. P., Krahenbuhl, J. L., Truman, R. W., & Williams, D. L. (2006). The continuing challenges of leprosy. Clinical Microbiology Reviews, 19(2), 338-381. https://doi.org/10.1128/CMR.19.2.338-381.2006
5. Rao PN, Suneetha S. Pure neuritic leprosy: Current status and relevance. Indian J Dermatol Venereol Leprol. 2016;82:252-61. https://doi.org/10.4103/0378-6323.179086
6. Mahajan PM, Jogaikar DG, Mehta JM. A study of pure neuritic leprosy: clinical experience. Indian J Lepr 1996;68:137-41. 7.
7. Palit A, Kar HK. Prevention of transmission of leprosy: the current scenario. Indian J Dermatol Venereol Leprol. (2020) 86:115-23. doi: 10.4103/ijdvl.IJDVL_326_19. https://doi.org/10.4103/ijdvl.IJDVL_326_19
8. Ooi W, Saini S. Case Report: Diagnostic Pitfalls in an Atypical Case of Primary Neuritic Leprosy. Am J Trop Med Hyg. 2021 Jun 21;105(2):490-493. doi: 10.4269/ajtmh.20-1130. PMID: 34152999; PMCID: PMC8437164. https://doi.org/10.4269/ajtmh.20-1130.
9. Garbino JA, Marques W Jr, Barreto JA, Heise CO, Rodrigues MM, Antunes SL, et al. Primary neural leprosy: systematic review. Arq Neuropsiquiatr. 2013;71(6):397-404. https://doi.org/10.1590/0004-282X20130046. https://doi.org/10.1590/0004-282X20130046
10. Pitta IJR, Hacker MAV, Andrade LR, Spitz CN, Vital RT, Sales AM, Antunes SLG, Sarno EN, Jardim MR. Follow-up assessment of patients with Pure Neural Leprosy in a reference center in Rio de Janeiro-Brazil. PLoS Negl Trop Dis. 2022 Jan 11;16(1):e0010070. doi: 10.1371/journal.pntd.0010070. PMID: 35015773; PMCID: PMC8752023. https://doi.org/10.1371/journal.pntd.0010070
11. Jardim MR, Antunes SL, Santos AR, Nascimento OJ, Nery JA, Sales AM, Illarramendi X, Duppre N, Chimelli L, Sampaio EP, Sarno EP. Criteria for diagnosis of pure neural leprosy. J Neurol. 2003 Jul;250(7):806-9. doi: 10.1007/s00415-003-1081-5. PMID: 12883921. https://doi.org/10.1007/s00415-003-1081-5
12. Jaiswal N, Chakraborti S, Nayak K, Pai S, Shelley BP, Sreeram S, Rakshith KC, Suresh BV, Misri ZK, Alva H, Shankar N. Hansen's Neuritis Revisited - A Clinicopathological Study. J Neurosci Rural Pract. 2018 Jan-Mar;9(1):42-55. doi: 10.4103/jnrp.jnrp_438_17. PMID: 29456344; PMCID: PMC5812158. https://doi.org/10.4103/jnrp.jnrp_438_17
13. Hui M, Uppin MS, Challa S, Meena AK, Kaul S. Pure neuritic leprosy: Resolving diagnostic issues in acid fast bacilli (AFB)-negative nerve biopsies: A single centre experience from South India. Ann Indian Acad Neurol. 2015;18:292-7. https://doi.org/10.4103/0972-2327.162284
14. Cruz RCDS, Bührer-Sékula S, Penna MLF, Penna GO, Talhari S. Leprosy: current situation, clinical and laboratory aspects, treatment history and perspective of the uniform multidrug therapy for all patients. An Bras Dermatol. 2017;92(6):761-73. https://doi.org/10.1590/abd1806-4841.20176724. https://doi.org/10.1590/abd1806-4841.20176724