Indian J Lepr 2022, 94 : 343-348

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Case Report

Sensory Ganglionopathy in Hansen's Disease: Report of a Patient and Review of Literature

SK Chaudhary[1], J Kalita[1], UK Misra[2]

Received : 09.02.2022 Accepted : 30.08.2022

Hansen's neuropathy usually affects cooler parts of the body, but presentation with poly-ganglionopathy is rare. We report a patient with ganglionopathy associated with pure neuritic Hansen's disease and discuss it in the light of the reported literature. A 42-year-old male presented with painful distal paraesthesia and ataxia for three months. His sensory nerve conduction was unrecordable. Thyroid hormones, vasculitis profile, HIV serology and hepatitis workup were normal. MRI of nerves and whole-body positron emission tomography were also normal. Slit smear examination of the affected skin after appropriate staining and microscopic examination showed the presence of AFB (*M. leprae*). He was treated with rifampicin, clofazimine, dapsone and prednisolone, and at six-month follow-up, he was asymptomatic. We conclude that leprosy should be considered in the differential diagnosis of sensory poly-ganglionopathy, especially in tropical countries, as it is amenably treatable.

Keywords : MRI, Leprosy, Neuropathy, Ganglionopathy, Pseudoathetosis

Introduction

The classical triad of Hansen's disease (HD) is maculo-anaesthetic patch, thickened nerve and positive acid-fast bacilli in slit smear or biopsy of skin or nerve (Eichelmann et al 2013). Leprosy neuropathy involves distal nerve endings and later involves dermal cutaneous sensory and mixed nerves by centripetal spread. *M leprae* has affinity for cool 28°C-32°C areas of sensory nerve and skin, giving it a typical clinical picture with diagnostic significance. It affects small and unmyelinated nerve fibres (Aδ and C) (Pandya & Bhatki 1994). In HD, joint position sensation and sense of vibration are generally spared (Wolf et al 1985). Large fiber sensory neuropathy and polyganglionopathy manifesting with ataxia and hyperalgesia are rare in HD (Pandya & Bhatki 1994, Rice et al 2016) and often attributed to malignancy, autoimmune disorders and drug toxicity (pyridoxine, cisplatin and paclitaxel). We could get only a fewl case reports on ganglionopathy with HD (Rice et al 2016, Bafna et al 2020, Khadilkar et al 2007) highlighting the rarity of this condition. In this communication, we report this patient, and discuss in the light of the available literature (Table 1).

[1] Dr Sarvesh K Chaudhary*, DM
[1] Dr Jayantee Kalita*, DM
[2] Dr Usha K Misra**, DM
* Department of Neurology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Raebareli Road, Lucknow, Uttar Pradesh - 226014.
** Department of Neurology, Vivekananda Polyclinic & Institute of Medical Sciences, UttarPradesh – 226007 Apollo medics superspecialty Hospital Kanpur Road, Lucknow.
Corresponding Author : Dr Usha K Misra, **Email :** drukmisra@rediffmail.com

Case Report

A 42 years old chartered accountant presented with walking instability for three months. He noted marked paresthesia in legs and hands and felt as if walking on bubbles and had a sensation of a film on his hand and fingers. He had difficulty gripping objects, signing and using the computer keyboard. He was non-diabetic, nonalcoholic and did not take any recreational drugs. He had tubercular lymphadenopathy in childhood and received nine months of antitubercular treatment. There was no history of any skin lesions or rash. His height was 175 cm, weight 80 kg, pulse 80/min, and blood pressure 120/70 mmHg without the postural drop. He walked with a stamping gait. He has no cutaneous hypopigmented or erythematous patches, skin atrophy, ichthyosis, xerosis, hair loss or autonomic changes. Peripheral nerves were not thickened. Muscle power was 5/5 in both lower limbs, shoulder, elbow and wrist, but had weakness of small muscles of both hands with agripstrength of 70%. There was generalized areflexia. Sensations of pinprick and touch were reduced by 20% in hands and feet (Figure 1A).

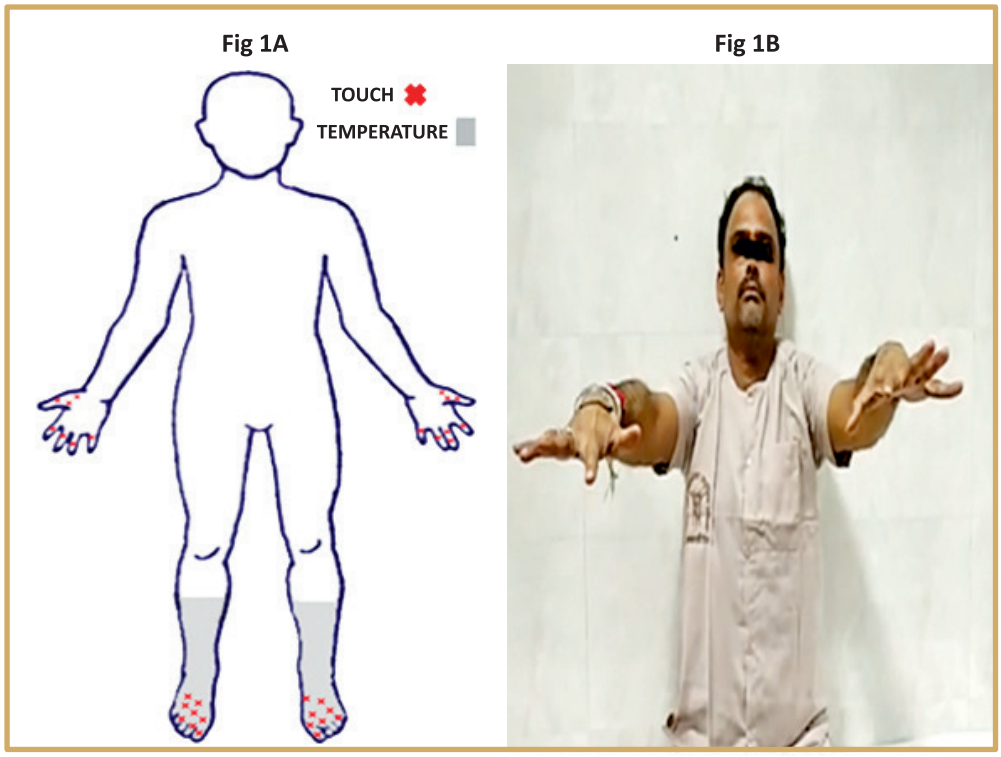

Figure 1A & 1B : Sensation of pinprick and touch were reduced by 20% over fingers and toes (Figure 1A) and temperature below knee by 50% position and vibration sensations were impaired in toes and fingers. Romberg's sign was positive. He had pseudo-athetotic movements in hands worsened by eye closure (Figure 1B).

Table 1 : Reports of Polyganglionopathy in Hansen's Disease

Author	No: of cases	Leprosy type	Lepra reaction	Clinical features	Confirmation of ganglionopathy	Treat-ment
			Ganglionopathy			
Khadilkar et al 2007	1	BT, left radial neuropathy	No	Sensory loss & weakness in radial distribution	MRI enhancement of left dorsal root ganglion at C5-6 (left)	MDT
Rice et al 2016	1	BT	No	Ataxia Weakness of left upper limb	MRI revealing C5-C7 and ganglionitis and myelitis	MDT+ steroid
Bafna et al 2020	1	Multi-bacillary	No	progressive weakness, tingling and numbness of all four limb	MRI cervical dorsal root ganglia enhancement and T2 hyper intensity at c6	MDT+ steroid
			Proprioception loss? Ganglionopathy			
Pandya & Bhatki 1993	7	BT AND BL	no	Ataxia, weakness and anesthetic patch	Lumbar biopsy in one showing lymphocytic infiltra-tion and no bacilli	MDT in 2 patients
Misra et al 2003	1	BT	yes	Ataxia and pseudoathetosis	Neurophysiology, no ganglion biopsy/MRI	MDT+ steroid
Van-Brakel et al 2005	7	All Multi-bacillary	No	Proprioception loss, weakness and areflexia	Neurophysiology	All on MDT
Khadilkar et al 2008	19	2pure neuritic, 4LL, 9BL, 3TT, 1BT	3	Proprioception loss, weakness (16) and areflexia (12)	Neurophysiology	
			Autopsy and biopsy study			
Liu Tze-Chun & QiuJu 1984	21 Spinal ganglia (14 ab-normal in LL, 2 in TT)	19 LL, 2 TT	-	All type of leprosy cases autopsy, lymph node biopsy	Neuron loss, foamy neuron, round cell infiltration and *M. leprae* in few	-

Supplementary Table 1 : Nerve conduction study of patient with ganglionopathy associated with Hansen's disease

Nerve	Distal latency (ms)	Amplitude (mv)	Conduction block	Nerve velocity m/sec	F-wave
Right median motor	4.3	4.6		42.1	32
Right ulnar motor	2.8	6.8		45.3	30.7
Right common peroneal	5.1	0.3		29.5	61
Right posterior tibial	5.6	0.4		36.3	
Left median motor	3.2	5.3		40	30.9
Left ulnar motor	2.7	3.8		44.4	31
Left common peroneal	5.3	0.2		30.7	57
Left posterior tibial	5.6	0.2		33.0	
Right median sensory	NR				
Right ulnar sensory	NR				
Right sural	NR				
Left median sensory	NR				
Left ulnar sensory	NR				
Left sural	NR				

NR = Not Recordable

Temperature sensation was ireduced by 50% below the knee, and joint position and vibration sensations were impaired in the toes and fingers. Romberg's sign was positive. He had pseudo-athetosis of hands-on eye closure (Figure 1B).

His hemoglobin was 14 gm/dl, total leucocyte-counts 9800/mm^3, erythrocyte sedimentation rate 13mm at the first hour, platelets 192 000/mm^3, fasting blood sugar 97 mg/dl, post-prandial 127 mg/dl, serum creatinine 1.02 mg/dl, bilirubin 0.9//dl, alanine transaminase 10U/L and alkaline phosphatase 192 U/L. His thyroid stimulating hormone was 2.2 miu/l, and antinuclear antibody, ENA and HIV serology were negative. Screening for hepatitis B and C was negative. Slit and smear were taken from both ear lobes and eyebrows. Smear revealed *M leprae.* MR neurography and whole-body positron emission tomography were normal. Sensory nerve conductions of the median, ulnar and sural were unrecordable bilaterally. H-reflex was not recordable. Motor nerve conductions of median, ulnar, posterior tibial and peroneal nerves revealed mild to moderate slowing with reduced compound muscle action potentials (supplementary Table 1). Electromyography of tibialis anterior, vastus lateralis, first dorsal interosseous and biceps was normal. He received dapsone 100 mg, clofazimine 50 mg, folic acid 5 mg and prednisolone 20 mg daily, and rifampicin 600 mg and clofazimine 300 mg monthly. Prednisolone was tapered after six weeks. At six months, he was asymptomatic, but multidrug therapy was continued for 12 months.

Discussion

The polyganglionopathy in in the present patient was suggested by sensory ataxia, pseudoathetosis, areflexia, mild weakness of hands and unrecordable sensory nerve conduction. The association of polyganglionopathy and Hansen's disease was confirmed by *M leprae* on slit and smear examination. Hansen's neuropathy usually presents with hypo-pigmented anesthetic skin lesions and thickened nerves in tuberculoid leprosy. These patients characteristically have mononeuropathy, mononeuritis multiplex or overlap neuropathy. Among the large nerve trunk, most commonly affected are the ulnar, peroneal, median, posterior tibial, superficial radial, great auricular, and facial nerves (Dastur 1955). Sensory polyganglionopathy is a rare condition and is reported in paraneoplastic, autoimmune disorders, anti-cancer drugs, and infections, including HIV, HTLV, Epstein–Barr and Varicella-Zoster (Sghirlanzoni et al 2005, Rubin & Daube 1999, Ramos et al 1999, Shimazaki et al 2002).

Sensory polyganglionopathy in HD has been reported in three patients whose ages ranged between 20 and 68 years, one had multibacillary, and two paucibacillary, and none had it during lepra reaction. MRI was d.one in three and revealed root and ganglion hyperintensity on T2 (Rice et al 2016, Bafna et al 2020, Khadilkar et al 2007). Four studies on 34 patients have reported the loss of proprioception in 34, ataxia in 7 and pseudoathetosis in 3; of them, 21 had multibacillary and 13 had paucibacillary; and four of them had lepra reaction (Pandya & Bhatki 1994, Misra et al 2003, Khadilkar et al 2008, Lockwood et al 2012). In a large cohort of 303 patients with leprosy, seven patients had impaired proprioception, and tendon reflexes were abnormal, which were unrelated to lepra reaction (van Brakel et al 2005).

An autopsy study, however, reported sensory ganglionoapthy in 16 out of 21 patients. There was round cell infiltration, nuclear pyknosis, cytoplasm swelling, and neuronal loss. In a few patients, *M leprae* was also detected. The authors suggested that the lesions in peripheral nerves may spread through the blood stream, lymphatic channels or by direct extension along the nerves (Liu & Qiu 1984). The studies reporting sensory polyganglionopathy and propioception loss are summarized in Table 1. The above-mentioned reports on clinical, MRI and histopathological examination suggest that sensory polyganglionopathy in leprosy is rare and may be due to retrograde degeneration, immunological reaction as it occurred in lepra reaction or direct invasion of dorsal root ganglia by the bacilli.

Sensory polyganglionopathy is rare in Hansen's Disease but should be kept on the list of differential diagnoses, especially in leprosy prevalent areas, because it needs specific treatment.

Acknowledgement

We thank Mr Shakti Kumar for his secretarial help.

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How to cite this article : Chaudhary SK, Kalita J, Misra UK (2022). Sensory Ganglionopathy in Hansen's Disease: Report of a Patient and Review of Literature. *Indian J Lepr.* **94**: 343-348.