Report

Notalgia paresthetica: a study on pathogenesis

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Abstract

Background Notalgia paresthetica is a sensory neuropathy involving the dorsal spinal nerves. The characteristic symptom is pruritus on the back, occasionally accompanied by pain, paresthesia, and/or hyperesthesia, which results in a well-circumscribed hyperpigmented patch in the symptomatic area. The etiology of this condition has not yet been completely defined.

Objective Possible mechanisms that could explain the pathogenesis of notalgia paresthetica were investigated through clinical examination and various diagnostic tests.

Methods Ten cases of notalgia paresthetica underwent dermatologic, neurologic, and orthopedic examination. This was followed by skin biopsy, electrodiagnostic investigation, and radiography of the spine.

Results All patients had a typical symptomatology and dermatologic picture. Neurologic examination and standard electrodiagnostic investigation results were normal in all cases. Histopathology was compatible with postinflammatory hyperpigmentation; there were no amyloid deposits. In seven cases, degenerative changes in the vertebrae were observed and, in all of these cases, these changes were most prominent in the vertebrae which corresponded to the dermatome of the cutaneous lesion.

Conclusions The striking correlation of notalgia paresthetica localization with degenerative changes in the spine suggests that spinal nerve impingement may contribute to the pathogenesis of this entity.

Introduction

Notalgia paresthetica (NP) was first defined by Astwazaturow in 1934 as a sensory neuropathy involving the dorsal spinal nerves.[1] The characteristic symptom is pruritus on the back, occasionally accompanied by pain, paresthesia, and/or hyperesthesia.[1–4] The dermatologic finding, which consists of a well-circumscribed hyperpigmented patch, is thought to be secondary to the chronic rubbing and scratching due to pruritus.[5–7]

Although the condition is not believed to be rare, NP is not frequently reported and investigational studies on its pathogenesis are few.[5,8,9] Thus, the exact pathomechanism of the condition has yet to be defined. In this study, we aimed to increase our understanding of the pathogenesis of this curious neuropathy by examining various clinical and laboratory findings in 10 cases.

Patients and methods

Ten NP patients, seven women and three men, were included in the study. The mean age was 44.50 ± 16.78 years (range, 21–74 years). The duration of NP was 9.35 ± 10.12 years (range, 1–30 years). All cases were questioned for various neurologic/dermatologic symptoms and the presence of similar complaints in family members. Dermatologic examination of each case included a recording of the dermatomal distribution of the lesion. A punch biopsy of lesional skin was followed by electromyography (EMG). Skin biopsy specimens stained with hematoxylin and eosin and Congo red were evaluated under light microscopy by an observer blind to the clinical diagnoses. The device used for electrodiagnostic tests was a Nihon-Kohden Neuropack II (Tokyo, Japan). A chart of electrodiagnostic algorithm, developed in accordance with the literature in our neurophysiology laboratory and routinely employed to investigate any neurologic pathologies of the back, was used.[10–12] This electrodiagnostic study protocol is shown in Table 1. Regardless of the localization of NP, all levels of the spine were thus examined. Electrodiagnostic findings in each case were recorded separately and were compared with the normal values of our clinical neurophysiology laboratory. Values exceeding the normal mean by two standard deviations (SD) were accepted as abnormal. The neurologist conducting the electrodiagnostic tests

Table 1 Electrodiagnostic study protocol

Nerve	Stimulation	Recording (and needle EMG studies)	Distance
Median S	Wrist	2. Finger	110 mm
Ulnar S	Wrist	5. Finger	110 mm
Median M	Wrist	Abductor pollicis brevis	70 mm
	Antecubital fossa		
Median F wave	Wrist	Abductor pollicis brevis	
Ulnar M	Wrist	Abductor digiti minimi	
	Sulcus condylaris		
Ulnar F wave	Wrist	Abductor digiti minimi	
Dorsal scapular	C4 level	Levator scapulae	
Dorsal scapular	C4 level	Rhomboideus major	
Thoracicus longus	C6 level	Serratus anterior	
Accessorius	C4 level	Trapezius inferior	
Musculocutaneous	C6 level	Biceps brachii	
Radial M	C7 level	Triceps	
Deltoid	C6 level	Deltoid	
Suprascapular	C6 level	Infraspinatus, supraspinatus	
Thoracodorsal	C7 level	Latissimus dorsi	
Subscapular inferior	C6 level	Teres major	
	C4–T12	Paraspinalis	

EMG, electromyography; M, motor; S, sensory.

was not blind to the diagnoses of the patients due to the nature of testing which enabled him to inspect each patient's back. Work-up was completed by physical examination and radiographic evaluation (X-ray and magnetic resonance (MR) for all patients) of the spine by an investigator who was blind to the NP localization.

Results

Clinical findings

In all patients, a characteristic hyperpigmented patch on the back was observed (Figs 1 and 2). There was mild lichenification in one case. In two cases, there were bilateral patches. One patient had a similar pruritic lesion in the right hypochondrium which had appeared following a cholecystectomy operation. Pruritus was the common symptom. Two patients also complained of episodic pain and one patient described a tingling–burning sensation. One patient had mild regression of his pruritus following skin biopsy. Family history was negative in all cases (Table 2).

Histology

The epidermis appeared normal in nine cases and showed mild hyperkeratosis in one patient (Patient 1). There were no necrotic keratinocytes. In the papillary dermis, a very mild inflammatory infiltrate, including a moderate number of diffusely spread melanophages, was observed. No amyloid deposits were seen in any of the preparations. No gross alterations in cutaneous nerves were observed with the stains employed.

Neurologic examination and EMG

There was no positive history for any cause of peripheral neuropathy (toxic, vascular, familial, diabetic, drug induced) in any of the patients. Detailed neurologic examination did not reveal any sensory or motor loss. Results of electrophysiologic examination were within the normal range in all cases.

Evaluation of the spine

Orthopedic examination of the spine was normal in all cases. Radiographic evaluation of the vertebral column showed various pathologies in seven patients (Figs 3 and 4). For each of these seven patients, the segmental location at which the severest of these changes was observed was in accordance with the NP dermatome (Table 3). There was no history of spine trauma that would account for the radiographic findings in any of the patients.

Discussion

Although generally accepted to be a sensory neuropathy, the pathogenesis of NP has not been completely elucidated.

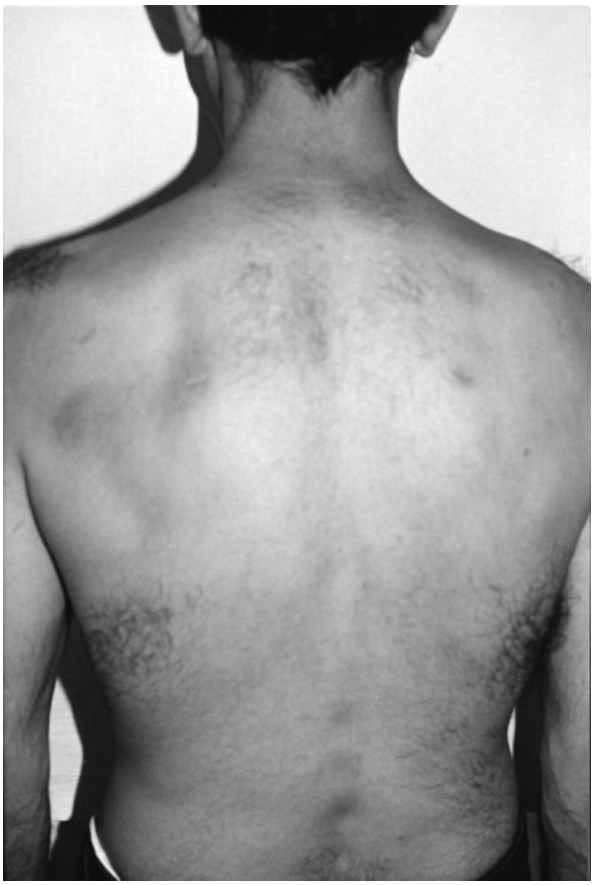


Figure 1 Patient 8 with the characteristic hyperpigmented patch on the back

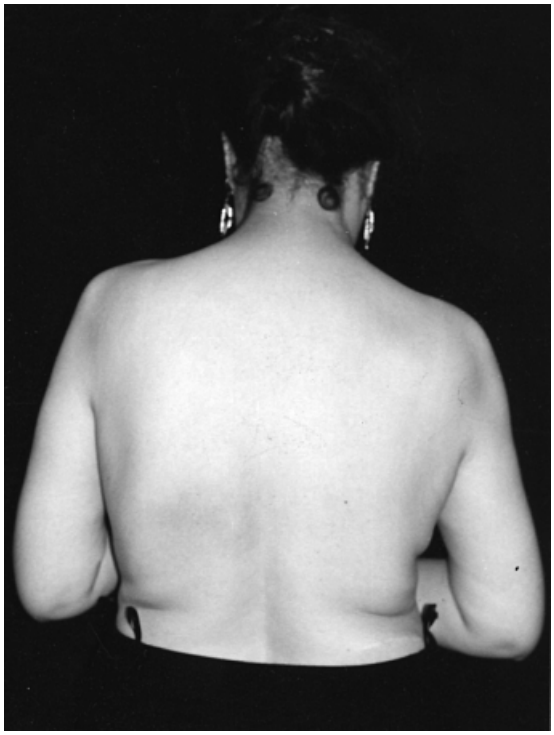


Figure 2 Patient 9

Factors implicated include a hereditary component, increased dermal innervation, viscerocutaneous reflex mechanism, neurotoxicity of certain chemicals, and spinal nerve injury due to trauma or entrapment.[5,13–17] Unfortunately, the above-mentioned factors have been the subject of individual reports and have been investigated separately. No study has yet evaluated these various parameters comparatively in the same group of patients. With this preliminary study we intended to determine how relevant all of the suggested factors were to this entity.

Symptomatology consisted of chronic pruritus in all cases, accompanied by episodic pain in two patients and a tingling–burning sensation in one. This initial finding was suggestive of a disorder of neurogenic origin. No hereditary component or toxic, metabolic, or medicamentous cause which would result in neuropathy was discovered. Reports dealing with the role of heredity and chemicals are few and anecdotal.[14–16] Although the evaluation of 10 cases is not sufficient to exclude these factors completely, we believe that they do not play a significant role in the pathogenesis of NP.

The histopathologic findings, consisting of pigmentary incontinence and a mild inflammatory infiltrate in the papillary dermis, are in accordance with postinflammatory hyperpigmentation which is a result of the chronic rubbing and scratching. The absence of amyloid deposits in all cases, even in patients with a 30-year history of NP, lead us to conclude that, as Westermark *et al.*[18] have previously reported, NP and macular amyloidosis are two different entities.

The presence of vertebral column pathologies in seven of the 10 patients, in accordance with the NP dermatome, is the most striking finding in our series. The various degenerative spinal changes could possibly contribute to the pathogenesis of NP by causing spinal nerve compression. It has been suggested that, because the spinal nerves emerge through the multifidus spinae muscle at right angles, they are prone to injury by trauma or entrapment.[14] Eisenberg *et al.*[13] reported a single case with NP in whom nerve root impingement precisely correlated with the clinical findings. A very recent review of 12 NP patients by Raison-Peyron *et al.*[19] has revealed dorsal spinal X-ray changes in all of the nine cases who underwent radiographic evaluation. In all seven of our patients with spinal pathology, the radiographically observed degenerative changes were severe enough to cause spinal nerve compression. It may be argued that, as degenerative

Table 2 Clinical characteristics of the cases

Patient	Age	Sex	Family history	NP duration (years)	Symptomatology			Other clinical findings
					Pruritus	Pain	Paresthesia	
1	68	M	–	30	+	+		Lichenification Mild regression following biopsy
2	74	F	–	10	+			Similar patch at site of cholecystectomy
3	31	F	–	2	+		+	
4	29	F	–	2	+			
5	21	M	–	1.5	+			
6	47	F	–	10	+			
7	46	F	–	1	+	+		
8	38	M	–	10	+			
9	52	F	–	24	+			
10	39	F	–	3	+			

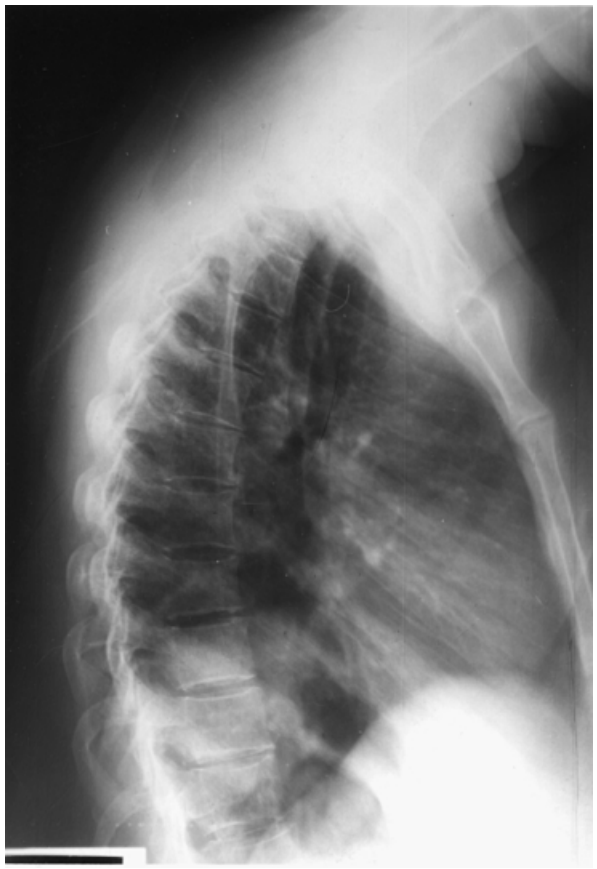


Figure 3 X-Ray of Patient 8 showing degenerative changes in the thoracic segments, most prominent at T1–5

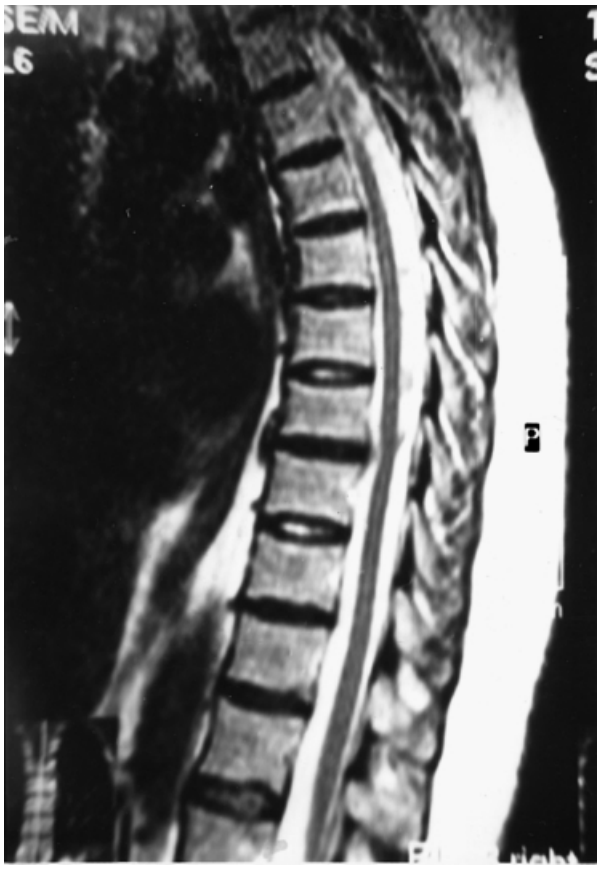
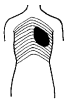
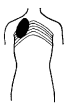


Figure 4 Magnetic resonance of Patient 9 showing disk protrusion at T6

changes of the spine are quite common in the elderly, such radiographic findings in older NP patients are incidental; however, it must be emphasized that the interesting finding in our study was not just the presence of spinal degenerative changes in NP patients, but rather the striking match between NP localization and the level at which spinal changes were most prominent. We also believe that radiographic evaluation of the spine by an investigator who

Table 3 Relationship of NP localization to spinal changes

Patient	NP localization (A)	Radiographic findings (B)	Graphic representation of dermatomes (A + B)	Relationship
1	Bilateral C6–8	Degenerative changes in the cervicothoracic segments,		Yes
2	Right T4–8 Right hypochondrium T8–10	Extensive degenerative changes in the thoracic segments, minimal thoracic scoliosis		Yes
3	Left C6–8	Normal		No
4	Right T3–7	Normal		No
5	Right T10–12	Normal		No
6	Bilateral T2–4 Right T6	Degenerative changes in the thoracic segments most prominent at T1–6		Yes
7	Right C5–7	Herniated nucleus pulposus at C5		Yes
8	Left C7–T4	Degenerative changes in the thoracic segments most prominent at T1–5		Yes
9	Left T6–7	Degenerative changes in the thoracic segments, protrusion of nucleus pulposus at T6		Yes
10	Left C4–5	Cervicothoracic degenerative changes and minimal protrusion of nucleus pulposus at C5		Yes

was blind to the NP localization also contributes to the conclusion that our findings are not just chance results.

No radiographic findings were observed in the three youngest patients and spinal examination was also normal; however, Patient 5 had a 2-year history of intermittent mild upper back pain and Patient 3 had been treated for cervical pain. Thus, we tentatively speculate that spinal pathologies which cannot easily be diagnosed radiographically, such as cervical fibrous bands or muscle spasms due to repetitive microtrauma, in sports for example, could also contribute to NP.

Electrodiagnostic results of all cases were within normal limits. In the case of electrodiagnostic studies, the search for a suspected radiculopathy could prove to be a challenge. As stated by Wilbourn and Aminoff,[20] ''electrodiagnostic studies are not as sensitive for detecting many other entities, such as carpal tunnel syndrome. The symptomatology and radiological findings may not present as electrodiagnostic reflections. So, a normal neurophysiological work-up does not necessarily rule out neurological disease.'' Electrodiagnostic findings in NP patients vary. Massey and Pleet[8] have reported positive waves in seven out of nine patients as evidence of paraspinal denervation. Marcusson *et al.*[7] have found normal EMG results in both patients they reported. A single case presented by Streib and Sun[21] showed needle EMG findings compatible with spinal neuropathy, whereas another case published by Eisenberg *et al.*[13] showed normal EMG.

Although well aware that an evaluation of 10 NP cases is not sufficient to explain the entire pathogenesis of this disorder, we conclude that spinal changes are worth investigating as a contributing factor. Keeping in mind that the largest number of NP cases reported in one study is 14, we believe that our results will contribute to a better understanding of this entity. Future studies with sex- and age-matched controls will surely yield more definite opinions as to how relevant the changes in the spine are to the changes in the skin. We suggest that all dermatologists and neurologists who come across a case of NP include physical and radiographic examination of the spine in their work-up. We would also like to remind orthopedic surgeons and neurosurgeons who have patients with a puzzling hyperpigmented patch on their back, in addition to spinal complaints, that the two very different symptoms could be the result of a single pathology.

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