Acta Dermatovener (Stockholm) 60: 119–122, 1980

VIBRATORY PERCEPTION IN PATIENTS WITH GENERALIZED SCLERODERMA

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Abstract. The vibratory perception threshold (VPT) was measured on eight different test-spots in 13 patients suffering from generalized scleroderma of the acrosclerosis type without clinical evidence of peripheral neuropathy. VPT was significantly elevated in skin areas severely involved in the sclerodermic process, whereas normal on test-spots with less affected skin. The pattern of impaired vibratory perception differs from that usually seen in peripheral neuropathy and it is concluded that the pattern most probably reflects the viscous-elastic properties of the test-spots, indicating that cutaneous sclerosis exerts a damping effect on external vibrations. It is suggested that this phenomenon could be utilized as an objective gauge for the severity of the sclerosis.

Key words: Vibration sense; Scleroderma; Peripheral neuropathy

There have been conflicting reports on a possible affection of the peripheral nervous system in patients with generalized scleroderma. Polyneuropathy has been described in a few well-documented case reports (6, 11), while Gordon & Silverstein (3) were unable to demonstrate any clinical manifestations of neuropathy in 130 patients. Electromyographically, several studies have shown changes compatible with a myopathy in patients with generalized scleroderma (5, 9, 15, 16), whereas Christopher & Robinson (1) claimed that the motor and sensory conduction velocity was significantly retarded in their patients—a change suggestive of neuropathy. However, to our knowledge the latter observation has not been supported by other studies and it should, moreover, be considered with some reservation, since the patient material and control group did not match in age distribution. Richter (11) described severe histopathological epi- and perineural fibrosis with demyelination and axonal degeneration in his two patients with clinical neuropathy. This was recently confirmed by Fleischmajer et al. (2), who found cellular infiltrates in the perineurium of patients with generalized and localized scleroderma. In a study by Pawlowski (10), a fragmentation of nerve fibres of the skin was a frequent finding.

One of the earliest signs of peripheral neuropathy is an impairment of the vibratory perception, especially on the toes. This has been demonstrated for example in patients with diabetes mellitus (4, 14), uraemia (7), and in vitamin B_{12} insufficiency (13). However, the fact that the actual amplitude of vibrations is markedly influenced by the viscous-elastic properties of the skin and subcutaneous tissue (8) is of particular importance in generalized scleroderma, in which dermal thickening may be expected to exert a damping effect on the vibration amplitude. Hence, elevated vibratory perception thresholds may erroneously be interpreted as indicating peripheral neuropathy.

To elucidate this problem, we measured the vibratory perception threshold (VPT) on eight different test-spots in patients with generalized scleroderma of the acrosclerosis type without clinical evidence of peripheral neuropathy. The VPT was significantly elevated on the most sclerotic test-spots, while remaining normal on test-spots with apparently normal skin. The pattern of elevated VPT differed considerably from that characteristic of peripheral neuropathy.

MATERIAL AND METHODS

Thirteen consecutive patients, 10 females and 3 males, aged 18–69 (mean = 53) years were studied. None of them exhibited symptoms or signs of peripheral neuropathy, nor did they suffer from disorders where subclinical neuropathy might be suspected. All patients fulfilled the criteria of systemis scleroderma of the acrosclerosis type, i.e. with predominance of skin affection on hands, feet, and face. The duration of the skin disease ranged from 3 to

Table I. *Clinical data in 13 patients with acrosclerotic systemic scleroderma*

No.	Sex/ age (y.)	Duration (y.)	Severity of skin changes	Involvement of	
				Oesophagus	Lung
1	M/59	8	Severe	+	+
2	F/18	5	Mild	+	−
3	F/69	17	Mild	+	−
4	F/47	20	Moderate	+	−
5	F/38	7	Severe	(+)	−
6	F/57	14	Severe	+	+
7	F/42	9	Severe	+	−
8	F/56	12	Moderate	−	−
9	F/56	4	Moderate	(+)	−
10	F/69	11	Mild	−	−
11	F/64	10	Moderate	+	+
12	M/56	3	Severe	−	(+)
13	M/55	3	Moderate	+	−

20 years. The cutaneous affection was mild in 3, moderate in 5 and severe in 5 patients. All of them complained of Raynaud's phenomenon. All patients were treated with glutamine and/or dimethylcystein. Clinical data are summarized in Table I.

Vibratory perception threshold (VPT). The vibration stimulus consisted of harmonic mechanical oscillations of 100 Hz delivered by an electromagnetically driven vibrator (Biothesiometer, Chagrin Falls, Ohio). The stimulus strength was expressed as the square of the voltage of the energy supply, according to the equation $A = k\ (V^2/m)$, where A = amplitude of vibrations, m = mass, and V = volts (8). The threshold for vibrations was measured as described by Nielsen (7) on the following test-spots: The pulp of digits 1–5, the radial styloid process, the pulp of the big toe, and the medial malleolus. The skin temperature was normal. VPT was not measured on pulps with ulcerations. The skin overlying the radial styloid process and the medial malleolus showed minimum changes in 3 patients, whereas the pulps of the digits were involved in all, and the big toe in most patients.

In normal subjects, the VPT increases with age. The normal regression equations of VPT ($\log V^2$) on age (years) for the selected test-spots are given in Table II, which also shows the 95% of normal variations ($= \pm 2\ S_{y.x}$). In the patients, each observation represents the mean of four measurements. To account for the normal age variation, data are expressed as the deviation from the normal mean value for the age of the patient ($\Delta VPT = \Delta \log V^2$). The significance of the average deviation of ΔVPT from zero was evaluated with the Student's t-test.

RESULTS

Although scleroderma usually affects the patient symmetrically, the severity of skin changes need not be the same on the right and the left extremities. Therefore, VPT values measured on right and left test-spots were analysed as independent observations.

The average age-adjusted deviation of VPT on the eight test-spots is shown in Table II, and representative plots are shown in Fig. 1. ΔVPT was significantly increased on the pulps of digits 1–5 ($P < 0.001$–< 0.01), most pronounced on digit 1, where half the observations were above the 95% limit of normal variation (Fig. 1, PP). In comparison, only 10% of single observations exceeded the upper limit of normal on the pulps of digits 4 and 5. ΔVPT on the pulp of the big toe showed greater individual variation, and the average deviation from normal only reached statistical significance at the 5% level, mainly due to the elevated values in 2 or 3 patients (Fig. 1, PH). Over the radial styloid process and the medial malleolus, the average ΔVPT did not differ significantly from zero, and the 95% range of normal variation was exceeded in only a few patients. Using the mean values of VPT of the same test-spot on the right and the left side revealed no change in the pattern of deviations, except that the average deviation of the VPT on the big toe was no longer significantly different from zero. There was no correlation between ΔVPT and the general classification of the scleroderma as mild, moderate, or severe.

DISCUSSION

To our knowledge, the vibratory perception threshold has not previously been measured in patients with generalized scleroderma. The present study shows that in these patients the mean VPT was significantly elevated on the pulps of the digits as compared with normal persons of the same age. A borderline impairment of the vibratory perception was recorded on the big toe, while VPT was normal over the radial styloid process and the medial malleolus. With few exceptions, however, most VPT values were only mildly increased.

We do not consider the impaired vibratory perception in sclerodermic patients to be an indication of a peripheral neuropathy, for the following reasons: 1) None of the patients in this material presented other symptoms or signs of peripheral neuropathy; 2) the distribution of impaired vibratory perception differed considerably from that in patients with peripheral neuropathy. The latter group is characterized by an increase of the VPT

Table II. *The regression of the vibratory perception threshold (VPT = y = log V²) on age (x = y.) in normal subjects, and the age-adjusted deviation (ΔVPT = Δlog V²) from the normal mean VPT in 13 patients with generalized scleroderma*

N = number of persons; S_{ux} = standard error of estimate; n = number of test-spots, right and left side; P = significance of difference from zero (Student's t-test). For individual observations, see Fig. 1

	Normal subjects ($y=a+bx$)				Sclerodermic patients (Δ VPT)			
Test-spot	N	a	b	S_{ux}	n	Mean±S.E.	t	P
Digit 1	100	0.8217	0.0121	0.176[a]	26	0.3640±0.0636	5.7261	<0.001
Digit 2	100	0.7631	0.0138	0.2020[a]	23	0.2754±0.0641	4.2968	<0.001
Digit 3	100	0.7539	0.0147	0.2059[a]	23	0.2443±0.0643	3.8004	<0.001
Digit 4	100	0.7943	0.0148	0.2224[a]	23	0.2093±0.0637	3.2882	<0.005
Digit 5	100	0.8551	0.0158	0.2305[a]	23	0.1587±0.05521	3.0439	<0.01
Radial styloid process	45	1.2161	0.0102	0.1679[b]	26	0.0668±0.0581	1.1498	n.s.
Pulp, big toe								
Males	44	1.0066	0.0249	0.2325[c]	25	0.2556±0.0931	2.7447	<0.05
Females	41	1.2277	0.0149	0.2089[c]				
Medial malleolus								
Males	44	1.4454	0.0177	0.2111[c]	25	0.1278±0.0789	1.6192	n.s.
Females	41	1.4472	0.0138	0.2224[c]				

[a] Roland & Nielsen (12).
[b] Nielsen (unpublished data).
[c] Nielsen (7).

distally in the legs, the pulp of the big toe being the site of the earliest and most severe affection, whereas VPT on the pulps of the digits is usually normal or only mildly elevated even in more advanced stages (7); 3) in peripheral neuropathy, VPT on the medial malleolus usually becomes impaired in parallel with the VPT on the big toe. In the present material, the VPT was normal over the malleolus as well as over the radial styloid process. This may partly mean that the skin overlying these test-spots was less affected by the scleroderma or not affected at all. However, another possibility is that external vibrations are easily transferred to the Pacinian corpuscles in the periosteum of the bone, irrespective of changes in the thin layer of skin and subcutaneous tissue interposed between the vibrator and the closely underlying bone. These receptors showed no evidence of neuropathy; 4) the significant impairment of the VPT on the pulps of the digits and, less pronounced, on the big toe was fully compatible with the distribution and the severity of skin changes in generalized scleroderma of the acrosclerosis type.

Thus, we suggest that the pattern of impaired vibratory perception reflects the viscous-elastic properties of the test-spots, indicating that sclerosis of the skin and subcutaneous tissue interposed between the stimulator and the receptors exerts a damping effect on external vibrations. In fact, this phenomenon could be utilized as an objective gauge for the severity of the acrosclerotic skin changes. By applying a fixed voltage to the vibrator, the

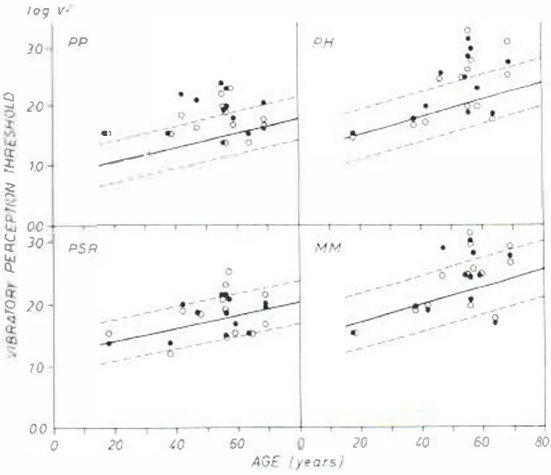

Fig. 1. Vibratory perception thresholds in patients with generalized scleroderma on four test-spots: *PP* = pulp, digit 1; *PSR* = radial styloid process; *PH* = pulp, big toe; *MM* = medial malleolus. ●, Right and ○, left extremities. —, Mean VPT and ---, 95% range of normal variation, adjusted for age. Regression equations are given in Table II.

actual peak-to-peak amplitude of vibrations can be measured in μm as described by Nielsen (8). With a constant pressure against the skin the amplitude will depend on the viscous-elastic properties of the test-spot. Increased stiffness will damp the amplitude of vibrations.

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Received July 19, 1979

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