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Clinical Appearance of Skin Lesions and Disturbances of Pigmentation in Localized Scleroderma

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Skin manifestations of localized scleroderma were assessed clinically in 58 patients presenting 214 circumscribed scleroderma lesions (23 patients with localized morphoea plaques, 15 with generalized morphoea, 11 with linear scleroderma of trunk and extremities, 9 with scleroderma en coup de sabre). Characteristic differences between different types of localized scleroderma with respect to age at debut, regional distribution, symmetry and linearity, degree of sclerosis of the lesions, and the presence of inflammatory change of colour, pigmentation and visible atrophy of underlying subcutaneous tissue were found. Separate brown-pigmented spots resembling atrophoderma Pasini-Pierini were observed in 49% of the patients. A regional distribution chart of linearity in localized acleroderma was elaborated. The frequent finding of disturbances of pigmentation, the character of the linear distribution-pattern, and affections of underlying anatomical structures is discussed to indicate a predisposing defect in the migration of crest cells during embryonal life. *Key words: Scleroderma; Circumscribed; Morphoea; Pigmentation; Predisposition.* (Received April 25, 1984.)

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The characteristic circumscribed skin lesions of localized scleroderma consist of white, thickened and indurated plaques, lines, bands or guttate lesions with increased content of collagen in the dermis. The clinical spectrum of manifestations of this rather rare disorder is wide. Localized scleroderma is generally assumed not to involve visceral organs (1).

In 1930, O'Leary & Nomland reported on clinical findings in a series of patients with localized scleroderma (2). During the fifties, Christianson et al., Curtis & Jansen, and Stava published extensive studies based on reviews of records covering several years (3, 4, 5). A study on 13 cases of linear scleroderma with illustrations of the regional distribution was published in 1948 by Rubin (6). In the present study, the clinical appearance of skin manifestations of localized scleroderma including disturbances of pigmentation is assessed in a prospective study.

MATERIAL AND METHOD

The patient material includes 58 patients with a clinical and microscopical diagnosis of localized scleroderma admitted to the Department of Dermatology, Rigshospital, during the period February 1980—January 1984 (Table I). All the patients had presented one or more typical lesions of scleroderma with white colour, induration, and clinically increased skin thickness (Fig. 1).

The follwing classification of localized scleroderma was used.

Morphoea:

1. *Localized morphoea plaque* (LMP), i.e. one or a few circumscribed plaques typically of medium size, located to a few regions.
2. *Generalized morphoea* (GM), i.e. several circumscribed plaques typically large and located to several regions.
3. *Guttate morphoea*, i.e. numerous drop-like (guttate) scleroderma lesions occurring in regions.

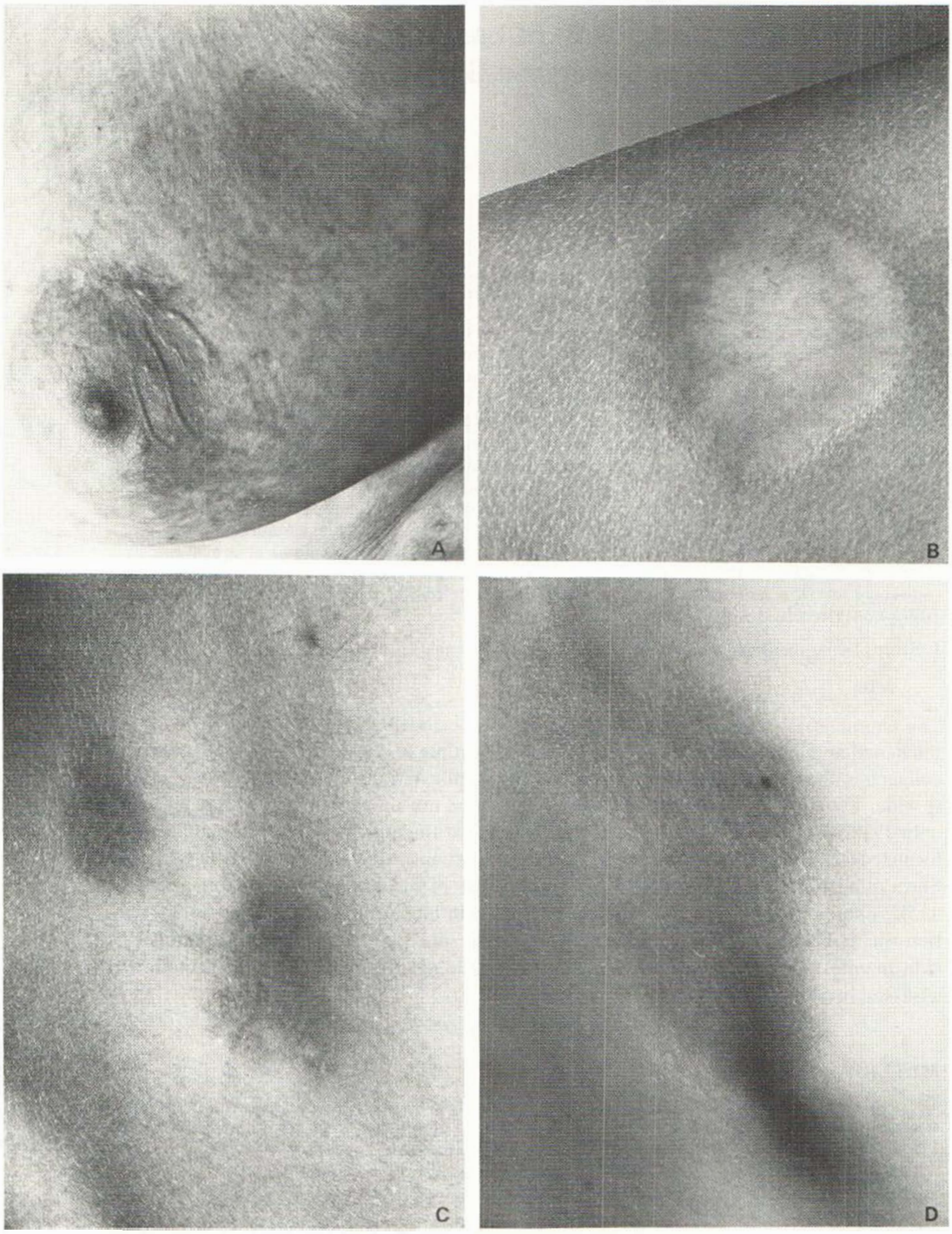

Fig. 1. (A) Very early plaque of morphoea exhibiting lesional redness. (B) Fully developed plaque or morphoea with white colour, thickening and induration (C) Pigmented spots of the right side of the lower abdomen in a patient with morphoea (LMP). (D) Pigmented spots at the right side of the back between the spine and the scapula. The spots are partly confluent. The superior one shows linearity.

Table I. *Sex, age (mean and range) and age at debut of localized scleroderma (mean and range) of 58 patients included in the study*

	No. of patients	Sex		Age (years)	Age at debut (years)
		Females	Males		
Morphoea					
Localized plaque	23	13	10	28.3 (8–70)	25.3 (6–69)
Generalized	15	13	2	58.9 (36–76)	54.3 (33–73)
Linear types					
Extremity and trunk	11	10	1	28.6 (6–57)	13.8 (6–32)
En coup de sabre	9	8	1	22.6 (8–51)	14.7 (4–29)

Linear types:
1. *Linear scleroderma of extremities and trunk (LET)*, i.e. one or a few linear or band-like lesions of scleroderma located to the extremities or to the trunk.
2. *Linear scleroderma "en coup de sabre" (LCS)*, i.e. one or a few lines of scleroderma located to the front or forehead.

The following notes were made: Number of scleroderma lesions in each patient, the regional distribution, the clinical degree of sclerosis classified as advanced or slight, symmetry and linearity, the clinical appearance of the lesion centre (faint redness, pigmentation, visible underlying atrophy of the subcutaneous tissue), the clinical appearance of the perilesional area (lilac ring, pigmentation, visible underlying atrophy of the subcutaneous tissue). The presence and location of *pigmented spots* (PS), i.e. circumscribed and distinct lesions of brown pigmentation were registered (Fig. 1). The degree of sclerosis was not assessed in linear scleroderma. Lesions were accepted as being symmetrical if they were located to a similar part of symmetrical body regions and if they were both either plaques or lines, but the lesions need not be exactly identical. Linearity was registered if only one border of a plaque or its periolesional pigmentation was linear. Assessment of colour and disturbances of pigmentation were performed in bright daylight. Atrophy of the subcutaneous tissue was only registered when clinically obvious. In the majority of patients with GM it is not possible to define PS since the perilesional pigmentation is often widespread. Perilesional changes were not registered in LET and LCS.

In some of the patients X-ray examination of the thoracolumbar spine was performed to detect congenital anomalies.

Table II. *Number (in each patient mean and range) and regional distribution of 214 scleroderma lesions (plaques and lines) registered in 58 patients with different types of localized scleroderma*

	No. of lesions		Regional distribution				
	total	In each patient	Trunk	Extremity	Posterior vertex	Front	Lower face
Morphoea							
Localized plaque	47	2.0 (1–8)	35	11	1	–	–
Generalized	123	8.2 (3–12)	98[a]	22	3	–	–
Linear types							
Extremity and trunk	22[b]	2.0 (1–6)	4	18	–	–	–
En coup de sabre	22[c]	2.4 (1–4)	2	–	–	17	3

[a] Includes 5 plaques located to crena ani, 4 to the midline, 2 to the umbilicus.
[b] Two plaques in two patients included, one located to the trunk, one to an extremity.
[c] 5 plaques in 4 patients included, 3 located to a cheek, two to the trunk.

Table III. *Symmetry and linearity of lesions of localized scleroderma registered in 58 patients presenting 214 lesions*

	Symmetry		Linearity	
	No. of patients	No. of lesions	No. of patients	No. of lesions
Morphoea				
Localized plaque	2	3	9	12
Generalized	15	44	14	55
Linear types				
Extremity and trunk	1	1[a]	–[c]	–[c]
En coup de sabre	2[b]	2[a]	–[c]	–[c]

[a] Plaques registered in different regions not included.
[b] 3 patients presented symmetrical lines of the front with pigmentation only.
[c] Exhibiting linearity per definition.

Table IV. *Degree of clinical sclerosis in patients with morphoea*

38 patients presenting 170 plaques were examined

	Degree of sclerosis (no. of plaques)		No. of patients with plaques of advanced sclerosis
	Advanced	Slight	
Morphoea			
Localized plaque	41	6	18
Generalized	8	115	4

Table V. *Clinical appearance of scleroderma lesions in 58 patients with different types of localized scleroderma presenting 214 scleroderma lesions in total*

	Faint redness		Pigmentation		Visible atrophy of subcutaneous tissue	
	No. of patients	No. of lesions	No. of patients	No. of lesions	No. of patients	No. of lesions
Morphoea						
Localized plaque	7	8[a]	16	22	3	3
Generalized	2	6[b]	13	81	1	3
Linear types[c]						
Extremity and trunk	–	–	10	17	7	14[d]
En coup de sabre	–	–	9	15	9	14[e, f]

[a] Two intense.
[b] Two intense.
[c] Only linear lesions included.
[d] 3 cases presented contractures of large joints and 3 shortening of involved extremity (range 1–3.5 cm).
[e] In 9 lesions underlying depression of frontal bone by palpation.
[f] Two cases had eye involvement at the same line, and one facial hemiatrophy of Romberg.

Table VI. *Clinical appearance of the perilesional area in 38 patients with morphoea presenting 170 plaques*

	Lilac ring		Pigmentation		Visible atrophy of subcutaneous tissue	
	No. of patients	No. of lesions	No. of patients	No. of lesions	No. of patients	No. of lesions
Morphoea						
Localized plaque	5	8	13	16	2	2
Generalized	5	16[a]	13	77	1	2

[a] 4 intense.

RESULTS

As it appears from Table I, GM has its debut in elderly people, while linear types of scleroderma appear in childhood or in adolescence. LMP may appear throughout life, however, with a preference for adolescence.

As it appears from Table II, LMP and GM mainly affect the trunk with a similar ratio between affection of trunk and extremities, while linear types of scleroderma have a peripheral location.

As it appears from Table III, symmetry is common in GM in contrast to other types of scleroderma including LMP. Many plaques in patients with morphoea exhibited linearity.

As it appears from Table IV, the degree of sclerosis was frequently advanced in LMP as compared to GM, where plaques typically exhibited slight sclerosis.

As it appears from Table V, increased pigmentation of the lesions was frequent in all types of localized scleroderma. The pigmentation was either spotty, central, peripheral or diffuse. Some plaques of morphoea, preferably LMP, showed faint redness. In a few cases, though, redness was intense. Visible atrophy of the subcutaneous tissue was uncommon in morphoea but frequent in linear types.

As it appears from Table VI, increased pigmentation of the perilesional area was

Table VII. *40 pigmented spots observed in 21 patients with localized scleroderma (generalized morphoea patients not included), number and regional distribution*

	Pigmented spots No. of patients	No. of spots and percent of scleroderma lesions	Regional distribution		
			trunk	Extremity	Other
Morphoea					
Localized plaque	11	21[a] (44.7%)	18	3	–
Linear types					
Extremity and trunk[b]	6	13 (59.1%)	10	2	1
En coup de sabre[c]	4	6 (27.3%)	6[d]	–	–
Total	21	40 (44.0%)	34	5	1

[a] In 6 spots underlying atrophy of subcutaneous tissue.
[b] In one case pigmented coup de sabre line with no evidence of sclerosis.
[c] In 3 cases contralateral pigmentation en coup de sabre.
[d] Two spots presenting underlying atrophy of subcutaneous tissue.

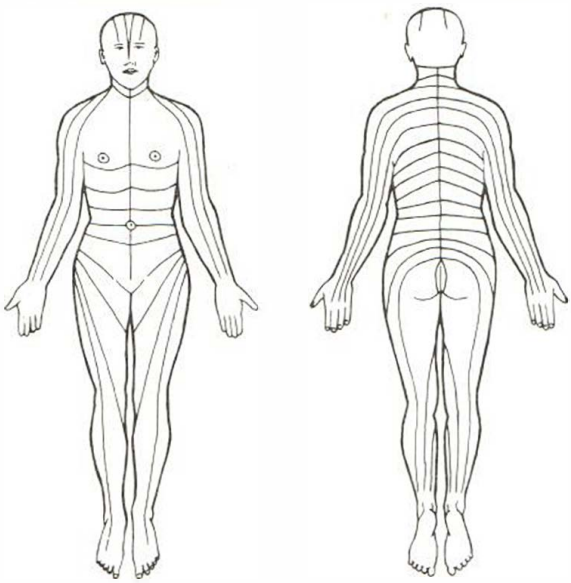

Fig. 2. Linearity in localized scleroderma according to the present study. A typical pattern of linearity was not observed on the upper chest, on the lower abdomen, and in the lower face. Palms, soles and ears were not involved.

frequent in morphoea, in particular GM, in which pigmentation was often extensive. Lilac ring, an inflammatory zone, was observed around some of the plaques. Visible atrophy of the subcutaneous tissue occurred around a few plaques.

As it appears from Table VII, PS were found in about half of the patients with LMP and LET, while these separate lesions of disturbed pigmentation seemed less frequent in LCS. In all types of localized scleroderma PS were mainly located to the trunk, in some cases associated with underlying atrophy of the subcutaneous tissue.

X-ray examinations showed spina bifida in 4 patients out of 36 examined, in one associated with arcolysis and spondylolisthesis. Spina bifida occurred in two patients with LMP, one patient with LET and one with LCS.

Registrations made according to history were the following:

Diseases in close relatives. Rheumatoid arthritis 1, rheumatic fever 1, systemic lupus erythematosus 1, synovitis 1, migraine 1, multiple sclerosis 1, congenital myotonia 1, diabetes mellitus 1, contact dermatitis 1, atopic dermatitis 1.

Previous diseases in the patients. Rheumatoid arthritis 1, rheumatic fever 2, arthritis urica 1, osteoarthritis 2, Scheuermann disease 2, herniated lumbar disc 2, epiphysiolysis of the tibia 1, migraine 3, hypersedimentation 1, contact dermatitis 4, atopic dermatitis 1, urticaria 1, psoriasis 1.

Possible provocatory factors. Menarche 4, pregnancy 3, menopause 1, loss of weight (29 kg) 1.

Initial and associated symptoms and diseases. Arthralgia 6 (LMP 2, GM 1, LET 2, LCS 1), Raynaud's phenomenon 6 (GM 3, LET with extensive involvement of one extremity 3), guttate morphoea 4 (LMP 2, GM 2), lichen sclerosus et atrophicus 2 (GM 2).

DISCUSSION

In spite of variability and overlap in clinical manifestations, different types of localized scleroderma were found to exhibit characteristic features.

Disturbances of pigmentation occurred in most patients. A special lesion, the pigmented

spot, was found in half of the patients, in particular on the trunk. Recently, the author found the skin thickness of such spots to be reduced, as measured by pulsed ultrasound (7). In the study of Christiansen et al. café au lait spots were registered in 8 (3.4%) of the patients with localized scleroderma. Pigmented spots seem undistinguishable from or a special type of atrophorderma Pasini-Pierini (8, 9). In a later study, Pierini et al. reported on atrophoderma associating morphoea, which they named "l'atropho-sclérodermie" (10). Idiopathic atrophy may be segmental (11).

The skin lesions of morphoea patients exhibited linear and band-like features, symmetric and midline affection in a significant number of cases. Based on charts of the patients of this study a regional distribution chart of linear affection in localized scleroderma was elaborated (Fig. 2). This chart is in good agreement with illustrations of cases of linear scleroderma published by Rubin (6). The lines of the chart are different from the lines of the skin of Langer, the innervation fields of peripheral and cranial nerves, and the lines of Voigth between innervated fields. The chart shows several similarities with the innervation fields of the spinal roots according to Foerster and according to Keegan & Garrett (12, 13). However, spinal innervation fields cannot explain the rare though characteristic lesions of the nipples, the umbilicus, and the coup de sabre lines of the front. The chart also shows similarities with the Blaschko chart of linear dermatoses elaborated from observation of patients with different linear and segmental dermatoses also including cases of herpes zoster (14). V- and S-shaped lines of the trunk indicated by Blaschko were not seen in this study of localized scleroderma.

Bettmann and later Stava assumed that the lines seen in localized scleroderma have an embryonal background (15, 5). Rubin's finding of a high frequency of spina bifida in selected and severe cases of linear scleroderma might support this view (6). During embryonal life and growth, mesenchymal cells originating in the somites proliferate in a transverse direction to form the trunk and a longitudinal direction to form the extremities, followed by migration of primitive melanocytes and ectodermal cells from the neural crest to form melanocytes of the skin and peripheral nerves. Recently, Johnston described the migration pattern of crest cells in the cranium, following vertical lines in the front much alike the coup de sabre lines (16). The author has reported two cases of scleroderma en coup de sabre with eye involvement at the same line, both indicating an embryonal predisposition related to the migration of crest cells (17, 18). Various segmental disorders of pigmentation—some associated with neural defects—are assumed to be due to defects in the migration of neural crest cells during embryonal life (19). Patients with localized scleroderma seem to have no increased incidence of neurological defects and diseases. However, some studies have shown a high occurrance of abnormal EEG (20, 21, 22, 23, 24, 25). A case of multiple morphoea plaques of the face, abnormal EEG and convulsive fits have been reported (26).

Altogether, the high frequency of disturbances of pigmentation in localized scleroderma, affections of underlying anatomical structures, and the special linear distribution on the trunk, the extremities, and in the front with lines radiating to the spinal region may indicate a predisposing defect in crest cell migration during embryonal life.

REFERENCES

1. Helwig EB, Piper WN. Progressive systemic sclerosis—visceral manifestations in generalized scleroderma. Arch Dermatol 1955; 72: 535–46.
2. O'Leary PA, Nomland R. A clinical study of one hundred and three cases of scleroderma. Am J Med Sci 1930; 180: 95–122.
3. Christianson HB, Dorsey CS, O'Leary PA, Kierland RR. Localized scleroderma—a clinical study of two hundred and thirty-five cases. Arch Dermatol 1956; 74: 629–39.

4. Curtis AC, Jansen TG. The prognosis of localized scleroderma. Arch Dermatol 1958; 78: 749–757.
5. Stava Z. Zirkumskripte Sklerodermie. Dermatol Wochenschr 1957; 139: 513–23.
6. Rubin L. Linear scleroderma—association with abnormalities of the spine and nervous system. Arch Dermatol Syph 1948; 58: 1–18.
7. Serup J. Decreased skin thickness of pigmented spots appearing in localized scleroderma (morphoea)—measurement of skin thickness by 15 MHz pulsed ultrasound. Arch Dermatol Res 1984: 276: 135–7.
8. Canizares O. Idiopathic atrophoderma of Pasini and Pierini. Arch Dermatol 1959; 79: 614–5.
9. Jablonska S, Szczepanski A. Atrophoderma Pasini-Pierini. In: Jablonska S, ed. Scleroderma and pseudoscleroderma Warsaw: Polish Medical Publishers, 1975; 521–36.
10. Pierini LE, Abulafia J, Mosto SJ. Atrophodermie idiopathique progressive et états voisins. Ann Derm Syph (Paris) 1970; 97: 391–416.
11. Epstein E. Linear segmental atrophy. Arch Dermatol 1956; 74: 411–13.
12. Foerster O. The dermatomes in man. Brain 1933; 56: 1–39.
13. Keegan JJ, Garrett FD. The segmental distribution of the cutaneous nerves in the limbs of man. Anat Rec 1948; 102: 409–37.
14. Blaschko A. Die Nervenverteilung in der Haut in ihrer Beziehung zu den Erkrankungen der Haut—Beilage zu den Verhandlungen der Deutschen Dermatologischen Gesellschaft VII Congress zu Breslau im Mai 1901. Wien, Leipzig: Wilhelm Braumüller, 1901.
15. Bettmann S. Bandförmige Sklerodermie und Naevuszeichnung. Arch Dermatol Syph 1922; 142: 235–51.
16. Johnston M. The neural crest in abnormalities of the face and brain. In: Bergsma D, Langman J., Paul NW, eds. Morphogenesis and malformation of face and brain. Original Article Series vol. XI, No. 7. New York: Alan R. Liss, 1975: 1–18.
17. Serup J, Alsbirk PH. Localized scleroderma en coup de sabre and irido-palpebral atrophy at the same line. Acta Derm Venereol (Stockh) 1983; 63: 75–77.
18. Serup J, Serup L, Sjö O. Localized scleroderma en coupde sabre with external eye muscle involvement at the same line. Clin Exp Dermatol 1984; 9: 196–200.
19. Metzker A, Morag C, Weitz R. Segmental pigmentation disorders. Acta Derm Venereol (Stockh) 1983; 63: 167–70.
20. Taylor RM, Pacella BL. The electroencephalogram in scleroderma. J Nerv Ment Dis 1949; 109: 42–47.
21. Harnack K. Zerebrale Symptome bei Hemiatrophia faciei progressiva und Sklerodermia circumscripta. Dermatol Wochenschr 1962; 145: 46–50.
22. Stava Z. Stein J. Electro-encephalography in scleroderma. Dermatologica 1961; 123: 375–90.
23. Zimmermann H, Müller D, Schäfer A. Zur Pathogenese der Sklerodemie unter Berücksichtigung bioelektrischer Untersuchungen. Dermatol Wochenschr 1965; 151: 858–62.
24. Sollberg G. Neurologische und elektrophysiologische Untersuchungen bei progressiver Sklerodermie und Morphaea. Arch Klin Exp Dermatol 1967; 229: 20–32.
25. Gottwald E, Sturm U. EEG-Befunde bei 44 ausgewählten Patienten mit Sklerodermie. Nervenarzt 1981; 52: 219–27.
26. Dickmans-Burmeister D. Multiple Herde von Morphaea mit cerebralen Anfallsleiden. Z Hautkr 1972; 47: 71–86.