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High-resolution Magnetic Resonance Imaging in Patients with Facial Haemiatrophy

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The aim of this clinical study was to evaluate the use of high-resolution magnetic resonance imaging (MRI) in the diagnosis of facial haemiatrophy. A total of 14 patients with clinically suspected facial haemiatrophy were investigated using high-resolution MRI. The T1- (500/25) and T2- (2200/50) weighted images were analysed visually and numerically. The results of the affected skin portions were compared with the contralateral skin and correlated with the clinical results. The subcutis could not be delineated by high-resolution MRI in 9 patients with facial haemiatrophy. The dermis was not discernible in 6 cases and was "smooth" in a further 6 patients. The signal-to-noise ratio of affected skin portions and contralateral skin or clinical severity did not correlate. The higher the clinical severity, the more pronounced was the magnetic resonance ratio between dermis and subcutis thickness. Thus high-resolution MRI is a useful method for objective description of pathological changes in clinically suspected facial haemiatrophy. ***Key words: surface coil; facial haemiatrophy.***

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Progressive facial haemiatrophy (PFH) or Romberg's disease was first described by Parry in 1824 (1–3) and Romberg in 1846 (4). Typically, the first symptoms occur in early adulthood, but they have also been described in the first few months of life (5). Most common as the early signs of disease are circumscribed skin lesions with consecutive reduction of subcutical fat, followed by muscle and bone involvement. Central or peripheral neurological signs may also occur. The skin alterations typically progress over 2–10 years, while neurological symptoms appear later or progress over a longer period.

The aim of this study was to describe the morphological changes of cutaneous lesions of PFH by high-resolution magnetic resonance imaging (MRI) and to analyse whether there was a correlation between MRI findings and clinical disease severity.

MATERIALS AND METHODS

A total of 14 patients (9 females and 5 males) with clinically suspected PFH were examined by high-resolution MRI. The mean age of the patients was 30 years (range 5–62 years). Magnetic resonance images were acquired at the site of the clinically most extensive skin alterations. The location was the forehead in 7 patients, temples in 5 and the cheek in 2.

All examinations were conducted using a 1.5 T Magnetom SP 63 (Siemens, Erlangen, Germany). A linearly polarized receiver coil with 1 coil loop and a diameter of 2.5 cm was used (Siemens, Erlangen, Germany). The coil has an integrated pre-amplifier with optimized noise factor to reduce cable-related signal loss. Patients were scanned in the supine position. The patient's head was positioned with the skin lesion directed upward. The corresponding skin portion on the other side of the head was later scanned in the same mode. T1- (500/25) and T2- (2200/50) weighted transverse images with a "field of view" of

Table I. *Summary of patient data, clinical and magnetic resonance imaging (MRI) staging, thickness and signal noise ratios (SNR) of the affected and normal sides*

Patient	Stage	Stage	$Thickness_{affected}/Thickness_{normal}$			Epidermis-$SNR_{affected}/SNR_{normal}$		Subcutis-$SNR_{affected}/SNR_{normal}$	
Age	Clinical	MRI	Epidermis	Dermis	Subcutis	T1 image	T2 image	T1 image	T2 image
f 30	1	2	0.70	0.65	0.75 (cheek)	1.47	–	0.90	–
f 52	1	1	0.85	1.05	0.33	0.31	0.44	0.42	0.35
f 23	1–2	2	0.44	0.32	0.69 (cheek)	0.54	0.90	0.95	1.05
f 27	2	3	0.95	0.50	0	0.91	0.71	1.00	0.74
m 62	2	1	0.76	0.92	0.28	0.98	0.57	1.12	0.91
m 11	2	3	1.11	Discernible	0	0.53	–	–	–
m 11	2	3	0.60	Discernible	0	0.91	0.71	0.75	0.41
f 60	2–3	2	1.06	0.60	0.09	1.2	–	0.47	–
f 45	3	3	0.67	0.50	0	0.79	0.75	0.47	0.37
f 25	3	3	0.43	0.43	0	0.56	0.41	0.78	0.80
f 19	2–3	3	0.54	Discernible	0	0.93	0.74	1.12	0.67
f 38	3	3	0.45	Discernible	0	0.48	0.58	0.45	0.62
m 11	2	3	0.53	Discernible	0	0.64	0.78	0.25	0.18
m 5	3	3	1.00	Discernible	0	0.37	1.01	0.68	0.76
Median			0.685	0.55		0.715	0.71	0.75	0.67

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25 mm and 3 acquisitions were acquired. The imaging matrix was 256×256 pixels, which resulted in a spatial resolution of 0.098×0.098 mm at a 25 mm FOV. The slice thickness was 2 mm. The number of slices was 6.

T1 and T2 images with maximal representation of the lesion were selected for evaluation (i.e. the thinnest layer). The thickness of all 3 skin layers was measured and derived as the mean of 5 measurements. The thickness of the layers was measured on the basis of the magnetic resonance images using a specially designed computer program. The skin was visually rated as interdigitated, smooth, or not discernible from the epidermis. Signal intensity was measured in all 3 skin layers and in the background with SD (noise). The signal to noise ratio ($SNR = SI_x - Si_{background}$/noise) was calculated in the T1- and T2-weighted images. Finally, the skin thickness and SNR of the affected side were compared with the normal side, calculating thickness and SNR ratios. All data were reproduced as medians. A skin biopsy was performed only in 4 cases with clinically suspected borreliosis.

Concerning the morphological findings in high-resolution MRI we defined 3 stages: stage 1 was defined as a thinner subcutis than the unaffected side, with an otherwise normal appearance and interdigitating dermal border zone. This was graded as a mild change. In stage 2 the subcutis was thinner than on the normal side and the dermal border zone had a smooth contour. Stage 3 was characterized by a missing subcutis and a smooth dermis and was graded as skin skeletonization.

RESULTS

Using clinical criteria, 2 patients were in stage 1, 5 in stage 2, 4 in stage 3, 1 in stage 1–2 and 2 in stage 2–3. Suspected borreliosis was not corroborated in the 4 biopsies.

In 9/14 patients 2 and in 5/14 patients 3 skin layers of different thickness were discernible by high-resolution MRI. In these cases the epidermis appeared as an intermediate signal, the dermis as a low signal with an interdigitating border zone and the subcutis as a high signal layer with connective tissue (Figs 1–2). In 9 patients, there was a complete loss of subcutis signal and the interdigitating border zone of the dermis, so that the SNR ratio could not be determined. This resulted in a "smooth" contour of the dermis, which was not detectable in 6 cases. In the remaining

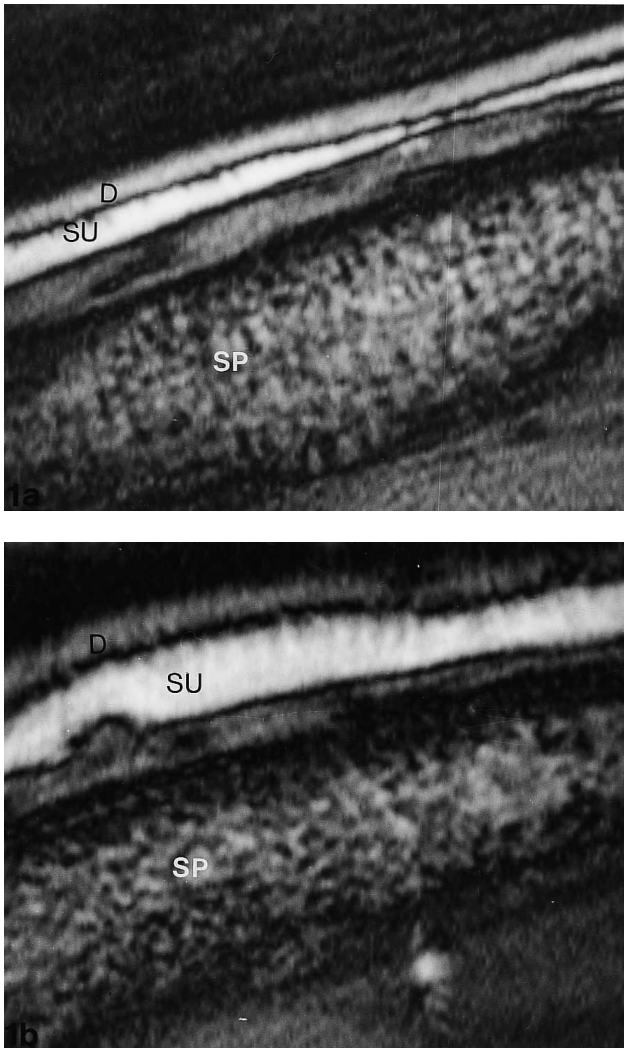

Fig. 1. A 60-year-old woman with clinical stage 2–3 progressive facial haemiatrophy. Magnetic resonance imaging stage 2. MRI of the skin (temple). T1-weighted transverse image (500/25, ACQ 3, matrix 256×256, slice thickness 2 mm). (*a*) Affected side. Both subcutis and dermis are thin, in particular, there is a variable subcutical thickness. There is no dermal interdigitating. There is a chemical shift artifact between the dermis and the subcutis. The epidermis cannot be discerned. (*b*) Healthy contralateral skin. Hypointense dermis, underlying subcutis. D: dermis; SU: subcutis; SP: spongiosa.

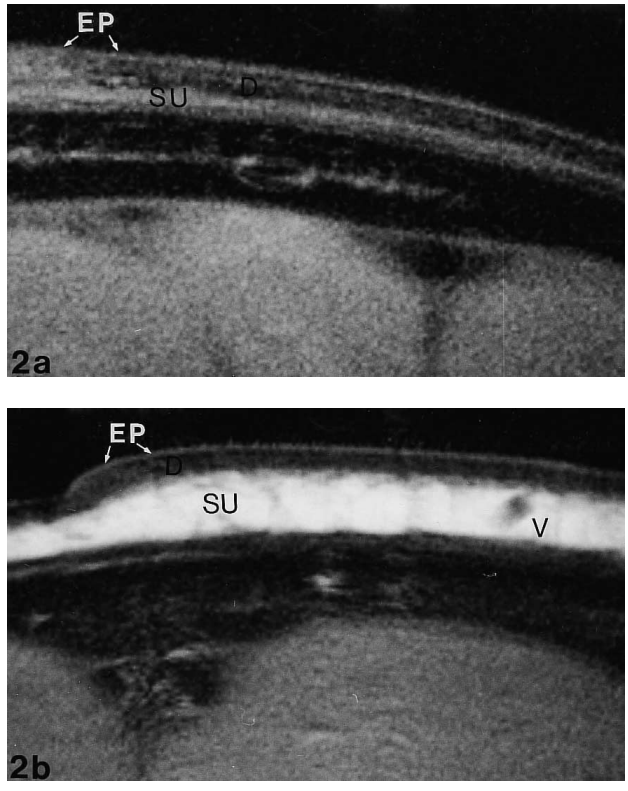

Fig. 2. An 11-year-old boy with clinical stage 2 progressive facial haemiatrophy. Magnetic resonance imaging stage 3. MRI of the skin (frontal bone). T1-weighted transverse image (500/25, ACQ 3, matrix 256×256, slice thickness 2 mm). (*a*) The subcutis can be distinguished with definitely reduced thickness at some sites only. Minimal chemical shift artifact. Reduced dermal thickness and loss of interdigitating. (*b*) Unaffected contralateral side. Three-layered skin: hyperintense epidermis, hypointense dermis; underlying subcutis. EP: epidermis; D: dermis; SU: subcutis; V: vessel.

5 cases the thickness ratios of the subcutis varied between 0.09 and 0.75, demonstrating a lower subcutis thickness and less connective tissue on the affected side. The thickness ratios of the dermis showed similar results, correlating with a lower thickness of the affected side. In 2 patients, the cheek, and not the forehead or the temple was affected, as in all other patients. The subcutis of the cheek is thicker than that of the forehead or temple, which may explain the relatively high subcutical ratio of 0.75 or 0.59. The calculated SNR ratios (epidermis and subcutis) of affected skin portions and contralateral skin did not correlate with clinical severity (Table I).

In 9 patients MRI stage 3, in 3 patients stage 2 and in 2 patients stage 1 was found. The MRI grading system partially overestimated the clinically severity of the disease in stage 3. In summary, the results of the MRI grading system emphasized the good correlation between the delineation of the skin layers and the clinical findings.

DISCUSSION

Wartenberg (2) has called the vanishing subcutical layer a "key" symptom of PFH. In severe cases, large areas of complete fat tissue involution may occur, with resulting visualization of the bony skull in great detail ("skeletonization"). Concerning the clinical findings in our MRI grading system, stage 3 was defined as a finding in which the subcutis could not be discerned at all. The thickness of epidermis, dermis and subcutis were measured and the respective ratios of the affected to the unaffected side calculated.

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