Appendix S1

SUPPLEMENTARY MATERIALS AND METHODS

This is a sub-study of the European Network for Localized Scleroderma funded by the European Academy of Dermatology and Venereology (EADV) in 2012. A retrospective review was performed using the digital databases of LS patients with linear subtypes, such as LSECDS, PFH, or a combination of both types. These patients were diagnosed and treated at 5 German tertiary referral centres for LS (Bochum, Erlangen, Köln, Oberhausen, and Wuppertal) over an observation period of 17 years (1 January 2000, through 1 January 2017). The local ethics review boards approved the study. To be eligible for this retrospective study, the typical clinical criteria for LSECDS or PFH had to be present, as previously published (S1). A diagnosis of LSECDS was made in patients with band-like sclerotic or atrophic lesions located on the frontoparietal region, ranging paramedian from the eyebrows into the hair-bearing scalp (S1). PFH was defined as atrophy of the subcutaneous tissues of the unilateral face with minimal or absent overlaying cutaneous changes (S2). If clinical features of both conditions were present, a diagnosis of LSECDS/PFH overlap was made. Punch biopsies for histopathological analysis were taken only in patients with an inconclusive clinical status. In all patients, a detailed medical history (including current medication, rheumatic or other autoimmune diseases, and other co-morbidities), physical examination, and clinical inspection of the entire skin were performed.

Neurological analysis

Neurological involvement was classified as epilepsy/seizures, headache, migraine, vision involvement, and cranial nerve involvement. Radiological screening for cerebral/CNS involvement was performed using MRI or CT. CNS abnormalities were then classified as areas of gliosis, white matter lesions, abnormal cortical size and folding, and vascular malformation/abnormality, as reported previously (S3).

Serological evaluation

Standard serological analysis was performed once at first diagnosis of LS in all 96 patients and included a complete blood cell count, antinuclear antibodies (ANA; a titre≥1:160 was considered positive), screening for antibodies against extractable nuclear antigens (ENA, including anti-Ro and anti-La antibodies, anti-Smith antibodies, anti-U1-ribonucleoprotein antibodies, anti-histone antibodies, anti-SCL-70 antibodies, anti-centromere antibodies, and anti-Jo-1 antibodies), anti-double-stranded deoxyribonucleic (anti-Ds-DNA) antibodies (performed by enzyme immunoassay), anti-smooth muscle antibodies (ASMA), rheumatoid factor, circulating immune complexes (CIC), anti-thyroid antibodies (thyroid peroxidase and anti-human thyroglobulin antibodies), complement components 3 and 4, C-reactive protein, and routine blood chemistry testing.

SUPPLEMENTARY REFERENCES

S1. Kreuter A. Localized scleroderma. Dermatol Ther 2012; 25: 135–147.
S2. Laxer RM, Zulian F. Localized scleroderma. Curr Opin Rheumatol 2006; 18: 606–613.
S3. Amaral TN, Peres FA, Lapa AT, Marques-Neto JF, Appenzeller S. Neurologic involvement in scleroderma: a systematic review. Semin Arthritis Rheum 2013; 43: 335–347.