

Oral presentation

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13.4 High frequency of CNS involvement in linear scleroderma of the face

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Background

Single case reports have reported neurological symptoms or cerebral imaging abnormalities in linear scleroderma of the face (LSF). Aim of the study was to investigate the frequency and clinical features of CNS involvement in a large cohort of patients with LSF.

Methods

Patients included in the JUVenile Scleroderma International (JUSI) database of PRES with LSF were evaluated. Clinical and laboratory information were analyzed in order to define frequency, type, timing of onset and possible risk factors of CNS involvement.

Results

121 patients with LSF, followed at 12 centers entered the study. F:M ratio was 2, 1:1, mean age at onset 5, 8 years.

Among 77 patients whose we obtained complete clinical and instrumental updated documentation (MRI, CT scan and EEG), 46 (59.7%) had CNS involvement. 33 (42.9%) presented with symptoms such as epilepsy in 16 (20.8%), chronic headache in 11 (14.3%), hemiparesis with or w/o epilepsy in 3 (3.9%) or behavioural disorders in 3 (3.9%). Five patients (15.1%) had seizures before the onset of the skin changes, 39.4% presented neurologic symptoms by

the first year and 84,8% by 5 years since the disease onset. 13 asymptomatic patients presented only EEG or cerebral imaging abnormalities. All these findings were ipsilateral to the skin lesion. No significant correlation was found between CNS involvement and autoantibodies, age at onset, gender, family history for autoimmune diseases and environmental factors.

Conclusion

LSF is frequently associated with CNS abnormalities. A careful clinical and instrumental evaluation should be recommended for every patient at onset and during the follow-up.