Pediatric Rheumatology

Poster presentation

Extracranial linear localized scleroderma associated with longstanding epileptic encephalopathy unresponding to methotrexate and prednisone

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We report a boy, who achieved developmental milestones without delay, presenting complex partial seizures (left-deviation of the head, staring gaze, oral automatism and tonic-clonic generalization) at 7 years: his blood/cerebrospinal fluid tests were all within normal limits, enclosed those for inborn errors of metabolism. Progressive signs of psycho-motor regression were simultaneously observed by caregivers in combination with a band of linear ivory-coloured indurated lesion on the dorsal region of the right foot, which was diagnosed as linear localized scleroderma (LS). Immunological tests performed at 8 years revealed only low-titre positivity of anti-nuclear antibodies. Various brain CT and MRI scans resulted negative. In the following years tonic-clonic seizures were observed with daily recurrence in spite of different antiepileptic drugs. At 16 years we evaluated this patient for the first time: asymmetrical tonic fits had multiple daily frequency and therapy consisted of clobazam/methsuximide; EEG revealed dysrupted electrical cerebral activity; brain MRI was normal; the skin lesion was extended to the whole right inferior limb. Methotrexate (MTX) and prednisone were instituted (respectively at the dosage of 15 mg/m^2/week and 1.5 mg/kg/day): MTX was maintained for 1,5 year with low-dose prednisone (0.5 mg/kg/day). Semeiological features of seizures remained unchanged and no control upon their frequency could be reached, whilst LS remained stable.

The exact nature of multi-resistant epilepsy associated with extra-cranial linear LS remains a matter of debate and the immunologic process leading to the pathologic collagen deposition might be dissociated from the specific neurological disturbance.