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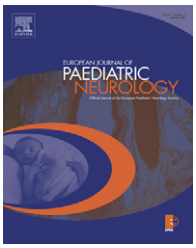
Official Journal of the European Paediatric Neurology Society



Case study

Hippocampal atrophy and developmental regression as first sign of linear scleroderma "en coup de sabre"

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ARTICLE INFO

Article history:
Received 29 May 2007
Received in revised form
20 August 2007
Accepted 7 December 2007

Keywords:
Hippocampal atrophy
Developmental regression
Linear scleroderma
Children

ABSTRACT

An 8-year-old girl with linear scleroderma "en coup de sabre" is reported who, at preschool age, presented with intractable simple partial seizures more than 1 year before skin lesions were first noticed. MRI revealed hippocampal atrophy, controlaterally to the seizures and ipsilaterally to the skin lesions. In the following months, a mental and motor regression was noticed. Cerebral CT scan showed multiple foci of calcifications in the affected hemisphere. In previously reported patients the skin lesions preceded the neurological signs. To the best of our knowledge, hippocampal atrophy was not earlier reported as presenting symptom of linear scleroderma. Linear scleroderma should be included in the differential diagnosis in patients with unilateral hippocampal atrophy even when the typical skin lesions are not present.

1. Introduction

Linear scleroderma "en coup de sabre" is a localised type of scleroderma affecting the face and the head in a linear pattern. Involvement of underlying structures may cause facial hemiatrophy and deformity.[1] Acquired progressive facial hemiatrophy, also named Parry-Romberg syndrome is often thought to be an overlapping condition with linear scleroderma "en coup de sabre".[2]

In patients with linear scleroderma "en coup de sabre" as well as in those with Parry-Romberg syndrome neurological symptoms such as epilepsy and mental retardation have been reported before. Cerebral imaging shows ipsilateral cerebral lesions. In most cases, the characteristic skin involvement is recognised before the development of neurological disease.[3,4] Very few cases are described with epilepsy as first clinical symptom.[5,6]

We report an 8-year-old girl with linear scleroderma "en coup de sabre" presenting at preschool age with intractable simple partial seizures and regression more than 1 year before skin lesions were first noticed.

2. Case study

The proposita was born at term from non-consanguineous parentage. The family history was uneventful. She never presented with febrile seizures. Milestones were reached at normal ages. Her initial motor and mental development was normal. At age 30 months she started at the nursery school and performed normally for her age.

At the age of 34 months she presented with simple partial seizures. She first had a sensation of pain in the left corner of the mouth and the left arm, rapidly followed on most

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doi:10.1016/j.ejpn.2007.12.001

occasions by twitching of the left corner of the mouth and the left arm. She remained conscious during seizures. The episodes lasted from seconds to minutes. Initially the frequency was once a week, later there were several episodes in 1 day. Between seizures she complained of headaches and was irritable. Clinical examination was normal. Routine analyses of blood and cerebrospinal fluid were normal.

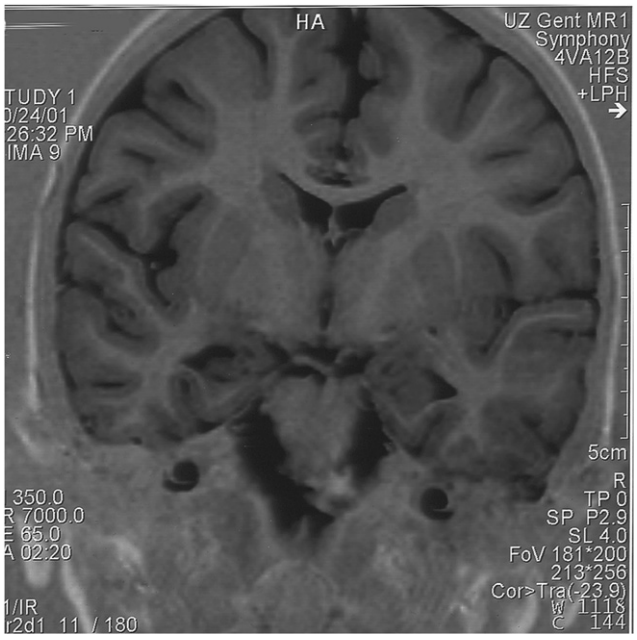

Fig. 1 – MRI (age 35 months) showing right hippocampal atrophy.

Electroencephalography revealed a right temporal focus with sharp slow-waves. A focus with the same localisation was seen on SPECT and PET scan. Cerebral MRI, within the first month after seizure onset, revealed hippocampal atrophy at the right side (Fig. 1). No significant signal changes were noticed on T2 or FLAIR sequences.

The epilepsy was resistant to several anti-epileptic medications (carbamazepine, clobazam, vigabatrine, sulthiam, clonazepam, topiramate, lamotrigine, nitrazepam and sodium valproate). Finally, she became seizure free after 1 year with a combination of sodium valproate, nitrazepam and topiramate.

Since the onset of her epilepsy, a regression of her cognitive functions had been noticed. This was the case even when she became seizure free, and when the doses of the anti-epileptic medications were lowered. While she performed well for her age in the first year of nursery school, she had a significant delay in her second year. Her total IQ (SON-R) at age 5 year was estimated to be 61. From then on she followed special education, first in a school for children with mild cognitive delay, later on for children with a moderate delay.

At the age of 4 years, her mother detected a localised area of alopecia in the right frontal zone of the scalp (Fig. 2a). The skin was ivory-coloured, with the clinical characteristics of a sclerotic plaque. Despite treatment with high-potency topical corticosteroids, a slow progression of the skin lesion was seen with extension to the forehead, the medial corner of the right eye and the right lateral side of the nose. The nose became mildly asymmetric with a smaller right nostril and mild retraction of the right upper lip (Fig. 2b). At that moment she was diagnosed

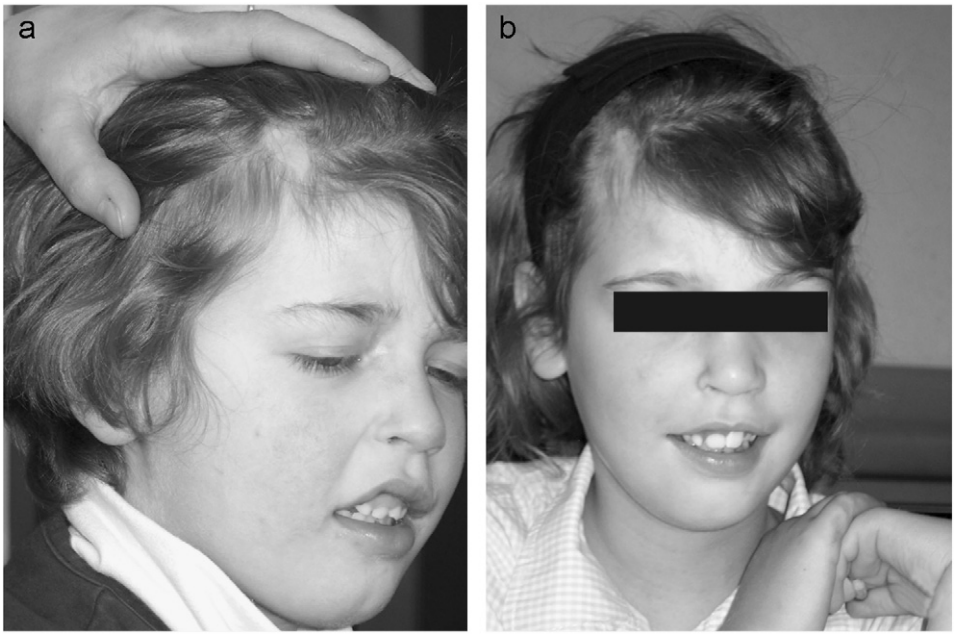

Fig. 2 – (a) Forty-nine months: ivory-coloured, sclerotic plaques right frontal on the scalp. (b) Eight years: progression of the skin lesion extending to the forehead, the medial corner of the right eye and the right lateral side of the nose. Mild asymmetry of the nose with smaller right nostril and mild upper retraction of the right upper lip.