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Progressive inflammatory lesions of the brain parenchyma in localized scleroderma of the head

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Summary. A patient with localized scleroderma of the head, uveitis, and Raynaud's phenomenon presented with generalized seizures, spastic hemiparesis, and local IgG production in the cerebrospinal fluid. Magnetic resonance imaging revealed progressive cortical and subcortical brain parenchymal lesions mainly adjacent to the cutaneous and bony lesions and probably of inflammatory origin.

Key words: Localized scleroderma – Magnetic resonance imaging – Central nervous system inflammation – Intrathecal IgG production

Introduction

Scleroderma is a heterogeneous group of chronic collagen tissue diseases which range from localized scleroderma to progressive systemic sclerosis (PSS). In addition to cutaneous manifestations, numerous forms of organ involvement are encountered. Overlapping syndromes with other connective tissue diseases such as systemic lupus erythematosus or dermatomyositis have also been described.

There have been only a few reports of central nervous system (CNS) involvement in both PSS and localized scleroderma [1–3]. One case was reported with acute necrotizing encephalitis associated with PSS [5]. Other pathological studies of the brain in PSS have revealed no distinctive pathology [9]. In localized scleroderma post-mortem brain examinations have been exceedingly rare [10]. On the other hand there have been several clinical observations on CNS manifestations in this disease, which have consisted mainly of different focal symptoms and electroencephalographic (EEG) abnormalities [4].

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We report here a patient with scleroderma and concomitant intrathecal IgG production where magnetic resonance imaging (MRI) revealed extensive brain abnormalities mainly adjacent to the cutaneous manifestations.

Case report

The 30-year-old female patient presented with generalized seizures of 2-year duration. She had had localized scleroderma with a band-like sclerotic lesion over her left forehead and parietal region (sclerodermie en coup de sabre) for 22 years. Also, a slowly progressive Raynaud's phenomenon had been observed over the course of 9 years. Transient visual loss had occurred in the left eye 2 years and in the right eye 1.5 years prior to admission, both with incomplete remission.

Clinical examination and plain radiography revealed circumscript skull atrophy beneath the localized scleroderma of the left forehead and parietal region as well as alopecia areata and enophthalmos due to loss of retro-orbital fat. Another sclerodermal lesion with alopecia areata was found in the right parietal region. There was a slight spastic hemiparesis of the right side. Ophthalmological examination disclosed evidence of previous periphlebitis of the right eye and pallor of the optic disc as well as subacute uveitis in the left eye.

Routine laboratory testing and extensive serological screening including anti-centromere and anti-Scl-70 antibodies were within normal limits except for an elevated antinuclear antibody titre, which is not infrequent in localized scleroderma [6]. Cerebrospinal fluid analysis was normal with regard to cell count, cell differentiation and quantitative protein analysis (albumin, immunoglobulins G, A and M). However, oligoclonal IgG was detected by isoelectric focusing, indicating local IgG production within the CNS. EEG was slightly dysrhythmic without focal alterations.

A fist MRI scan had revealed several hyperintense patchy areas extending throughout the cortical and subcortical brain parenchyma mainly in the left hemisphere. The hyperintense areas of the left hemisphere were located immediately beneath the sclerotic cutaneous and skull lesions. There was also some spatial connection between the right hemispherical hyperintense area and the right parietal sclerodermal lesion (Fig. 1a, b). Two years later, a repeat MRI showed that the right hemispherical lesions had almost disappeared, whereas cortical alterations of the left hemisphere were enlarged and confluent. In addition, a cystic parenchymal defect

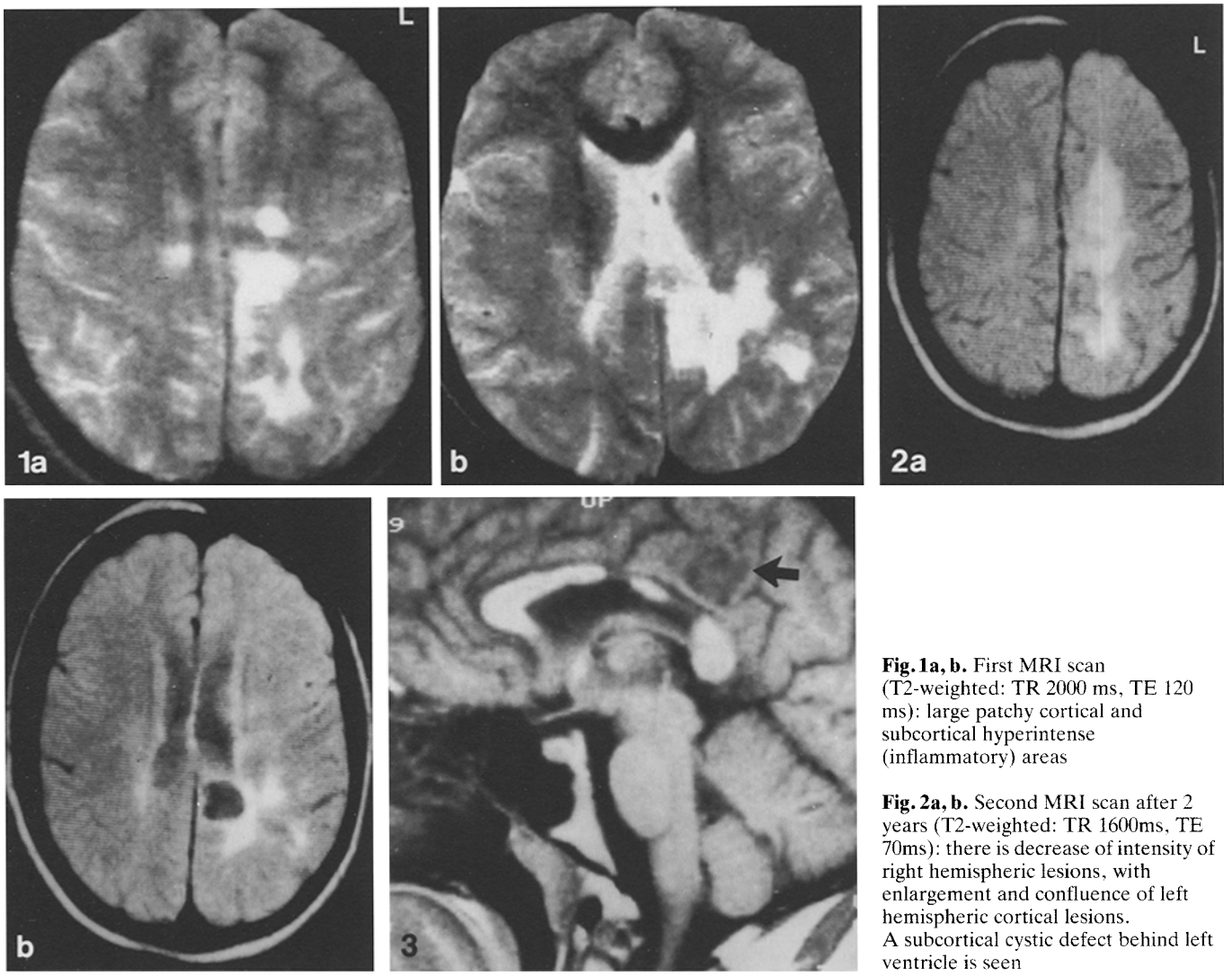

Fig. 1a, b. First MRI scan (T2-weighted: TR 2000 ms, TE 120 ms): large patchy cortical and subcortical hyperintense (inflammatory) areas

Fig. 2a, b. Second MRI scan after 2 years (T2-weighted: TR 1600ms, TE 70ms): there is decrease of intensity of right hemispheric lesions, with enlargement and confluence of left hemispheric cortical lesions. A subcortical cystic defect behind left ventricle is seen

Fig. 3. Second MRI scan, as in Fig. 2 (T1-weighted: TR 400 ms, TE 22 ms). Defects shown in the corpus callosum, the more rostral one in spatial contact with the subcortical cystic defect *(arrow)*

had developed behind the left lateral ventricle (Fig. 2a, b). Also, there were two defects in the corpus callosum (Fig. 3).

Discussion

To our knowledge this is the first report of a case of scleroderma where extensive circumscript brain parenchymal lesions have been detected by MRI which are localized at least in part immediately adjacent to cutaneous and bony alterations. Our observation is well in accord with former reports on focal neurological deficits, EEG abnormalities and findings of parenchymal involvement at autopsy. The clinical symptomatology of our case extended beyond the picture of localized scleroderma. It included symptoms of limited scleroderma (uveitis, Raynaud's phenomenon) [7].

In our opinion, the cerebral manifestations were not of recent origin, in particular since MRI findings showed comparatively little change over an observation period of 2 years. It might be that they developed simultaneously with the adjacent cutaneous alterations. Moreover, it has been speculated that the cutaneous manifestations are induced by primary cerebral malformations [8].

On the other hand, the local IgG production in the cerebrospinal fluid indicated an active inflammatory reaction in the CNS. Most likely this is attributable to the circumscribed brain parenchymal lesions, especially since no clinical signs or serological findings of an overlap syndrome or PPS were found. Also, there was no indication of an inflammatory CNS disorder of different aetiology. In particular, multiple sclerosis is unlikely considering the corpus callosal and cystic parenchymal defects. The appearance of seizures, subacute uveitis and Raynaud's phenomenon certainly are further indicators of disease activity. More attention should be paid to the question as to whether other localized scleroderma manifestations of the head are also connected with progressive intracerebral inflammatory reactions.

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