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Case report

Mild Cognitive Impairment as a single sign of brain hemiatrophy in patient with Localized Scleroderma and Parry–Romberg Syndrome

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ABSTRACT

Neurologic involvement is well recognized in Systemic Scleroderma and increasingly reported in Localized Scleroderma. MRI brain abnormalities are often associated with symptoms such as seizures or headaches. In some cases they may be clinically silent. We describe a 23 years old female with head, trunk and limbs scleroderma who developed Parry–Romberg Syndrome. Brain MRI showed ipsilateral temporal lobe atrophy without any prominent neurologic symptoms. Neuropsychological examination revealed Mild Cognitive Impairment. During the 7 years of follow up we have noticed progression of face atrophy but no progression of brain atrophy. Cognitive functions have been stable. This case highlight that major MRI brain abnormalities in LS may occur with only subtle clinical manifestation such as Mild Cognitive Impairment.

1. Introduction

Localized Scleroderma (LS) is a group of autoimmune disorders that affect skin, subcutaneous tissue and sometimes underlying bone. In contrast to Systemic Scleroderma, internal organ involvement is limited [1]. Many authors consider progressive hemifacial atrophy called Parry–Romberg Syndrome (PRS) as a subtype of LS [2].

Neurologic involvement is well recognized in Systemic Scleroderma. It is also one of the most common systemic manifestations in LS [1], especially in subtypes that affect head such as linear scleroderma "en coup de sabre" (LScs) or Parry–Romberg Syndrome [1,3]. MRI brain abnormalities are often associated with symptoms including seizures, headaches, and clear cognitive impairment. Less is known about association with Mild Cognitive Impairment.

We described a female with LS involving head, trunk and limbs, who developed PRS. Patient had ipsilateral focal brain atrophy with no prominent neurological symptoms. However, neuropsychological examination showed Mild Cognitive Impairment.

2. Case report

A 23-year-old Caucasian woman with LS was referred to the Neurology Outpatient Clinic due to asymmetry of the face. At

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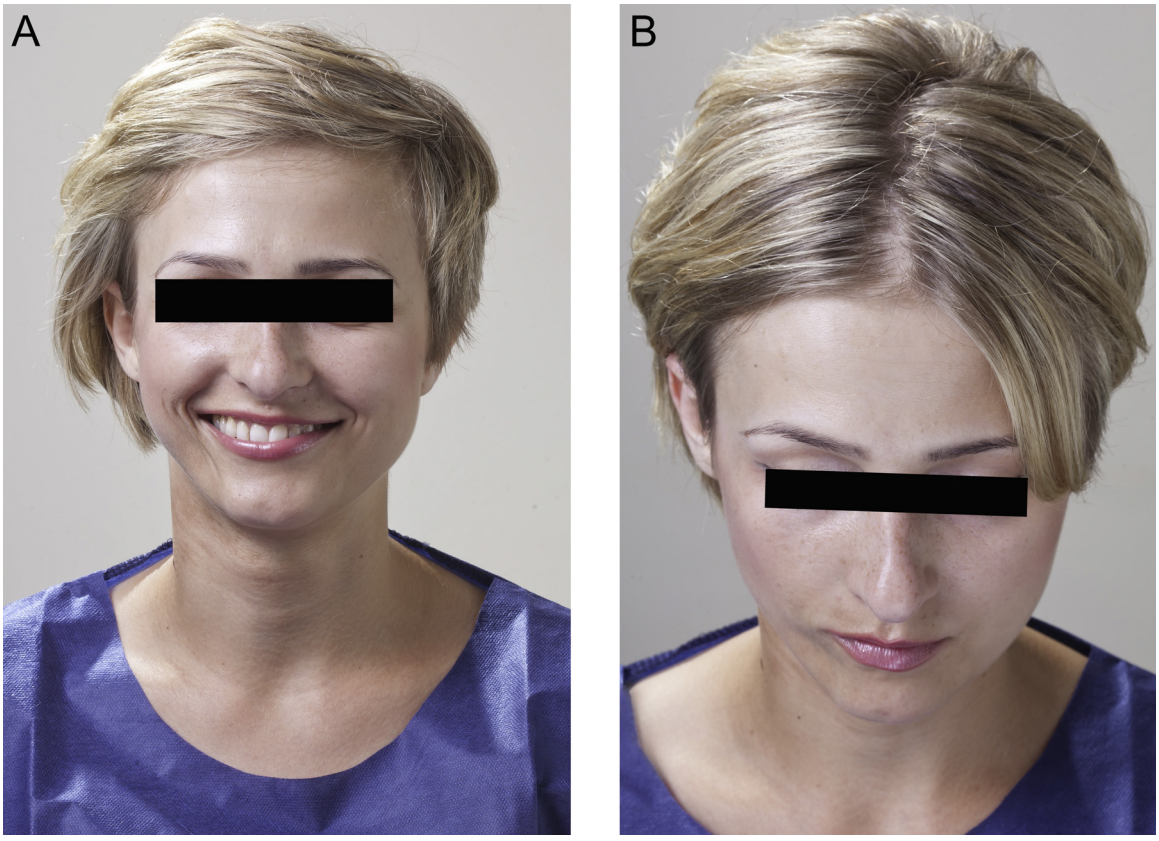

Fig. 1 – Right temporal lobe atrophy. (A) Frontal MRI T1-weighted frontal image and (B) axial MRI FLAIR image shows right temporal lobe atrophic changes.

age of 16 she noticed hyperpigmented and sclerotic skin lesions on the right cheek and neck. Skin biopsy was consisted with morphea (subtype of LS). New skin changes appeared bilaterally on the trunk and limbs. Treatment with piascledine and vitamin E was administered.

Neurologic examination was remarkable only for right hemifacial atrophy and right mouth deviation. Hyperpigmented, sclerotic lesions were noted on the neck and on the body (Fig. 1). Brain MRI showed atrophic changes of the right temporal lobe (Fig. 2) including enlargement of brain sulci,

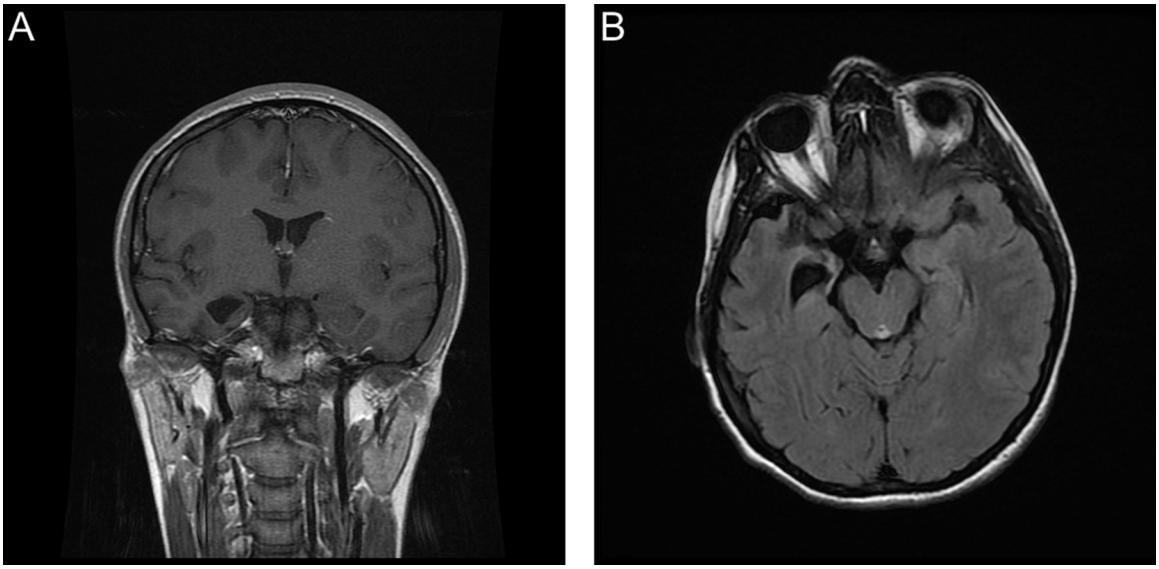

Fig. 2 – Right facial hemiatrophy in follow up. (A and B) Facial hemiatrophy on the right. Patient's mouth deviate toward the affected side and the upper right teeth are crooked (A).