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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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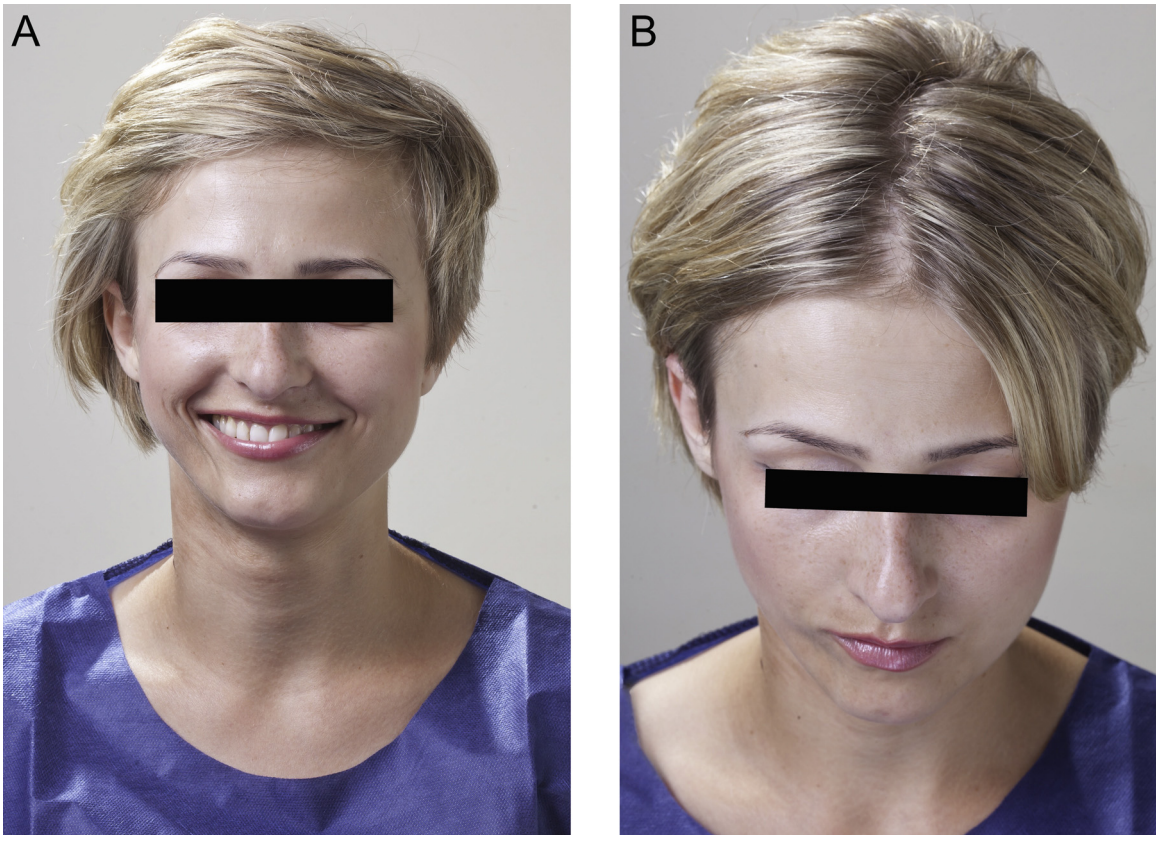

Fig. 1 – Right temporal lobe atrophy. (A) Frontal MRI T1-weighted frontal image and (B) axial MRI FLAIR image shows right temporal lobe atrophic changes.

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,

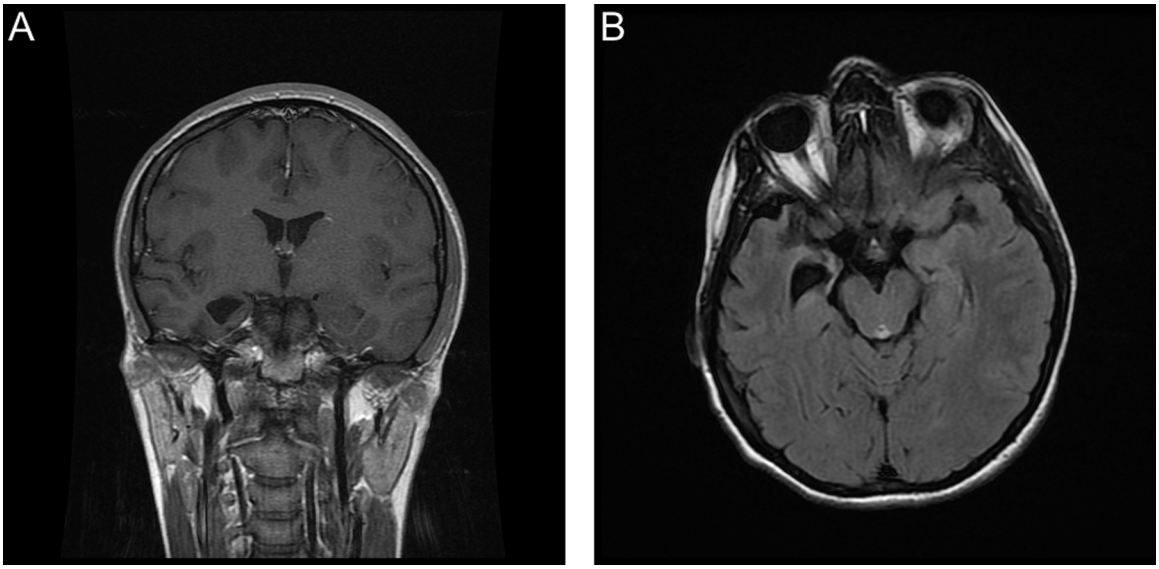

Fig. 2 – Right facial hemiatrophy in follow up. (A and B) Facial hemiatrophy on the right. Patient's mouth deviate toward the affected side and the upper right teeth are crooked (A).

right Sylvian fissure and temporal horn of right lateral ventricle. There was no signal change noticed on T2 or FLAIR sequences and no signs of underlying bone involvement. MR spectroscopy revealed evidence of demyelination – elevated level of Choline (Cho/Cr), Myo-inositol (MI/Cr), Lactic acid (Lac/Cr), and Lipid (Lip/Cr).

Neuropsychological examination showed moderate impairment of attention and concentration, low learning abilities and decline in memory test. The multi domains Mild Cognitive Impairment was recognized.

In blood sample we detected Anti-U1RNP antibodies. Rheumatoid factor, anti-Scl70, anti-dsDNA, anti-Jo1, anti-SM, anti-SS-A, anti-SS-B antibodies were absent. High resolution CT of the lung was normal and there was no other internal organ involvement.

We made a diagnosis of Localized Scleroderma and Parry–Romberg Syndrome with CNS involvement. Treatment with oral (prednisone 50 mg) and topical (clobetasol propionate) corticosteroids was started.

During the next 7 years the area of alopecia appeared on patient's forehead and new sclerotic lesions on the body. Facial hemiatrophy progressed. Methotrexate was added to the therapy but after 2 months patient refused such treatment. Three subsequent MRI performed between 2 and 5 years after diagnosis of PRS showed no progression of temporal lobe atrophy. Patient had no neurologic symptoms and did not notice any worsening in cognitive functioning. Follow-up neuropsychological examinations showed slight improvement in attention level and memory processes.

3. Discussion

Parry–Romberg Syndrome is a rare disorder characterized by atrophy of skin, subcutaneous tissue and sometimes bone on the one side of the face [2]. Neurologic involvement is one of the most frequent systemic manifestations of the disease. PRS coexists with other LS subtypes, especially with LScs in up to 53.6% patients [4].

Differential diagnosis between PRS and LScs is often challenging and some authors consider them to be different spectrum of the same disease [2]. PRS usually affects entire side of the face whereas LScs is limited to frontoparietal area and do not extend below the eyebrow [2,5]. Skin involvement in LScs includes hard, depressed and hyperpigmented sclerotic lesions with progressive softening over time. Atrophy of skin and deep underlying tissue is typical for PRS, but it can be also present in LScs [5].

MRI findings and neurologic symptoms are similar in PRS and LScs but are more frequent in the former [2,6]. Patients usually present with epilepsy, headaches and less frequently pyramidal signs, vision involvement, cranial nerve palsy, cognitive deterioration and behavioral disorders [1,2].

MRI findings in Localized Scleroderma include calcifications, white matter lesions and mostly ipsilateral brain atrophy [7,8]. Blaszczyk et al. compared brain involvement in PRS and LScs using MRI, angio-MRI and 99mTc-HM-PAO-SPECT. Brain abnormalities were similar in both diseases [9]. Lesions may progress, remit and relapse, or reach the stable phase. Generally MRI changes do not correlate with clinical symptoms [7]. Even prominent white matter lesions [7,10] or CNS tumor [11] can be clinically silent. Sakai et al. presented 47-year-old woman with LScs, who despite extensive white matter lesions had no neurologic symptoms [12]. They suggested that this might be explained by interstitial edema, which cause radiologic abnormalities without changes in myelin, neurons, or glial cells.

Brain atrophy of different severity was reported in PRS and other LS types [6,7,13–15]. It can affect the whole hemisphere or manifest as focal abnormal gyral pattern and blurring of gray-white matter. Brain atrophy is sometimes associated with clear cognitive deterioration [13,15] but less is known about Mild Cognitive Impairment. MRI changes associated with Mild Cognitive Impairment may be under-recognized in LS due to publication bias of severe cases. Also, investigation is often postponed until neurologic symptoms or signs of PRS or LScs occur and it rarely includes neuropsychological examination. Our patient had multi domains Mild Cognitive Impairment that improved in the follow-up. When the patient was seen first time, she was very concerned and worried about her medical condition. At that time she was also in a very stressful situation starting her business, working a lot, sleeping little. This situation might negatively influence her cognitive functioning.

When the follow-up neuropsychological examination was done, we noticed improvement, which can be explain by the fact that her business was doing well, she hired people working for her, therefore she had more time to relax. Also her mood improved. When seen first time she expressed general worries about her future, when seen second time she was satisfied and relaxed.

Since that time she does not complain about any change in cognitive functioning.

MR brain spectroscopy of our patient showed metabolic changes consistent with demyelination. In the absence of pathologic confirmation it is difficult to clarify their etiology. Few patients with LS had brain biopsy and they showed evidences of inflammation and vascular dysgenesis [16,17].

There are no recommendations which patients with LS should undergo brain imaging and detailed neurological and neuropsychological investigation. Our patient had anti-U1RNP antibodies. Hietarinta et al. studied different types of antibodies in SS patients and their association with neurologic findings. They suggested that subpopulation of SS patient with anti-U1RNP and those with anti-Scl70 antibodies are more prone to neurologic involvement [18]. Anti-U1RNP were also reported in 3% of LS patients but their associations with neurologic involvement was not investigated [19].

This case shows that the brain atrophy in LS can manifest as Mild Cognitive Impairment without other symptoms. Thus, detailed neurological investigation with brain imaging should be considered in patients with LS despite no prominent neurologic symptoms.

Conflict of interest

None declared.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

Contributors

KE wrote the manuscript, KMA was the treating physician of the patient and supervised writing of the manuscript. Authors reviewed and approved the final version of the manuscript.

Patient consent

A written informed consent was obtained from the patient.

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