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(CASE REPORT)

IPSILATERAL INFLAMMATORY WHITE MATTER LESIONS WITH HEMORRHAGIC FOCI IN LINEAR SCLERODERMA EN COUP DE SABRE: BRAIN MRI FINDINGS WITHIN THE PARRY–ROMBERG SPECTRUM

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ABSTRACT

Linear scleroderma en coup de sabre is a rare subtype of localized scleroderma that overlaps with Parry–Romberg syndrome. We report the case of a 35-year-old woman with long-standing linear scleroderma en coup de sabre who underwent brain MRI to evaluate possible intracranial involvement despite the absence of neurological symptoms.

MRI revealed ipsilateral white matter hyperintensities with thin peripheral contrast enhancement, suggestive of active inflammatory changes, associated with deep hemorrhagic foci on T2*-weighted imaging involving the left internal capsule and thalamic region. These findings were consistent with subclinical intracranial involvement within the Parry–Romberg spectrum.

This case emphasizes the importance of brain MRI in detecting and characterizing inflammatory and hemorrhagic brain abnormalities in patients with craniofacial linear scleroderma, even in the absence of neurological symptoms.

Keywords: Linear Scleroderma En Coup De Sabre, Parry–Romberg Syndrome, Brain MRI, White Matter Lesions, Susceptibility-Weighted Imaging, Subclinical Central Nervous System Involvement

1 INTRODUCTION

Linear scleroderma en coup de sabre is characterized by a linear atrophic lesion affecting the frontoparietal scalp and forehead. It is increasingly considered part of the same clinicopathological spectrum as Parry–Romberg syndrome, also known as progressive hemifacial atrophy (1).

Neurological involvement has been reported in a substantial proportion of patients and may present with seizures, headaches, focal neurological deficits, or may remain entirely subclinical (2). Brain MRI is the modality of choice for evaluating intracranial manifestations and may reveal a broad range of abnormalities, most commonly ipsilateral white matter lesions, cerebral atrophy, calcifications, and enhancement abnormalities (3).

We report a case of linear scleroderma en coup de sabre with ipsilateral inflammatory white matter lesions and deep hemorrhagic foci detected on brain MRI in an asymptomatic patient.

2 CASE PRESENTATION

A 35-year-old woman presented with linear scleroderma en coup de sabre that had begun at the age of 18 years with a pigmented lesion of the forehead and had progressively evolved with cutaneous atrophy and alopecic plaques involving the scalp. She had previously been treated with oral corticosteroids and topical agents. Because of ongoing cutaneous progression, treatment was escalated to methotrexate at a dose of 15 mg weekly, combined with repeated pulses of intravenous methylprednisolone (1 g daily for 3 days every two months). She reported no neurological symptoms. Dermatological examination revealed a 5 cm linear atrophic and pigmented plaque on the left hemi-forehead extending toward the scalp, associated with two sclero-atrophic alopecic plaques. Neurological examination was normal.

Given the progressive craniofacial disease and the known possibility of subclinical neurological involvement within the Parry–Romberg spectrum, brain MRI was performed to assess possible intracranial abnormalities. Brain MRI demonstrated multiple hyperintense lesions on T2-weighted and FLAIR sequences involving the subcortical and periventricular white matter of the left cerebral hemisphere, strictly ipsilateral to the cutaneous lesion. These lesions showed thin peripheral contrast enhancement after gadolinium administration, suggestive of active inflammatory involvement. The linear subcutaneous plaque in the left frontal region was also well visualized, corresponding to the clinically evident cutaneous lesion. Susceptibility-sensitive T2*-weighted images revealed focal hypointense foci in the left internal capsule and adjacent thalamic region, consistent with hemorrhagic changes. Diffusion-weighted imaging showed no restriction. No significant mass effect, cortical atrophy, or leptomeningeal enhancement was identified. MR angiography showed no vascular abnormality.

These findings were consistent with ipsilateral inflammatory white matter lesions and deep hemorrhagic foci associated with linear scleroderma en coup de sabre.

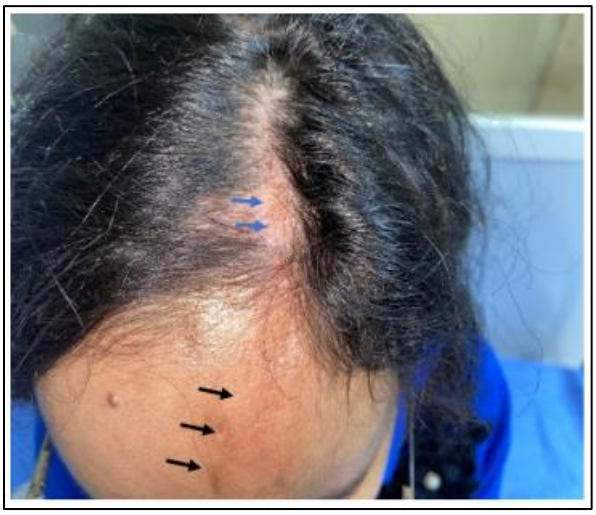

Figure 1: Clinical photograph showing a linear atrophic pigmented lesion on the left frontal scalp (black arrow) with associated alopecic plaques (blue arrow), consistent with en coup de sabre morphea.

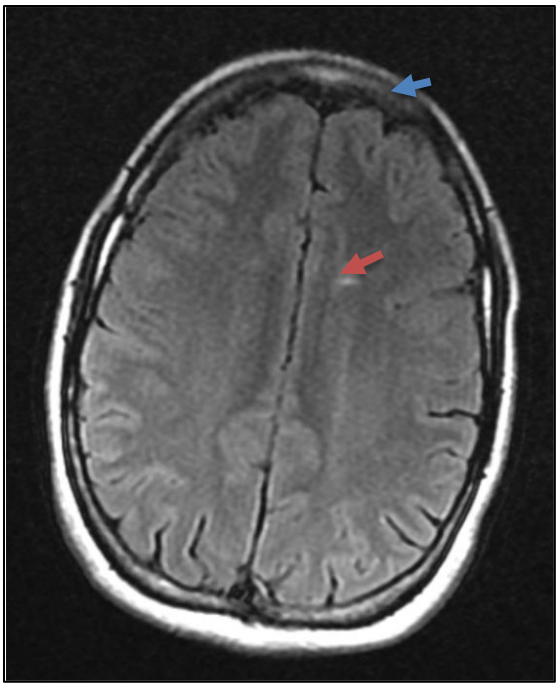

Figure 2: Axial FLAIR MRI showing hyperintense lesions in the left periventricular and subcortical white matter (orange arrow) with visualization of the linear subcutaneous plaque (blue arrow).

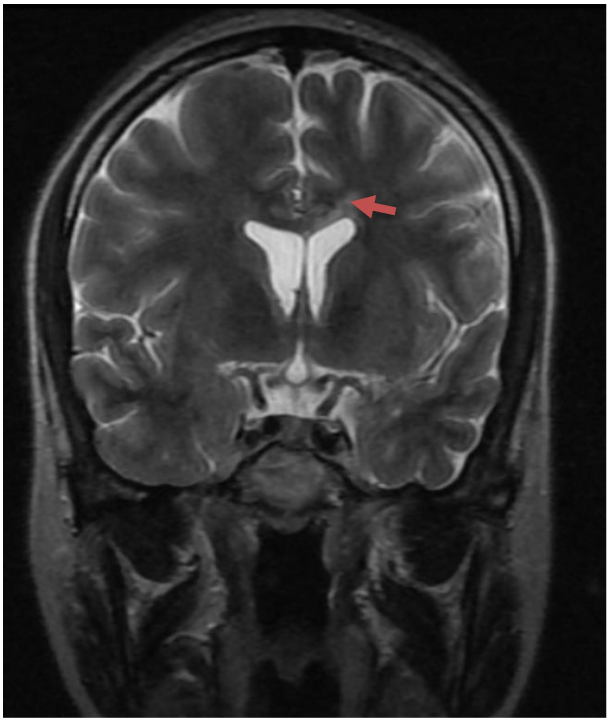

Figure 3: Coronal T2-weighted MRI confirming hyperintense white matter lesions in the left cerebral hemisphere (orange arrow).

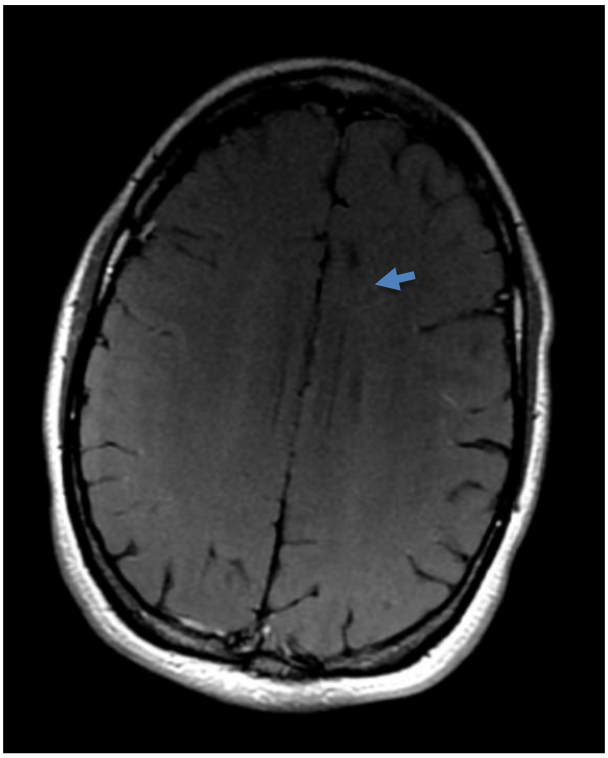

Figure 4: Axial contrast-enhanced T1-weighted image demonstrating thin peripheral enhancement of the white matter lesions (blue arrow).

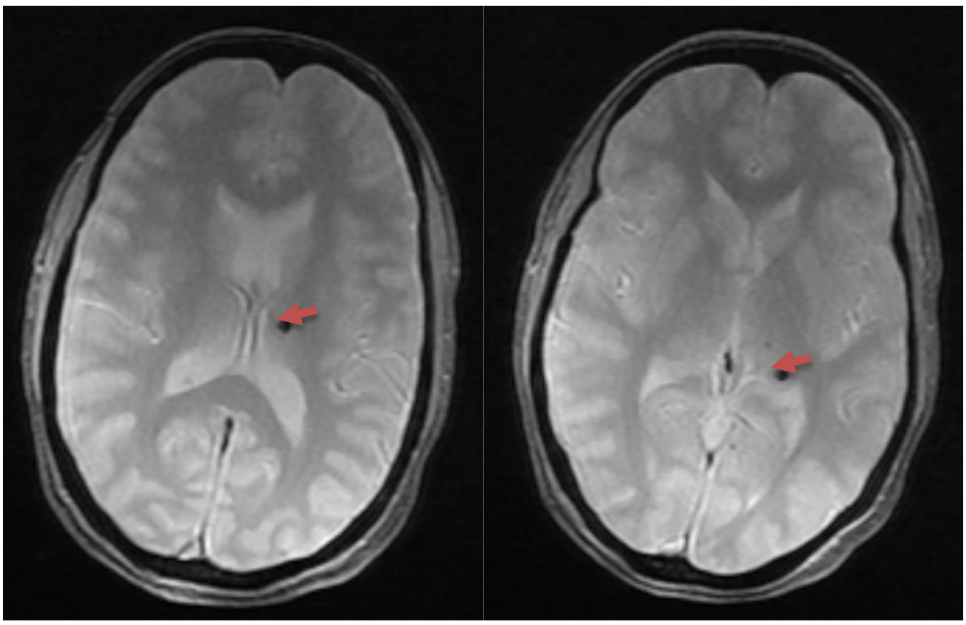

Figure 5: Axial T2*-weighted image showing hypointense foci in the left internal capsule and thalamic region (orange arrow), consistent with hemorrhagic changes.

3 DISCUSSION

Linear scleroderma en coup de sabre (ECDS) and Parry–Romberg syndrome are increasingly recognized as part of the same clinicopathological spectrum (1). Neuroimaging abnormalities are common and typically ipsilateral to the cutaneous lesion.

The most frequently reported MRI findings include T2/FLAIR white matter hyperintensities, cerebral atrophy, calcifications, and leptomeningeal or parenchymal enhancement (3). These changes can be detected even in

asymptomatic patients, highlighting the value of neuroimaging in identifying subclinical central nervous system involvement (2).

Linear scleroderma en coup de sabre is a rare subtype of localized scleroderma with a clear female predominance. Although it most often begins in childhood, adult-onset forms are well documented, as in our 35-year-old patient whose disease started at age 18. Neurological involvement, ranging from seizures and headaches to focal deficits or purely subclinical MRI abnormalities, occurs in a significant proportion of patients (4).

Regarding treatment, the combination of methotrexate (15–25 mg weekly) and systemic corticosteroids remains the first-line therapy for active craniofacial disease, aiming to halt inflammatory progression and prevent further atrophy or neurological complications. In our case, this regimen effectively controlled the ongoing cutaneous progression, while brain MRI revealed subclinical inflammatory and hemorrhagic lesions. For refractory cases, additional immunosuppressants or biologics may be considered. A multidisciplinary approach involving dermatology, neurology, and radiology with serial MRI monitoring is recommended. In the stable phase, reconstructive procedures such as autologous fat grafting are frequently performed to address residual facial atrophy (8).

In the present case, brain MRI demonstrated multiple ipsilateral subcortical and periventricular white matter hyperintensities on T2-weighted and FLAIR sequences with thin peripheral contrast enhancement, suggestive of active inflammation. In addition, T2*-weighted imaging showed deep hypointense foci in the left internal capsule and thalamic region, consistent with hemorrhagic changes. These findings were observed despite a completely normal neurological examination.

Although inflammatory white matter lesions are relatively well recognized in ECDS, associated hemorrhagic manifestations remain uncommon and are less frequently highlighted in the literature (5). The deep T2* hypointense foci in our patient likely represent hemosiderin deposition secondary to an underlying inflammatory microangiopathy or small-vessel vasculitic process (6).

The strict ipsilateral distribution of both the cutaneous and intracranial abnormalities supports a localized immune-mediated process affecting the skin and underlying brain parenchyma. The presence of contrast enhancement argues in favor of ongoing inflammatory activity rather than purely chronic gliotic changes.

This case emphasizes the important role of brain MRI in patients with craniofacial linear scleroderma. Even in the absence of neurological symptoms, MRI can detect and characterize both inflammatory and hemorrhagic intracranial lesions, which may have implications for therapeutic monitoring and long-term follow-up (7).

4 CONCLUSION

Linear scleroderma en coup de sabre can be associated with ipsilateral inflammatory white matter lesions and hemorrhagic foci on MRI, even in asymptomatic patients. Brain MRI should be considered in patients with craniofacial linear scleroderma in order to evaluate possible subclinical neurological involvement within the Parry–Romberg spectrum.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Disclosure of conflict of interest

The authors declare no conflicts of interest regarding this manuscript.

Statement of informed consent

Informed consent was obtained from the legal guardians of the patient included in this study.

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