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Linear Scleroderma "En Coup de Sabre" with Neurological Manifestations and Neurovascular Abnormalities: A Case Report

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1. Abstract

Localized scleroderma (LoS) is distinct from systemic sclerosis due to the lack of visceral involvement: among its subtypes, linear scleroderma "en coup de sabre" (LScs) has been associated with various neurological complications, including epilepsy, intracerebral lesions, and ophthalmologic symptoms. This article presents a rare and complex case of linear scleroderma associated with plaque-type morphea and concomitant neurovascular involvement in a 59-year-old female, emphasizing the radiological, neurological, and dermatological features.

2. Introduction

Localized scleroderma (LoS), also known as morphea, encompasses linear scleroderma, plaque-type morphea, and generalized morphea1. Unlike systemic sclerosis, LoS typically lacks esophageal, pulmonary, and vascular involvement. Linear scleroderma "en coup de sabre" (LScs) is characterized by a linear, atrophic, and sclerotic band typically affecting the frontoparietal region of the forehead and scalp: the term "en coup de sabre", French for "strike of the sword", describes the lesion's resemblance to a sabre wound [2]. This condition predominantly manifests in children and adolescents, with a higher incidence in females. The etiology of LScs remains unclear, though it is believed to involve autoimmune mechanisms leading to endothelial cell injury and the activation of pro-fibrotic pathways [3,4]. Clinically, patients present with a linear, indurated plaque that progresses to a depressed, sclerotic scar: the lesion often extends from the forehead to the scalp, resulting in alopecia and potential atrophy of underlying structures, including muscle and bone [2,5,6]. Neurological manifestations in LScs most commonly include complex partial seizures, followed by generalized tonic-clonic seizures; headache is also frequently reported. Additional findings include cranial nerve involvement and ophthalmologic abnormalities, pyramidal signs and psychiatric disturbances [7,8,9]. Radiological findings typically reveal cerebral calcifications, cortical atrophy, extracranial atrophy, and associated cortical depression [10,11]. The presence of cerebral aneurysms in association with LScs is exceedingly rare, with only a few cases documented in the literature [12]. The pathophysiological mechanisms underlying this association remain unclear; however, it is hypothesized that chronic inflammatory and fibrotic processes affecting the vascular endothelium may contribute to vessel wall fragility and aneurysm formation [12]. Here, we describe a case of LScs complicated by epilepsy, cerebral aneurysms, and coexisting plaque morphea.

3. Case Presentation

We present the case of a 59-year-old female patient, A. M., affected by focal scleroderma with neurological manifestations. Born at full term via dystocic delivery, with perinatal hypoxic distress due to an umbilical cord loop, A. M. exhibited mild cognitive impairment from birth, characterized by moderate ideomotor slowing and a fixation memory deficit. At the age of 25, she experienced her first tonic-clonic seizure and was started on phenobarbital 100 mg, with partial benefit. Subsequently, she developed multiple daily absence seizures. EEG recordings revealed a disrupted background rhythm, right hemispheric slowing, and occasional sharp theta waves. A CT scan showed right hemisphere atrophy with secondary enlargement of the lateral ventricle. Additionally, dysmorphia of the right greater wing of the sphenoid bone was observed, along with an arachnoid cyst extending to the pterygoid processes. Angiographic studies revealed a saccular aneurysm of the anterior communicating artery [Figure 1a-1b] giving rise to both anterior cerebral arter-

ies, focal ectasia of the M2 segment of the right middle cerebral artery involving the sphenoid malformation, hypoplasia of the right carotid siphon [Figure 1c], and reduced representation of the right intracranial circulation. A right-sided dural angiomatosis was also identified, corresponding to a triangular area of hypoplastic frontal skin tissue. Antiepileptic therapy was progressively adjusted to 150 mg of phenobarbital, achieving reasonable control over tonic-clonic seizures but less control over absence seizures. Concurrently, A. M. developed a psychiatric disorder characterized by obsessive-compulsive disorder and anorexia nervosa, for which she was started on escitalopram 10 mg. Subsequent brain MRI [Figures 2a-2b-2c-2d-2e] confirmed right hemispheric atrophy. A cerebral panangiography revealed reduced right intracranial circulation, aplasia of the right A1 segment of the anterior cerebral artery, and thrombosis of the right M2 segment of the middle cerebral artery. At the age of 53, following sudden onset of left hemifacial hyposthenia and hypoesthesia, she presented to the Emergency Department. An angio-CT scan revealed a newly thrombosed aneurysm in the M2 segment of the right middle cerebral artery. At 55 years of age, she experienced worsening psychiatric symptoms, including somatization of generalized anxiety disorder, with psychogenic non-epileptic seizures (PNES) mimicking the tonic-clonic seizures she had previously developed. Due to an increase in the frequency of tonic-clonic seizures, she underwent a new neurological evaluation and was started on lamotrigine, gradually titrated to 300 mg/day, achieving good control over both tonic-clonic and absence seizures. During clinical examination, attention was drawn to right frontal dermal hypoplasia; a second hypoplastic area was identified in the lumbar region, which was not associated with any malformation on MRI: a dermatological visit was requested. On 30 January 2025 the patient arrived in our rare diseases' clinic in Policlinico Umberto I for dermatologic evaluation: A.M. presented with a linear, atrophic, and hypopigmented lesion extending along the right frontoparietal region of the face and scalp, exhibiting a characteristic "en coup de sabre" appearance, consistent with linear morphea: the affected area showed marked dermal hypoplasia, with thinning of the overlying skin and increased visibility of the underlying vasculature [Figures 3a]. There was aFare clic o toccare qui per immettere il testo. lso evident cicatricial alopecia involving the affected scalp [Figures 3b]. On the dorsal aspect of the trunk, a well-demarcated, depressed plaque was also noted, measuring approximately 10x7cm with a smooth, atrophic surface and hyperpigmented appearance with hypertrichosis [Figures 3c]. Focal scleroderma with neurological involvement, particularly in LScs, is associated with a broad spectrum of neurological manifestations, including seizures, focal deficits, headaches, and stroke-like episodes. Radiologically, it is characterized by cortical and subcortical atrophy, gliosis and diminished cerebral perfusion11. Rarely, it can also be associated with the presence of intracranial aneurysms. This case illustrates the complexity of diagnosing and managing focal scleroderma with neurological sequelae. The persistent right hemispheric atrophy, congenital vascular anomalies, and associated psychiatric symptoms underscore the necessity for multidisciplinary management [10].

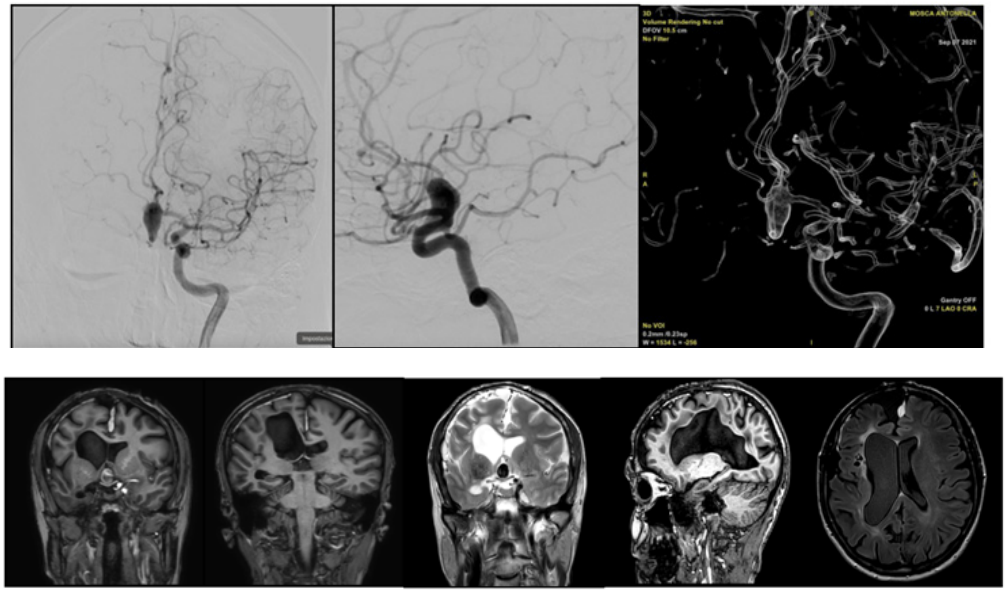

Figure 1a 1b: Left carotid angiography highlighting an anterior communicating artery (ACoA) aneurysm. Figure 1c. The angiographic study (3D rotational angiographic acquisition) revealed a hypoplastic right internal carotid artery, with a post-bulbar indentation resembling a WEB-like morphology. At the intracranial level, the right middle cerebral artery exhibited an occlusion of the proximal third, with a "cul-de-sac" appearance, consistent with spontaneous thrombosis of a large Sylvian aneurysmal sac. Furthermore, there was evidence of aplasia of the A1 segment of the right anterior cerebral artery.

Figure 2a: Coronal T1 section showing atrophy and ex vacuo dilation of the right lateral ventricle	Figure 2b: Coronal T1 section showing atrophy and ex vacuo dilation of the right lateral ventricle.	Figure 2c: Coronal T2 section highlighting right cortico-subcortical atrophy.	Figure 2d: Sagittal T1 section highlighting ex vacuo dilation and cortical atrophy	Figure 2e: Transverse FLAIR section highlighting atrophy and right-lateralized gliosis

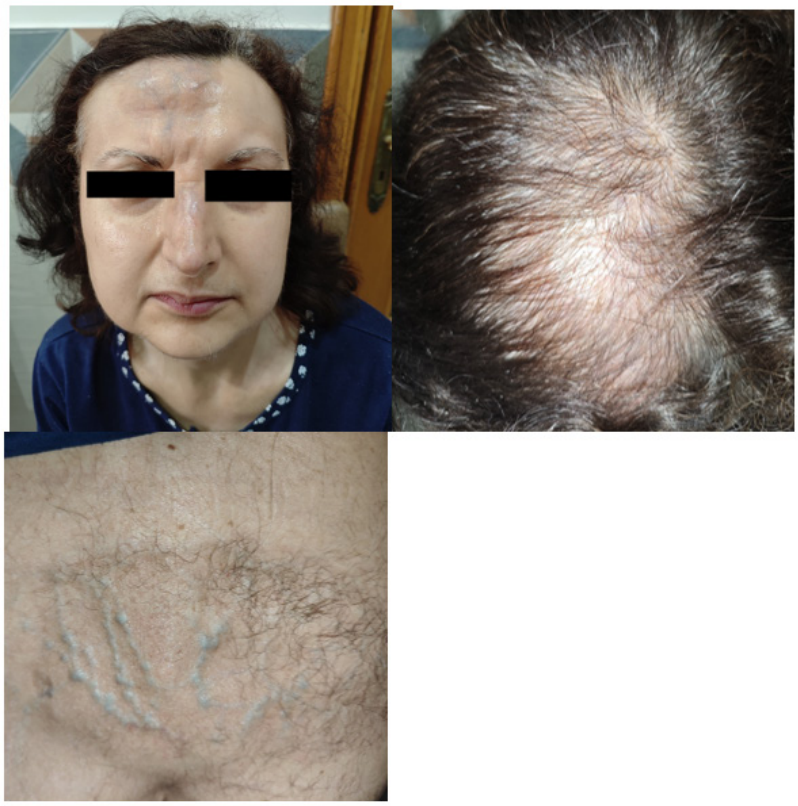

Figure 3a: Linear and atrophic “en coupe de sabre” lesion involving the right fronto-parietal region. Of note, skin thinning leads to visualize underlying vessels. Fiugre 3b. Cicatritial alopecia of the central-right side of the scalp with large reduction of active follicular units.
Figure 3c: Plaque-like morphea appearing as atrophic and hyperpigmented lesion on the back, associated with hypertrichosis.

4. Conclusion

This case highlights the need for heightened awareness of neurological and psychiatric involvement in localized scleroderma. Early neuroimaging, EEG monitoring, and psychiatric evaluation are essential components of care in such patients. Ongoing follow-up and tailored therapy can lead to improved outcomes despite the chronicity and complexity of the condition. To the best of our knowledge, this is the first documented case describing the coexistence of linear scleroderma “en coup de sabre”, plaque morphea, and cerebral aneurysms within the same patient other than the classical neurological complications frequently associated with LScs. Such a unique association may provide valuable insight into the pathophysiologic mechanisms linking cutaneous abnormalities to neurovascular and neurological dysfunction.

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