Multiple Sclerosis and Related Disorders 8 (2016) 96–98

Contents lists available at ScienceDirect

Multiple Sclerosis and Related Disorders

journal homepage: www.elsevier.com/locate/msard

Case report

Localized scleroderma en coup de sabre in the Neurology Clinic

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ARTICLE INFO

Article history:
Received 14 March 2016
Received in revised form
11 May 2016
Accepted 19 May 2016

Keywords:
Scleroderma, Localized
Scleroderma en coup de sabre
Neurological manifestations
Magnetic resonance imaging

ABSTRACT

Background: Localized scleroderma en coup de sabre (LScs) is a form of localized scleroderma thought to be an autoimmune disorder. Central nervous system involvement is not rare and neurological manifestations include seizures, focal neurological deficits, headache and neuropsychiatric changes.
Methods: Patients attending the Neurology Clinic with the final diagnosis of LScs with neurological manifestations were identified and clinical and imagiological records reviewed.
Results: Five patients (0.024%) had LScs with neurological involvement, presenting with transient focal neurologic deficits, seizures, headache or migraine with aura. Neuroimaging studies confirmed localized skin depression and showed bone thinning, white matter lesions, brain calcifications, sulcal effacement and meningeal enhancement. Three patients experienced clinical improvement after immunosuppressive therapy, and in two of these patients neuroimaging findings also improved.
Conclusions: Recognizing typical dermatologic changes is keystone for the diagnosis of LScs with neurological involvement. It is a diagnosis of exclusion and extensive etiological diagnostic evaluation should be performed. Treatment options, including conservative follow-up or immunosuppressive therapy, should be carefully considered.

1. Introduction

Localized scleroderma is a rare chronic disorder of unknown etiology, which presents with typical skin and underlying tissue sclerosis, classically considered not to present extra-cutaneous involvement (Takehara and Sato, 2005). Localized scleroderma en coup de sabre (LScs) is a form of localized scleroderma, involving the fronto-parietal area, with band-like sclerotic lesions with atrophy, furrow of the skin and outer diploe thinning (Unterberger et al., 2003). Recent reports confirm that central nervous system involvement is not rare, and presentations include epileptic seizures, headache, focal neurological deficits and neuropsychiatric changes (Appenzeller et al., 2004; Amaral et al., 2013).

2. Material and methods

During the period 2008–2013, 20,531 patients attended our Neurology Clinic for a first consultation. Diagnostic electronic database was used to identify patients with the diagnosis of LScs and 5 patients with LSsc with neurological involvement were found (0.024%). Clinical and imagiologic records were reviewed for data collection. Details that might disclose the identity of the patients were omitted. The study is in accordance to the local ethics committee requirements for clinical research.

3. Results

We present a brief case description of the 5 patients with LSsc with neurological involvement, and dermatological and neuroimaging findings are illustrated in Fig. 1.

3.1. Case 1

60-Year old man with sudden-onset aphasia and right hemiparesis lasting for 15–20 min. He reported 3 similar episodes in the previous 6 years. He had a localized atrophic skin area in the left fronto-parietal area that had been unchanged for several years. Brain magnetic resonance imaging (MRI) confirmed subcutaneous

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http://dx.doi.org/10.1016/j.msard.2016.05.013
2211-0348/

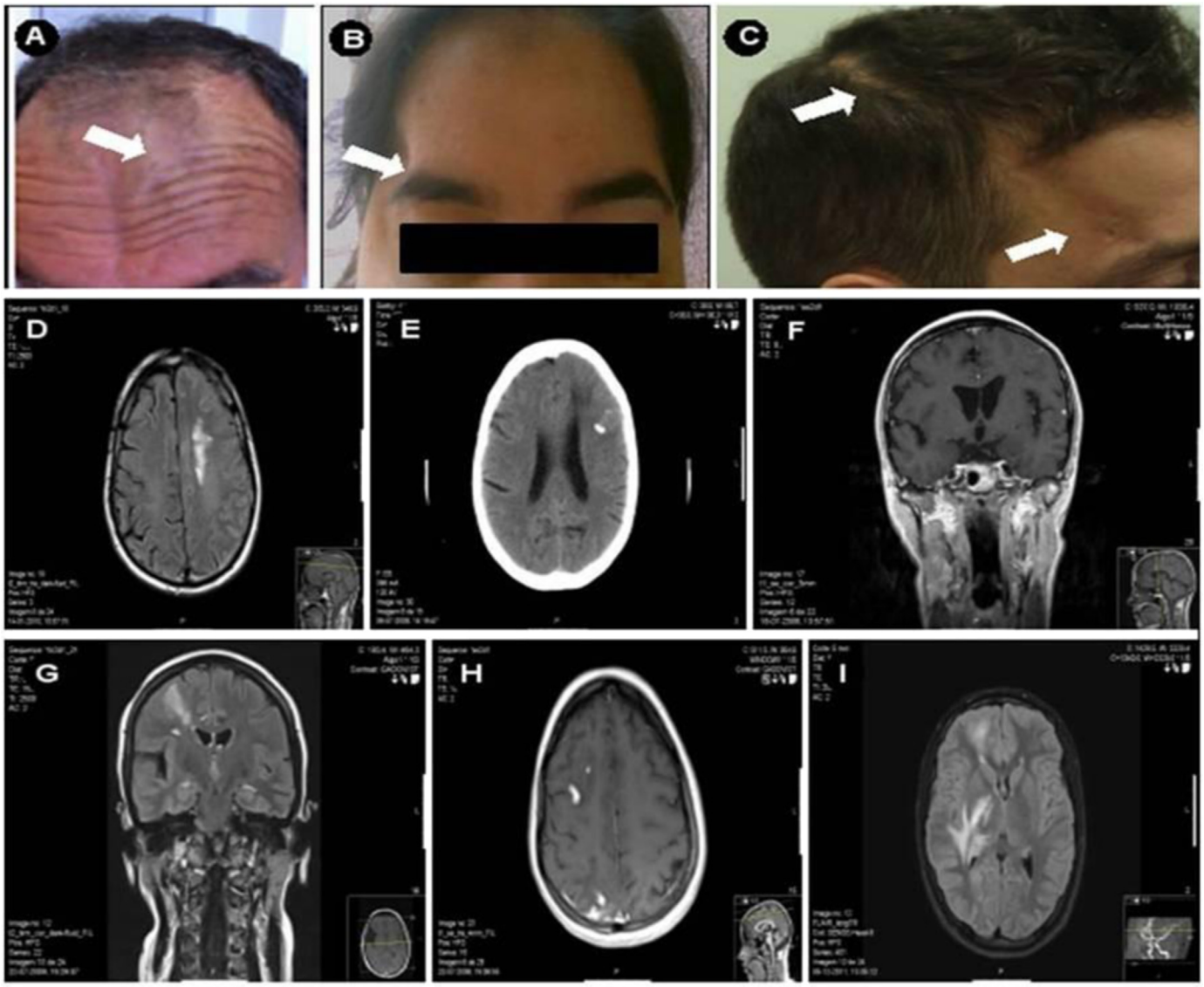

Fig. 1. A–C: dermatological changes of scleroderma en coup de sabre, with focal skin atrophy and hyperpigmentation and underlying bone depression (arrows; case 1, 3 and 5 respectively). D: Case 1, FLAIR image shows left hemispheric sulcal effacement and a hyperintense white matter lesion. E–F: Case 2, CT-scan (E) reveals left frontal sulci effacement and a left subcortical hypodense lesion with calcifications, and contrast-enhanced T1(F) demonstrates left frontal skin and bone atrophy, with underlying dural enhancement and sulci effacement. G–H: Case 3, FLAIR (G) and contrast-enhanced T1 (H) depict hyperintense, contrast-enhancing white matter lesions underlying right frontal skin and bone atrophy. I: Case 4, FLAIR shows hyperintense right hemisphere lesions involving internal and external capsules, peritrigonal region, thalamus and subcortical temporal and frontal white matter.

tissue and bone atrophy and showed left hemispheric sulcal effacement with a subcortical T2-hyperintense lesion and dural thickening with contrast enhancement. Vascular investigation, video-electroencephalogram (EEG) and cerebrospinal fluid (CSF) were normal with negative oligoclonal bands. He was treated with prednisolone but during steroid tapering a similar episode recurred, and methotrexate was started. He has been asymptomatic for 4 years and MRI remains unchanged, maintaining dural thickening with contrast enhancement.

3.2. Case 2

46 year-old woman with 1-month history of holocranial headache and paresis of the right upper limb. MRI revealed a partially calcified frontal lesion, with sulci effacement and no contrast enhancement, suggesting a low-grade tumor. Surgical lesionectomy was performed and histology showed multiple calcifications in otherwise normal brain tissue. Persisting headaches prompted a neurological assessment. She reported a gradual development of upper face asymmetry for several years, and skin atrophy on the left frontal area was noticed. MRI was reviewed and muscle and bone atrophy underlying the skin atrophy and left frontal and inter-hemispheric meningeal thickening with contrast enhancement were detected. Skin biopsy revealed lymphocytic infiltrates around hair follicles and accumulation of elastotic material. Symptomatic headache treatment was effective and MRI findings, including gadolinium study, remain unchanged for 5.5 years.

3.3. Case 3

29-Year-old woman, with history of migraine without aura, presented with severe headaches and status epilepticus with left motor seizures caused by right parietal vein thrombosis and cortical haemorrhage. After appropriate treatment, physical examination revealed a right eye adduction limitation which was

present since several years before, and a right fronto-parietal atrophic skin lesion with underlying bone deformity and mild alopecia which appeared at age 14 and progressed during 7 years. Acute phase MRI revealed multiple, bilateral T2 hyperintense enhancing lesions with surrounding edema, and new contrast enhancing lesions appeared 12 days later. Arterial-phase MR angiography (MRA) was normal, as was CSF study, including negative oligoclonal bands. Intravenous followed by oral steroids were started, and during follow-up immunosuppressive maintenance therapy with azathioprine was decided. Ophthalmoparesis persists, migraine frequency improved significantly and the last MRI was but considerably better than the first studies, revealing partial regression of the lesions (5-year follow-up) and gadolinium study revealed only one minor left occipital focus of contrast enhancement.

3.4. Case 4

16 Year-old girl with history of alpha-1 antitrypsin deficiency, admitted for recent episodes consistent with migraine with left visual and sensory aura. Neurological examination was normal. She had a right fronto-parietal atrophic cutaneous lesion with localized alopecia, which developed during the previous 2 years. MRI revealed right hemispheric, confluent, contrast enhancing white matter lesions, calcifications and leptomeningeal thickening. EEG, and cerebral angiography were normal and CSF was normal with negative oligoclonal bands. Skin biopsy only showed mild interstitial fibrosis. After intravenous steroid induction, oral prednisolone was maintained during 10 months. During the 2.5 years of follow-up a single generalized tonic-clonic seizure occurred (treated with levetiracetam) and MRI showed considerable reduction of lesion size and contrast enhancement being reduced to a small right sublenticular focus.

3.5. Case 5

30 Year-old man with history of IgA nephropathy and hypertension, reported a left sensory disturbance with gradual onset during 24 h (first involved the left hand, progressed proximally to the upper limb, left hemiface and ultimately the lower limb) which progressively improved after 1 month. Neurological examination was normal and physical examination showed a linear right frontal lesion extending to the parietal region, with skin atrophy, hyperpigmentation and alopecia, which developed over the period of 2 years when he was 6 years-old. MRI, including contrast, revealed no parenchymal or meningeal abnormality. The patient has been asymptomatic for 2 years and has not performed control MRI.

4. Discussion/conclusions

Localized scleroderma en coup de sabre with neurological involvement is rare and these 5 cases describe the clinical and imagiologic spectrum of its neurological manifestations. All patients developed neurologic symptoms several years (2–24) after appearance of dermatological changes. The most frequent neurological manifestations were focal neurological deficits and headache. Epileptic seizures occurred in two patients and cranial nerve palsy occurred in only one patient. The majority of patients with LScs and neurological symptoms have abnormal neuroimaging, similar to our case series (Appenzeller et al., 2004). Findings included localized skin depression (5/5), outer diploe thinning (4/5), T2 and FLAIR hyperintense white matter lesions (3/5), abnormal gyral pattern (2/5), calcification (2/5), dural thickening with contrast enhancement (2/5) and parenchymal contrast-enhancing lesions (2/5). In all patients extensive additional investigation excluded other causes for the neurological manifestations and CSF studies (performed in 3 patients) were normal as were vascular imaging studies (performed in 2 patients). Cerebral vascular imaging and CSF studies are pertinent in cases of scleroderma with neurologic involvement, since clinical and laboratory data support the hypothesis that vasculature is a primary target. Although rarely has vasculitis or CSF changes been reported in these patients (Amaral et al., 2013). Due to the retrospective nature of this report, the authors did not have the opportunity to be involved in the management of every patient. The investigation and follow-up was tailored by the assistant neurologist, who decided the most appropriate course of action concerning the specific case at hand.

There are no randomized controlled trials evaluating treatment and neurologic involvement in scleroderma is variable. Systemic immunosupressive therapy, namely steroids, methotrexate, azathioprine, mycophenolate mofetil and intravenous immunoglobulin were used in the treatment of LScs with neurological involvement (Amaral et al., 2013, Zulian et al., 2008). Immunosuppressive agents should be carefully considered in these patients, because at least some patients may have a benign course of neurological manifestations, which may improve spontaneously or with symptomatic treatment. Because patient 2 and 5 did not have any new significant symptoms and image was stable or normal, it was decided to have a conservative approach. Chronic immunosuppressant treatment in patients 1, 3 and 4 was the option due to repeated symptoms in patient 1 and ongoing clinical and imagiological activity in patients 3 and 4 (widespread bilateral involvement and new contrast enhancing lesions in patient 3, and lesion volume and mass effect in patient 4).

Disclosure of conflicts of interests

The authors report no conflict of interests.

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