SAGE Open Medical Case Reports

SAGE Open Medical Case Reports
JCMS Case Reports
Volume 9: 1–5

DOI: 10.1177/2050313X21993304
journals.sagepub.com/home/sco
SAGE

Adult-onset en coup de sabre scleroderma in a patient with linear localized scleroderma profunda: A case report and literature review

Roxana Mititelu[1], Zainab Ridha[2], Lydia Ouchene[3] and Elena Netchiporouk[1]

Abstract

The en coup de sabre variant of linear scleroderma typically occurs in children. We report a unique case of adult-onset en coup de sabre scleroderma in a patient with linear localized scleroderma profunda. The patient was treated with oral steroids and oral methotrexate improving her cutaneous disease. This case highlights the importance of a thorough cutaneous examination as this adult patient developed an entity traditionally believed to occur in childhood.

Keywords

Scleroderma, en coup de sabre scleroderma, adult-onset scleroderma, linear scleroderma, morphea, localized scleroderma

Introduction

Localized scleroderma, also known as morphea, is a cutaneous disorder characterized by fibrosis of the skin and less frequently of the underlying tissue. It can be subdivided into linear, circumscribed (plaque), generalized, pansclerotic, and mixed subtypes.[1] Linear scleroderma commonly affects the limbs or the face (en coup de sabre (ECDS) or progressive facial hemiatrophy) and may be associated with extracutaneous complications.[1] ECDS typically occurs in children and is characterized by a linear sclerotic plaque on the face.[1] Here, we present the third case of adult-onset (AO) ECDS linear scleroderma occurring in a patient with pre-existing long-standing linear scleroderma affecting the limb and the trunk.

Case report

A 52-year-old woman of Cuban origin was referred to our clinic for skin hardening for over 15 years. Her past medical history was pertinent for mild asthma and severe glaucoma. On skin examination, there were three ill-defined indurated, depressed plaques measuring 10 cm × 15 cm on the left mid-back, 7 cm × 7 cm on the left posterior shoulder, and 15 cm × 3 cm linear induration extending to the left forearm (Figure 1(a) and (b)). Examination of the oral mucosa and genitalia was unremarkable. Workup, including complete blood count, chemistry, renal and liver function tests, borrelia serology, antinuclear, and systemic sclerosis specific antibodies, was normal, except for mild eosinophilia. A punch biopsy of the left upper back lesion demonstrated a normal epidermis and superficial dermis. There was non-inflammatory extensive sclerosis in the deep dermis, subcutaneous tissue, and fascia.

The diagnosis of linear localized scleroderma profunda versus eosinophilic fasciitis was made. Pre-treatment screening for hepatitis B, hepatitis C, human immunodeficiency virus, QuantiFERON-TB Gold, and strongyloides serology were performed and subsequently revealed the presence of strongyloides antibodies, which were confirmed by fecal culture. The patient was treated with oral ivermectin resulting in a resolution of her respiratory symptoms and normalization of the eosinophil counts.

After successful eradication of strongyloidiasis, we planned to initiate methotrexate monotherapy, which was delayed due to scheduled hysterectomy for uterine fibroids.

[1]Division of Dermatology, McGill University Health Centre, Montreal, QC, Canada
[2]Faculty of Medicine, Université Laval, Quebec City, QC, Canada
[3]Faculty of Medicine, McGill University, Montreal, QC, Canada

Corresponding Author:
Elena Netchiporouk, Division of Dermatology, McGill University Health Centre, Montreal General Hospital, Montreal, QC, Canada, H3G 1A4.
Email: elena.netchiporouk@mail.mcgill.ca

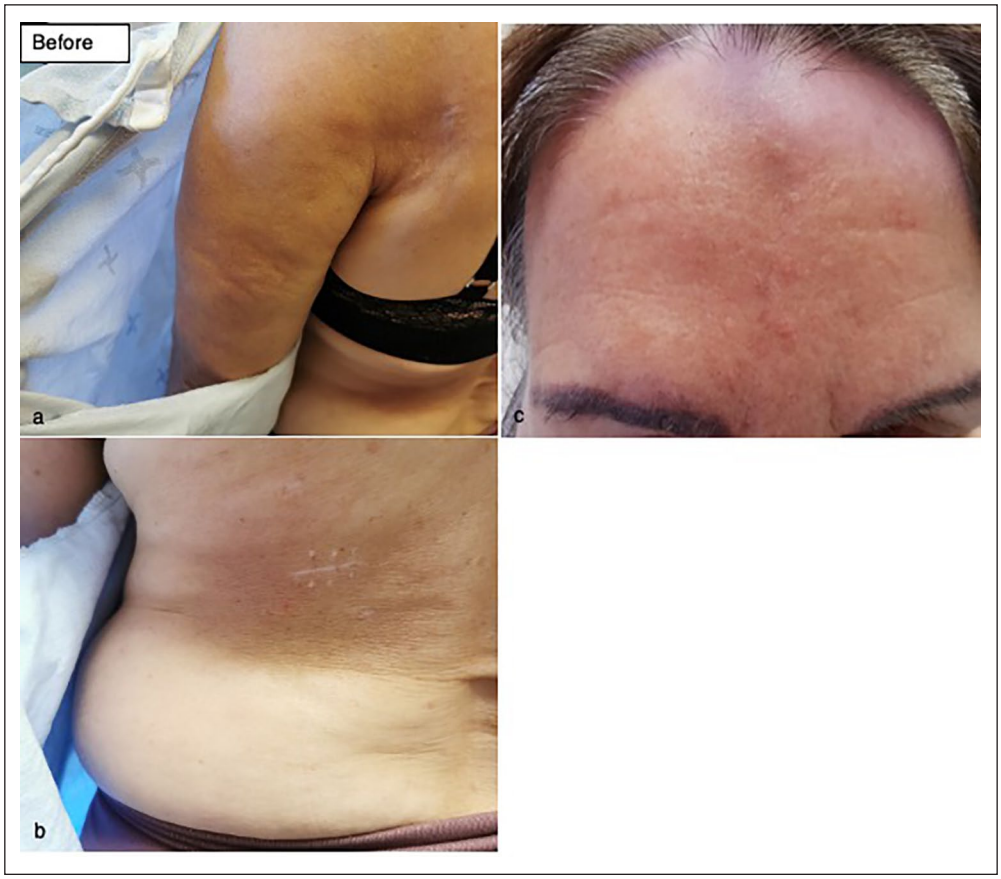

Figure 1. Prior to treatment, indurated depressed plaques were present on the (a) left shoulder and left triceps, (b) left mid-back, and (c) mid-forehead.

When the patient returned to our clinic, a new linear depressed plaque on the mid-forehead was noted (Figure 1(c)). She also complained of worsened vision in the right eye and new-onset migraine-type headache. Given the typical clinical appearance of the forehead lesion, a diagnosis of ECDS scleroderma was made. She was referred to her treating ophthalmologist, and a magnetic resonance imaging (MRI) of the brain was ordered. The MRI showed no skeletal or parenchymal abnormalities. Ocular examination revealed stable glaucoma. Oral prednisone 1 mg/kg/day (tapered over 3 months) and methotrexate 20 mg/week with folic acid and vitamin D supplementation were prescribed with regular follow-up. After 3 months of therapy, the size and induration of the truncal plaques had improved (Figure 2(a) and (b)). The linear groove over her forehead remained stable (Figure 2(c)). No new episodes of migraine headaches have occurred.

Discussion

ECDS scleroderma is a linear variant of localized scleroderma characterized by focal involvement of the face and can result in neurological, ocular, and dental complications.[1] It usually affects children. In 2017, a review of the literature suggested that the ECDS variant makes up only 2.4%–4% of AO localized scleroderma cases.[1–3] In contrast, in pediatric studies, ECDS scleroderma constitutes 3%–17.6% of cases.[1] To the best of our knowledge, less than 100 cases of patients with AO linear scleroderma ECDS have been reported.[2,4,5–17]

Patient descriptions of AO localized scleroderma are summarized in Table 1. Of note, while AO ECDS is very rare, ECDS localized scleroderma developing in a patient with pre-existing localized scleroderma is exceedingly rare. While exact estimates are not available, only two cases have been reported; one with pre-existing plaque localized scleroderma and second with linear limb localized scleroderma.[4,10] The majority of patients with AO localized scleroderma were females, and the age of onset ranged from 25 to 65 years. AO ECDS patient characteristics are summarized in Table 2. Extracutaneous manifestations in AO ECDS appear to occur less frequently than in the pediatric population.[1] Neurologic manifestations occured in 0%–30% of patients (Table 2), while ophthalmologic or dental disorders have not yet been described in AO ECDS. The scarcity of extracutaneous manifestations in AO ECDS may be explained by insufficient testing and/or data or by the fact that ocular and bone structures are not undergoing structural changes as seen during childhood.

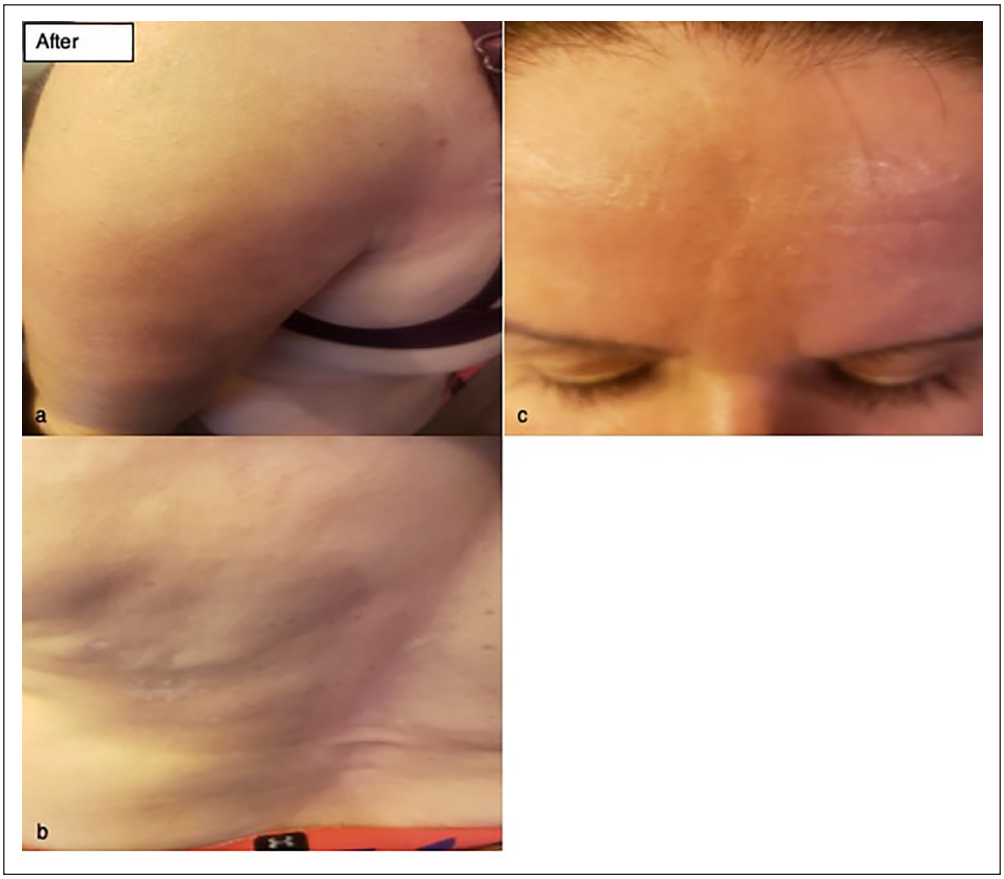

Figure 2. Three months post-treatment with oral methotrexate and oral corticosteroids, reduced size of the indurated plaques was observed on the (a) left shoulder and left triceps and (b) left mid-back. The linear grove mid-forehead (c) remained stable.

Notably, a retrospective cohort study of AO linear scleroderma highlighted that in patients with ECDS, less than half of them underwent MRI brain imaging, and less than a quarter had ophthalmologic evaluation.[4] Furthermore, there was a 27-month delay between symptom onset and the diagnosis by a dermatologist.[4] Thus, the discordance between the diagnosis and the appropriate management may be due to the rarity of the condition in addition to the lack of awareness of the disease incidence in the adult population.

Based on the European and Japanese treatment guidelines for localized scleroderma, generalized, linear, and deep subtypes generally require systemic treatment.[18,19] The recommended first-line therapy is methotrexate with or without systemic corticosteroids, and in case of treatment failure, mycophenolate mofetil is considered a second-line option. In a retrospective cohort study of AO linear scleroderma, most patients (59%) were treated with other agents including tetracyclines, topical corticosteroids, and topical calcineurin inhibitors, followed by methotrexate alone (14%), and subsequently by methotrexate with systemic corticosteroids (9%).[4] Moreover, patients treated with a regimen including methotrexate versus a regimen without methotrexate were more likely to have disease resolution (29% vs 4%).

While our patient reported a new onset of migraines and was followed for long-standing glaucoma, no objective evidence of locoregional complications of scleroderma was noted, and systemic therapy was started within 2 weeks of symptoms onset, which we believe helped to halt disease progression. Furthermore, the pre-existing localized scleroderma (linear trunk and limb) greatly improved with the use of methotrexate and oral corticosteroids, as seen in prior studies.[4]

We describe a case of a new AO ECDS localized scleroderma arising in a patient with pre-existing linear scleroderma of the trunk and limb. This case highlights the importance of recognizing the occurrence in adulthood of a typical childhood-onset variants, linear and specifically, the ECDS type. The literature also highlights the knowledge gap in regard to the investigation and management of linear scleroderma in adult patients. Further studies are needed to confirm the preliminary observations that AO ECDS localized scleroderma has a lower rate of extracutaneous involvement in comparison to pediatric populations, and to establish whether early investigation and subsequent initiation of

Table 1. Characteristics of adult-onset (AO) localized scleroderma.

Study	AO localized scleroderma, n (%)	Female:male ratio	Age at onset (years), median (IQR)	AO linear scleroderma (among all AO), n (%)	ECDS (among all AO), n (%)
Marzano et al.[5]	113 (47.2)	3:1	46 (17–77)	7 (6.2)	4 (3.5)
Arkachaisri et al.[7]	32 (44.4)	5.4:1	32.9 ± 14.5[a]	32 (100.0)[b]	6 (18.8)
Mertens et al.[2]	225 (65.4)	2.8:1	47 (18–86)	31 (13.8)	10 (4.4)
Mazori et al.[4]	61 (100.0)	5.1:1	35 ± 13[a]	61 (100.0)[b]	33 (54.1) + 1 case of concomitant extremity linear scleroderma
Kunzler et al.[6]	348 (59.9)	Not provided	31.1 (23.4–40)	95 (27.3)	27 (7.8)
Unterberger et al.[10]	case report	female	24	1[b]	Patient had a pre-existing plaque localized scleroderma
Miller et al.[11]	case report	male	65	1[b]	1
Taniguchi et al.[12]	9 (62.5)	8:1	36 (21–59)	9 (100.0)[b]	9 (100.0)
Arif et al.[13]	case report	female	26	1[b]	1
Rattanakaemakorn and Jorizzo[14]	1 (14.3)	male	38	1	1
Homayoon et al.[15]	case report	male	25	1	1
Abdelnour et al.[16]	case report	female	30	1	Patient had ECDS overlapping Parry–Romberg syndrome
Yamasaki et al.[17]	case report	female	25	1	1

ECDS: en coup de sabre; IQR: interquartile range.
[a]Mean age of onset, years, standard deviation.
[b]Study examines AO linear scleroderma only.

Table 2. Characteristics of patients with adult-onset (AO) en coup de sabre (ECDS) scleroderma.

Extracutaneous manifestation	% of ECDS patients
Neurologic (seizures, headaches, stoke, MRI evidence of brain involvement)	0–30[4,5,8–10,12]
Ophthalmologic (uveitis, enophthalmos, exophthalmos, adnexa abnormality)	0[4,5]
Dental disease	No reports
Associated diseases	
Lupus erythematosus, Hashimoto's thyroiditis, Sjögren syndrome, rheumatoid arthritis, relapsing polychondritis, antiphospholipid syndrome	2–7[4,5]

MRI: magnetic resonance imaging.

treatment can prevent sequelae. For now, it may be prudent to extrapolate the juvenile localized scleroderma management guidelines to AO head and neck localized scleroderma including assessment for neurologic, ophthalmologic, and dental complications.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article: Dr E.N. is an associated editor of the *Journal of Cutaneous Medicine and Surgery*.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Informed consent

The patient signed informed consent for the collection and publishing of the non-identifiable images. Upon request, the informed consent form can be provided.

References

1. Lis-Swiety A, Skrzypek-Salamon A, Ranosz-Janicka I, et al. Localized scleroderma: clinical and epidemiological features with emphasis on adulthood- versus childhood-onset disease differences. *J Eur Acad Dermatol Venereol* 2017; 31(10): 1595–1603.
2. Mertens JS, Seyger MMB, Kievit W, et al. Disease recurrence in localized scleroderma: a retrospective analysis of 344 patients with paediatric- or adult-onset disease. *Br J Dermatol* 2015; 172(3): 722–728.
3. Kreuter A, Wischnewski J, Terras S, et al. Coexistence of lichen sclerosus and morphea: a retrospective analysis of 472 patients with localized scleroderma from a German tertiary referral center. *J Am Acad Dermatol* 2012; 67(6): 1157–1162.
4. Mazori DR, Wright NA, Patel M, et al. Characteristics and treatment of adult-onset linear morphea: a retrospective cohort study of 61 patients at 3 tertiary care centers. *J Am Acad Dermatol* 2016; 74(3): 577–579.
5. Marzano AV, Menni S, Parodi A, et al. Localized scleroderma in adults and children: clinical and laboratory investigations of 239 cases. *Eur J Dermatology* 2003; 13(2): 171–176.

6. Kunzler E, Florez-Pollack S, Teske N, et al. Linear morphea: clinical characteristics, disease course, and treatment of the Morphea in Adults and Children cohort. *J Am Acad Dermatol* 2019; 80(6): 1664–1670.
7. Arkachaisri T, Fertig N, Pino S, et al. Serum autoantibodies and their clinical associations in patients with childhood- and adult-onset linear scleroderma. *J Rheumatol* 2008; 35(12): 2439–2444.
8. Doolittle DA, Lehman VT, Schwartz KM, et al. CNS imaging findings associated with Parry–Romberg syndrome and en coup de sabre: correlation to dermatologic and neurologic abnormalities. *Neuroradiology* 2014; 57(1): 21–34.
9. Lis-Swiety A, Brzezinska-Wcislo L and Arasiewicz H. Neurological abnormalities in localized scleroderma of the face and head: a case series study for evaluation of imaging findings and clinical course. *Int J Neurosci* 2017; 127(9): 835–839.
10. Unterberger I, Trinka E, Engelhardt K, et al. Linear scleroderma "en coup de sabre" coexisting with plaque-morphea: neuroradiological manifestation and response to corticosteroids. *J Neurol Neurosurg Psychiatry* 2003; 74(5): 661–664.
11. Miller K, Lehrhoff S, Fischer M, et al. Linear morphea of the forehead (en coup de sabre). *Dermatol Online J* 2012; 18(12): 22.
12. Taniguchi T, Asano Y, Tamaki Z, et al. Histological features of localized scleroderma "en coup de sabre": a study of 16 cases. *J Eur Acad Dermatol Venereol* 2014; 28(12): 1805–1810.
13. Arif T, Majid I and Haji ML. Late onset "en coup de sabre" following trauma: rare presentation of a rare disease. *Our Dermatol Online* 2015; 6(1): 49.
14. Rattanakaemakorn P and Jorizzo JL. The efficacy of methotrexate in the treatment of en coup de sabre (linear morphea subtype). *J Dermatolog Treat* 2018; 29(2): 197–199.
15. Homayoon D, Haybaeck J and Aberer E. Adult morphea en coup de sabre with accompanying regional polymyositis: a separate entity. *Australas J Dermatol* 2018; 59(2): e145–e146.
16. Abdelnour JG, Abdelnour YG, Kerollos RM, et al. Parry–Romberg syndrome associated with en coup de sabre in a patient from South Sudan–a rare entity from East Africa: a case report. *J Med Case Reports* 2019; 13(1): 1–5.
17. Yamasaki R, Yonekawa T, Inamizu S, et al. A case of overlapping adult-onset linear scleroderma and Parry-Romberg syndrome presenting with widespread ipsilateral neurogenic involvement. *Neuropathology* 2020; 40(1): 109–115.
18. Asano Y, Fujimoto M, Ishikawa O, et al. Diagnostic criteria, severity classification and guidelines of localized scleroderma. *J of Derm* 2018; 45(7): 755–780.
19. Knobler R, Moinzadeh P, Hunzelmann N, et al. European dermatology forum S1-guideline on the diagnosis and treatment of sclerosing diseases of the skin, Part 1: localized scleroderma, systemic sclerosis and overlap syndromes. *J Eur Acad Dermatol Venereol* 2017; 31(9): 1401–1424.