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

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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.

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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. 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. In contrast, in pediatric studies, ECDS scleroderma constitutes 3%–17.6% of cases. To the best of our knowledge, less than 100 cases of patients with AO linear scleroderma ECDS have been reported.

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. 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. Neurologic manifestations occurred 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 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.](image-url)
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. Furthermore, there was a 27-month delay between symptom onset and the diagnosis by a dermatologist. 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. 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%). 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.

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

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 groove mid-forehead (c) remained stable.
treatment can prevent sequelae. For now, it may be prudent
to extrapolate the juvenile localized scleroderma manage-
ment guidelines to AO head and neck localized scleroderma
including assessment for neurologic, ophthalmologic, and
dental complications.

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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)                       | 5:1               | 32.9 ± 14.5ª                      | 32 (100.0)ª                     | 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.ª        | 61 (100.0)                      | 5.1:1             | 35 ± 13ª                          | 61 (100.0)ª                     | 33 (54.1) + 1 case of concomitant extremity linear scleroderma |
| Kunzler et al.ª       | 348 (59.9)                      | Not provided      | 31.1 (23.4–40)                    | 95 (27.3)                       | 27 (7.8)                    |
| Unterberger et al.ª   | case report                     | female            | 24                                | 1ª                             | Patient had a pre-existing plaque localized scleroderma |
| Miller et al.ª        | case report                     | male              | 65                                | 1ª                             | 1                           |
| Taniguchi et al.12    | 9 (62.5)                        | 8:1               | 36 (21–59)                        | 9 (100.0)ª                      | 9 (100.0)                   |
| Arif et al.ª³         | case report                     | female            | 26                                | 1ª                             | 1                           |
| Rattanakaemakorn and Jorizzo14 | case report                     | 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.
ªMean age of onset, years, standard deviation.
³Study 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, stroke, MRI evidence of brain involvement) | 0–30ª,5,8–10,12 |
| Ophthalmologic (uveitis, enophthalmos, exophthalmos, adnexa abnormality) | 0ª,5 |
| Dental disease                                                | No reports        |

Associated diseases

- Lupus erythematosus, Hashimoto’s thyroiditis, Sjögren syndrome, rheumatoid arthritis, relapsing polychondritis, antiphospholipid syndrome
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MRI: magnetic resonance imaging.

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