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Indonesian Journal of Case Reports

Vol. 4, No. 1, 2026

En Coup de Sabre: A Classical Case of Linear Morphea in a Young Woman

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Article History

Received 7 April 2026
Revised 16 June 2026
Accepted 23 June 2026
Available Online 29 June 2026

Keywords:
Autoimmune skin disorder
Lichen sclerosus
Localized scleroderma
Craniofacial sclerosis
Methotrexate

Abstract

Morphea, or localized scleroderma, is a rare inflammatory disorder characterized by cutaneous sclerosis without systemic involvement. The craniofacial variant, en coup de sabre, is clinically significant due to its potential for deep-tissue extension and neurologic associations. A 22-year-old woman presented with a 10-year history of a progressively widening linear depressed lesion on the forehead. Clinical evaluation showed a well-demarcated atrophic plaque, while laboratory tests, including ANA, were normal. Histopathology revealed thickened collagen bundles with perivascular lymphocytic infiltration, consistent with sclerotic-phase linear morphea. The patient was diagnosed with linear morphea en coup de sabre and treated with methotrexate and corticosteroids, resulting in clinical improvement after 4 weeks. This case highlights the delayed presentation and slow progression of adult-onset craniofacial linear morphea. Despite minimal inflammatory activity, the lesion location carries a risk of deeper involvement, warranting early systemic immunomodulatory therapy. Adult-onset linear morphea en coup de sabre should be recognized in patients with slowly progressive linear atrophic forehead lesions. Early diagnosis and timely systemic treatment are essential to prevent progression and long-term deformity.

1. Introduction

Morphea, or localized scleroderma, is a rare chronic inflammatory connective tissue disorder characterized by excessive collagen deposition leading to skin and subcutaneous sclerosis. Unlike systemic sclerosis, morphea typically does not involve internal organs, sclerodactyly, or Raynaud's phenomenon. The disease demonstrates significant clinical heterogeneity, ranging from isolated indurated plaques to extensive fibrosis with potential functional impairment [1, 2].

Although the exact etiology of morphea remains unclear, various triggers such as infections, medications, and trauma are thought to induce vascular and immune dysregulation in genetically predisposed individuals. T-cell activation, along with the release of cytokines associated with interferon-γ (IFN-γ), plays a key role in initiating inflammatory and profibrotic pathways, ultimately leading to excessive collagen deposition [2, 3].

The reported incidence of morphea varies widely worldwide, ranging from approximately 4 to 27 cases per million annually. Nearly two-thirds of all cases occur in adults, whereas juvenile localized scleroderma has an estimated annual incidence of 3.4–9 cases per million children [3, 4]. Epidemiological data on morphea in Indonesia remain unavailable, reflecting a broader

limitation of population-based studies in many developing countries. However, data from Asia have been reported, particularly from Japan, where juvenile-onset morphea demonstrated an incidence ranging from 2.11 to 2.87 per 1,000,000 children aged <18 years between 2016 and 2019 [5]. Clinically, morphea is classified into several subtypes, including circumscribed, generalized, linear, deep, and mixed forms. Linear morphea *en coup de sabre* (LM) most commonly presents during childhood. However, up to 32% of cases may occur in adulthood, often with a more aggressive clinical course and a higher risk of systemic involvement [6–8].

A notable variant of linear morphea is en coup de sabre, characterized by a unilateral linear, atrophic, and sclerotic lesion typically involving the frontoparietal region. This subtype is clinically important due to its potential to extend beyond the skin, affecting subcutaneous tissue, bone, and even the central nervous system. Neurological manifestations such as seizures and headaches have been reported, with neuroimaging frequently revealing intracranial abnormalities [9, 10].

The diagnosis of morphea is established based on clinical presentation, laboratory investigations, dermoscopic findings, and histopathological examination. Laboratory tests may support the diagnosis and are useful for disease monitoring during follow-up. Antinuclear antibody (ANA) testing shows elevated titers in approximately 40–80% of patients, while antihistone antibodies (AHA) are elevated in about 32–39% of cases, particularly in linear morphea. Dermoscopy can aid in diagnosis, especially in patients with darker skin types, where lesion borders may be less clearly defined on routine clinical examination. Histopathological findings in morphea vary depending on the stage of the disease [11, 12].

The selection of therapy for morphea depends on the location, depth, and extent of the disease. Treatment should be initiated early, prior to the development of complications, and continued until disease activity has resolved. Methotrexate, with or without systemic corticosteroids, is considered the first-line therapy, particularly in moderate-to-severe cases or in linear morphea involving deeper tissues. Topical therapies, including potent corticosteroids, may be used in more limited or superficial disease. Alternative treatment options include mycophenolate mofetil and phototherapy, especially in patients who do not respond adequately to first-line treatment. The recurrence rate of morphea has been reported to range from approximately 28% to 44% within 16–20 months after discontinuation of therapy [11].

Despite its rarity, morphea can result in significant morbidity, particularly in cases with extracutaneous involvement. Early diagnosis and prompt initiation of immunomodulatory therapy, such as methotrexate with or without corticosteroids, are essential to prevent disease progression and long-term complications. This report aims to describe a case of adult-onset linear morphea *en coup de sabre*, focusing on its clinical presentation, diagnostic evaluation, and therapeutic response to systemic immunomodulatory treatment, as well as to emphasize the importance of early recognition and timely management in preventing irreversible craniofacial deformity.

2. Cases

2.1. History

A 22-year-old woman presented to the dermatology department with a chief complaint of a progressive linear depressed lesion on the forehead that had been present for approximately 10 years. The lesion first appeared in early adolescence as a painless, non-itchy, slightly indented 0.5 cm line with no redness. Over time, the lesion gradually enlarged and evolved into a well-defined, linear, atrophic depression extending vertically along the frontal region. The patient described the lesion as resembling a scar-like "cut mark." There was no history of preceding trauma, infection, or inflammation. She also denied associated symptoms such as headache, seizures, visual disturbances, or other neurological complaints. The patient sought medical attention two years ago at a private clinic and received topical corticosteroid treatment. However, no clinical improvement was observed. Her past medical history was unremarkable, with no history of autoimmune disease, chronic illness, or long-term medication use. There was no similar condition in the family, although a paternal relative had a history of unspecified autoimmune disease.

2.2. Examination

On physical examination, vital signs were within normal limits. Dermatological examination revealed a solitary, well-demarcated, hyperpigmented linear plaque with atrophy located on the midline frontal region, extending from the hairline toward the glabella. The lesion appeared depressed with irregular borders and a depth of approximately 0.5 cm (Figure 1). No erythema or induration was noted. No lymphadenopathy or systemic abnormalities were identified.

2.3. Investigations

Laboratory investigations, including complete blood count, liver and renal function tests, inflammatory

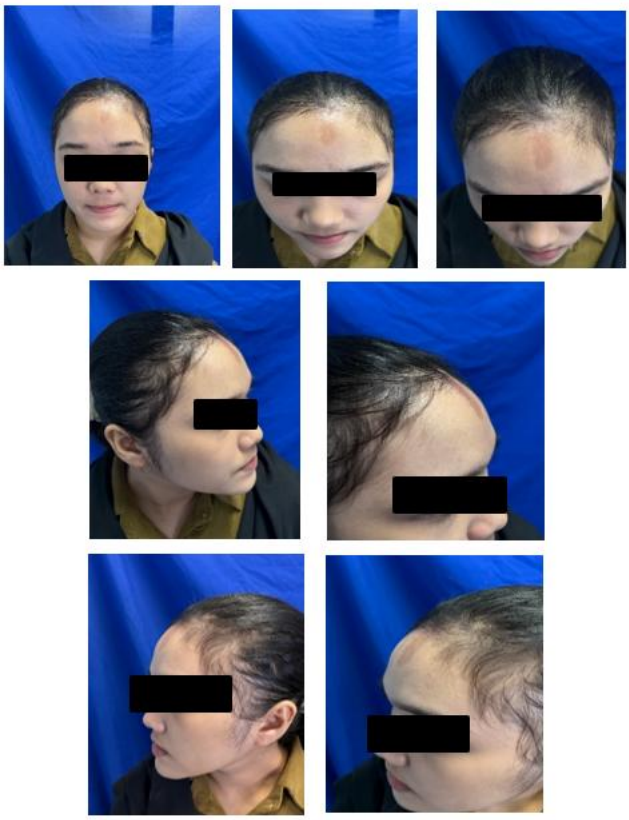

Figure 1. Initial presentation of linear morphea en coup de sabre. A linear, atrophic lesion on the midline forehead demonstrates a characteristic depressed "strike of the sword" appearance, extending from the frontal hairline toward the glabellar region, consistent with craniofacial involvement at first presentation.

markers, and ANA profile, were within normal limits. Histopathological examination of a skin biopsy demonstrated stage-dependent features of morphea. At low magnification (Figure 2a, 40×), there is dermal thickening with densely packed collagen bundles. At higher magnification (Figure 2b, 200×), mild orthokeratosis and perivascular lymphocytic infiltration are observed, supporting a sclerotic-phase lesion consistent with morphea. (Figure 2) Disease Assessment using the Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) showed mild disease activity, with a LoSAI score of 3, indicating minimal active inflammation, and a LoSDI score of 5, reflecting mild dermal and deep tissue atrophy with dyspigmentation.

2.4. Diagnosis and Management

Based on clinical and histopathological findings, a diagnosis of linear morphea en coup de sabre was established. The patient was treated with systemic methotrexate at a dose of 15 mg weekly combined with oral methylprednisolone 40 mg daily, along with folic acid supplementation. Topical therapy included corticosteroid-containing preparations. Patient education was provided regarding the chronic nature of the disease, treatment goals, and the importance of regular follow-up.

2.5. Follow-Up

During follow-up, the lesion demonstrated clinical improvement, characterized by stabilization of disease progression and partial softening of the affected area. No new lesions or extension of the existing lesion were observed, indicating a favorable therapeutic response (Figure 3).

3. Discussions

This case highlights the diagnostic and therapeutic challenges of adult-onset craniofacial linear morphea en coup de sabre, an uncommon but clinically significant presentation that may be underrecognized due to its slow progression, despite its potential for irreversible craniofacial deformity and deeper tissue involvement.

The clinical presentation of a slowly progressive linear atrophic lesion on the forehead, in the absence of systemic features and supported by histopathological findings, was consistent with linear morphea. The absence of systemic sclerosis features and a negative ANA profile further supported a localized disease process. Although morphea is primarily cutaneous [13], Torok et al. [14] reported that extracutaneous manifestations may occur in up to 25% of patients,

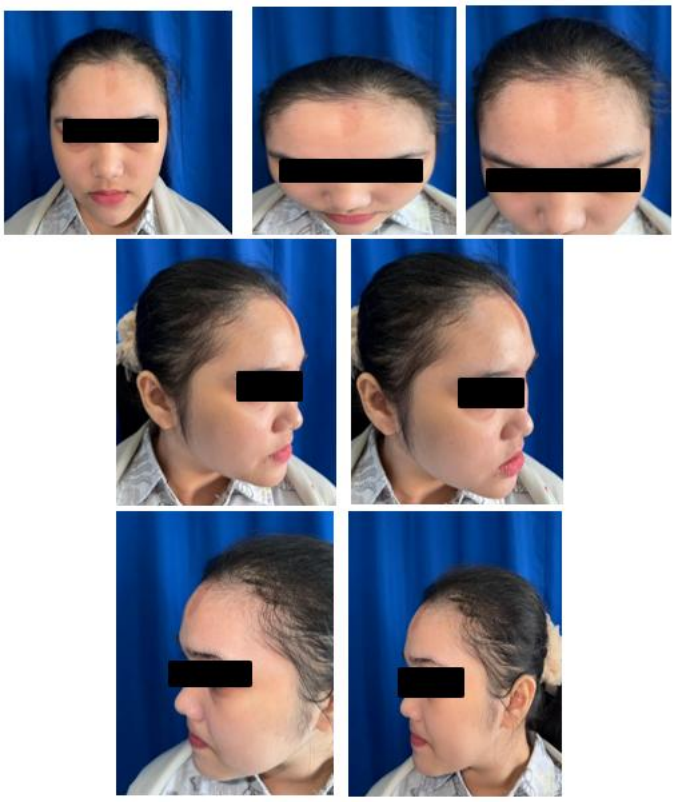

Figure 2. Clinical presentation after 4 weeks of therapy. The linear, plaque-sized hyperpigmented atrophic lesion on the midline frontal region shows noticeable improvement, with reduced depth of depression to approximately 0.2 cm, softer texture, and no further progression. The lesion appears more stable with partial cosmetic improvement compared to the initial presentation.

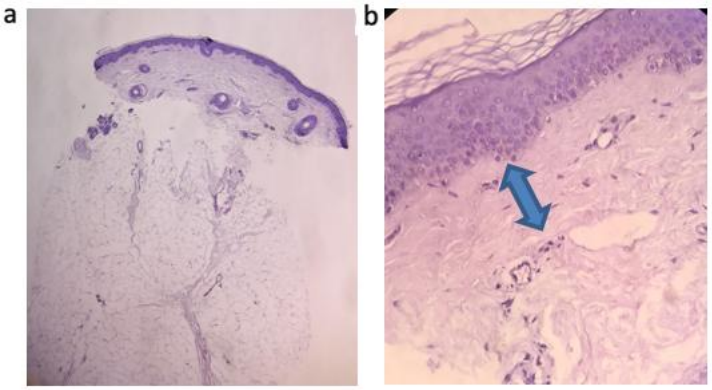

Figure 3. a) A tissue specimen from the forehead skin showed the epidermal layer with mild orthokeratosis, composed of squamous epithelial cells with ovoid nuclei, fine chromatin, and basophilic cytoplasm. In the subepidermal layer, thickening of collagen bundles was observed. Perivascular inflammatory infiltrates predominantly composed of lymphocytes were also noted. Hair follicles, skin adnexal structures, and adipose tissue appeared normal. H.E, 40x. b) Mild orthokeratosis is observed. Thickening of collagen bundles beneath the dermal papillae (blue arrow) is noted. Perivascular inflammatory infiltrates composed of lymphocytes are also present. H.E, 200x.

particularly in linear subtypes, underscoring the importance of careful clinical evaluation in cases such as this [14].

Linear morphea, particularly the en coup de sabre subtype, is a distinctive variant characterized by a linear, atrophic, and indurated lesion involving the frontoparietal scalp or forehead. It commonly begins in childhood or adolescence and follows a slowly progressive course before entering a quiescent phase. The characteristic “strike of the sword” appearance reflects progressive dermal fibrosis leading to a linear depression. The chronic evolution observed in this patient, with gradual enlargement over nearly a decade, reflects the natural progression of the disease. Mertens et al. [11] described that this subtype typically progresses from an early inflammatory phase to a fibrotic and atrophic stage, which aligns with the predominantly sclerotic features seen on histopathology in this case.

Histological findings of thickened collagen bundles and perivascular lymphocytic infiltration further support this stage of disease progression [11, 13].

The histopathological findings in this patient, characterized by thickened collagen bundles and perivascular lymphocytic infiltration, place the lesion within the sclerotic phase of morphea. This stage reflects underlying immune-mediated endothelial injury and fibroblast activation, resulting in increased production of profibrotic cytokines such as transforming growth factor-β and subsequent collagen deposition. Clinically, this correlates with the absence of erythema or induration and the predominance of a depressed, atrophic lesion at presentation. The identification of this phase is clinically relevant, as it suggests a transition from active inflammation toward irreversible tissue remodelling. Therefore, the initiation of systemic therapy with methotrexate in this patient was aimed at suppressing residual inflammatory activity and preventing further progression into a more advanced atrophic stage, where structural damage becomes less reversible [3].

Craniofacial involvement in linear morphea carries particular clinical significance due to its association with deeper tissue extension. Amaral et al. [15] demonstrated that neurologic abnormalities, including intracranial changes, may occur even in asymptomatic individuals, supporting the need for continued surveillance in such cases. While conditions such as Parry–Romberg syndrome share overlapping features with craniofacial linear morphea, including tissue atrophy and possible neurologic involvement, no clinical evidence of hemifacial atrophy or progressive facial asymmetry was observed in this patient. Therefore, the current findings are more consistent with localized en coup de sabre involvement rather than a broader overlapping syndrome [15, 16].

Disease activity and damage were assessed using the Localized Scleroderma Cutaneous Assessment Tool (LoSCAT), revealing mild activity (LoSAI score 3) and mild damage (LoSDI score 5). Skrzypek-Salamon et al. [17] validated LoSCAT as a useful tool for monitoring disease course; this case illustrates its limitations, as low activity scores may underestimate the clinical significance of lesions located in cosmetically and functionally sensitive regions such as the face. Therefore, therapeutic decisions should integrate not only quantitative scores but also disease subtype, anatomical location, and risk of progression [17, 18].

Systemic immunomodulatory therapy remains the mainstay of treatment for active or progressive linear morphea. Methotrexate, with or without systemic corticosteroids, is recommended as first-line therapy, particularly in cases involving the face or deep tissues [19]. Current evidence supports the use of systemic corticosteroids in combination with methotrexate (MTX) as an effective therapeutic approach for active localized scleroderma, particularly in progressive linear and generalized subtypes. Multiple studies have demonstrated that corticosteroids are beneficial in controlling the inflammatory phase of the disease and are generally well tolerated, especially when used as adjunctive therapy with MTX. Torok et al. [14] demonstrated that methotrexate combined with corticosteroids is effective in reducing disease activity and preventing progression, especially in linear morphea. In this case, the patient was treated with systemic methotrexate in combination with oral and topical corticosteroids, resulting in clinical improvement after 4 weeks of therapy [14, 19].

In this case, the prolonged, slowly progressive course over nearly a decade, combined with sclerotic-phase histopathological features and minimal inflammatory activity on clinical assessment, highlights the potential for delayed presentation in adult-onset craniofacial linear morphea. Despite relatively mild LoSCAT scores, the anatomically sensitive location and risk of deeper tissue involvement warranted early systemic therapy, illustrating that treatment decisions should be guided by clinical context rather than scoring systems alone. The favorable response observed after initiation of methotrexate and corticosteroids further emphasizes the importance of timely immunomodulatory intervention, even in cases that appear clinically quiescent. Additionally, the absence of neurologic manifestations at presentation underscores the need for continued longitudinal monitoring, given the possibility of subclinical or delayed extracutaneous involvement in craniofacial disease.

4. Conclusions

In conclusion, this case highlights the characteristic clinical presentation of en coup de sabre morphea as a slowly progressive linear atrophic lesion in the craniofacial region. The diagnosis was established through a combination of clinical findings and supportive histopathological examination, while laboratory investigations were primarily useful in excluding systemic involvement. The patient demonstrated a favorable therapeutic response to systemic immunomodulatory treatment with methotrexate and corticosteroids, as evidenced by clinical improvement after 4 weeks of therapy. This case underscores the importance of early recognition and prompt initiation of appropriate treatment to halt disease progression and minimize the risk of irreversible craniofacial deformity.

Author Contributions: Conceptualization, N.E., A.H., C.Y., and F.D.I.; writing—original draft preparation, W.W.; writing—review and editing, W.W.; supervision, N.E., A.H., and C.Y.; All authors have read and agreed to the published version of the manuscript.

Funding: This study does not receive external funding.

Ethical Clearance: Not applicable.

Informed Consent Statement: Written informed consent has been obtained from the patient for publication of this paper.

Data Availability Statement: The article contains all the necessary data to support the results.

Acknowledgments: The author team would like to thank the Department of Dermatology and Venereology, Faculty of Medicine, Universitas Syiah Kuala/Regional General Hospital Dr. Zainoel Abidin, for its assistance and collaboration in completing this case report.

Conflicts of Interest: All the authors declare no conflicts of interest.

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