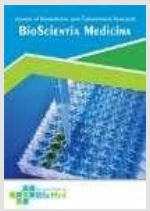
eISSN (Online): 2598-0580



Bioscientia Medicina: Journal of Biomedicine & Translational Research

Journal Homepage: www.bioscmed.com

En Coup De Sabre in 16-Year-Old Girls: A Case Report

Gardenia Akhyar[1], Fauzana Nazifah[1*], Ennesta Asri[1]

[1]Department of Dermatology and Venereology, Faculty of Medicine, Universitas Andalas, Padang, Indonesia

ARTICLE INFO

Keywords:
Autoimmune disease
Dermoscopy
En coup de sabre
Hyperpigmentation
Linear morphea

***Corresponding author:**
Fauzana Nazifah

E-mail address:
dokter.fauzana@gmail.com

All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/bsm.v7i2.770

A B S T R A C T

Background: *En coup de sabre* (ECDS) is one of the clinical variations of morphea. ECDS refers to a variant of linear morphea that usually involves the forehead and frontal scalp that may be associated with alopecia. This study aimed to describe a case of *en coup de sabre* with its diagnosis and management. **Case presentation:** A girl, aged 16 years, came to the dermatology polyclinic with the main complaint of a sunken brown spot on the left forehead and corner of the left eye, which has been expanding since 2 months ago. This complaint does not feel itchy or painful, but because it is getting more widespread, patients feel embarrassed and often cover it up. On dermatological examination, depressed hyperpigmented macules, plaque size, firm boundaries, and location on the left medial superior lid and left medial glabella were found. On dermoscopy examination, a reticular pigment network obliterating the follicular opening was found. The results of the histopathological examination showed the impression of a linear morphea type with *en coup de sabre*. This patient was diagnosed with linear morphea type with *en coup de sabre* **Conclusion:** *En coup de sabre* is a rare autoimmune disorder and is a subtype of morphea. Early detection and appropriate intervention will reduce disease progression.

1. Introduction

En coup de sabre (ECDS) is one of the clinical variations of morphea.[1] ECDS refers to a variant of linear morphea that usually involves the forehead and frontal scalp that may be associated with alopecia. Morphea, or local scleroderma, is an autoimmune disease with inflammation that primarily affects the skin and underlying tissue, which in turn ends in sclerosis.[1,2] Twenty to thirty percent of morphea begins in childhood, with linear morphea being the most common subtype.[3,4] The incidence rate is approximately 25-87% of all cases of children, with 70-80% clinical manifestations in the extremities and trunk. And 22-30% were obtained with *en coup de sabre* or progressive hemifacial atrophy (*Parry-Romberg* syndrome).[1,3] ECDS is usually a clinical diagnosis and can be confirmed by physical examination. Investigation in the form of histopathological evaluation is used to establish the diagnosis.[5] Treatment of ECDS depends on clinical manifestations, depth of lesion involvement, and extent of disease. Treatment primarily aims to inhibit disease progression in active skin lesions.[6,7] This study aimed to describe a case with *en coup de sabre* along with its diagnosis and management.

2. Case Presentation

A girl, aged 16 years, came to the dermatology polyclinic with the main complaint of a sunken brown spot on the left forehead and corner of the left eye, which has been expanding since 2 months ago. This complaint does not feel itchy or painful, but because it

is getting more widespread, patients feel embarrassed and often cover it up. There are no other complaints accompanying these spots. There was no history of a similar disease in the patient's family.

On physical examination, vital signs were within normal limits. On dermatological examination, a depressed hyperpigmented macule was found, with plaque size, firm margin, and location on the left medial superior lid and left medial glabella (Figure 1). The patient has type 4 Fitzpatrick skin. Evaluation of the children's dermatology life quality index (CDLQI) shows a score of 6, which means that skin disorders have a moderate effect on the patient's life. Assessment results based on localized scleroderma skin activity index (LoSAI) is 8 (mild), localized scleroderma skin damage index (LoSDI) is 6 (mild), physician global assessment of disease activity (PGA-A) is 8 (mild), and physician global assessment of disease damage (PGA-D) is 6 (mild). On dermoscopy examination, a reticular pigment network obliterating the follicular opening was found (Figure 2).

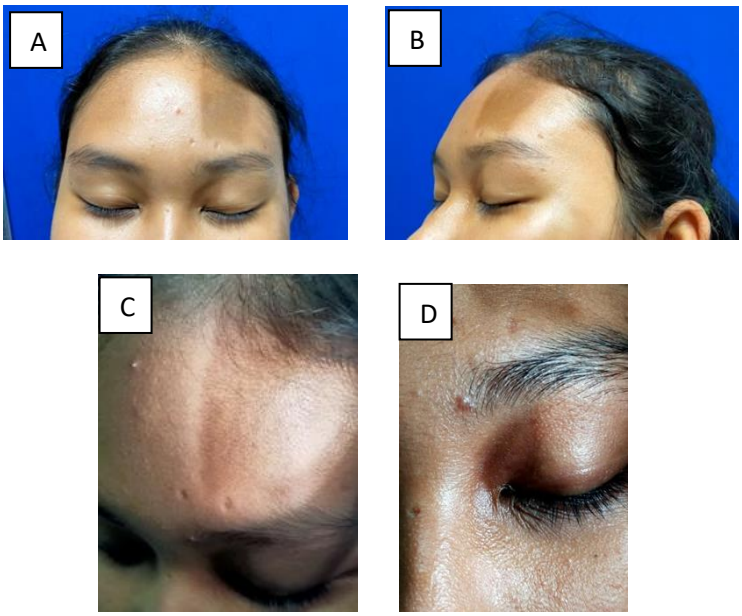


Figure 1. (A-D). Depressed hyperpigmented macules appear in the medial glabella sinistra and superior palpebral medial sinistra.

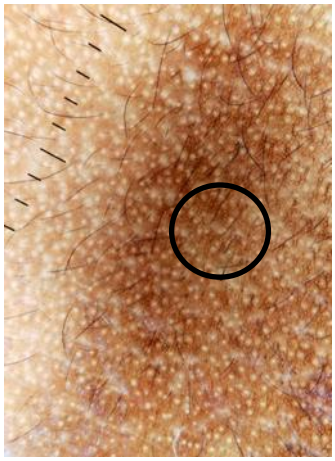


Figure 2. Reticular pigment network obliterating follicular opening (dark circles).

Routine laboratory evaluation of blood showed results within normal limits. Examination of ANA IF and ANA profile showed negative results. The results of the histopathological examination showed the impression of a linear morphea type with *en coup de sabre* (Figure 3). This patient was diagnosed with linear morphea type with *en coup de sabre*

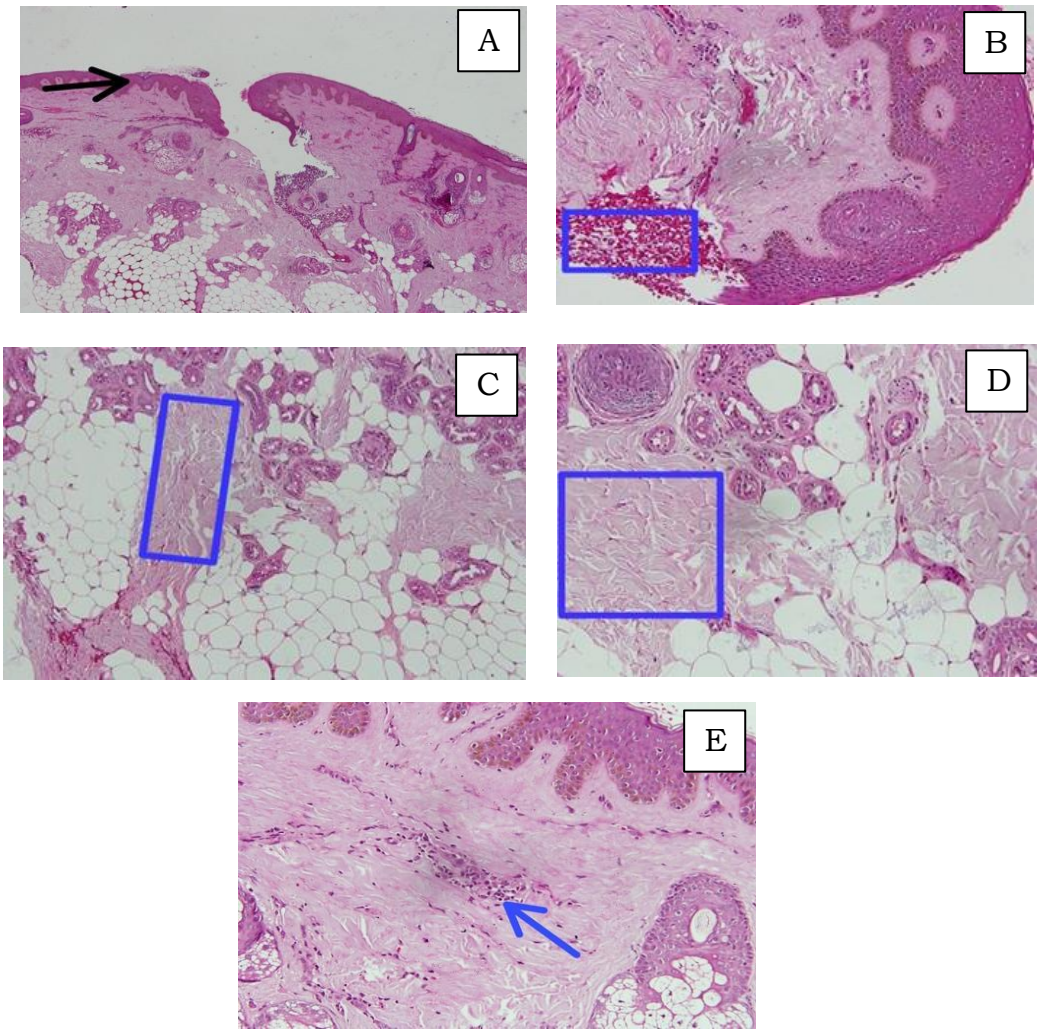


Figure 3. Results of histopathological examination of the skin. (A) Stratified squamous epithelium (black arrow); (B) Bleeding area (blue box); (C) Dense collagen bundles in the subcutis (blue box); (D) Periadnexal dense collagen bundles (blue boxes); (E) Light smear of lymphocytes and plasma cells (blue arrows). (Hematoxillin-eosin staining, 10x magnification).

This patient was treated with 1% pimecrolimus cream on brown spots twice daily for 3 weeks. In addition, patients and families are educated about the disease with *en coup de sabre.* Education includes an explanation that *en coup de sabre* is a chronic autoimmune disease, and its treatment is to prevent the disease from worsening. Therapy with *en coup de sabre* is a long-term therapy, and the results are not obtained quickly.

3. Discussion

En coup de sabre is a rare medical condition that causes skin depressions or visible straight lines on the scalp or face.[1-3] This condition usually appears in childhood or adolescence and can occur on one side of

the face or head.[8] *En coup de sabre* is caused by inflammation of the blood vessels under the skin that causes damage to the skin, subcutaneous tissue, bone, and brain. The exact cause of this condition is unknown, but several theories suggest that with *an en coup de sab*re. It can be related to infection, autoimmune inflammation, or physical trauma. Common symptoms seen with *en coup de sabre are* straight lines or depressions on the scalp or face, discolored or swollen skin, and baldness of the affected areas.[9-11]

En coup de sabre is a subtype of morphea. Extracutaneous manifestations occur in 22-56% of morphea patients.[10] Extracutaneous manifestations are wide-ranging, including musculoskeletal (inflammatory arthritis, articular disease, limb atrophy, joint contractures, limb length discrepancy, and gait disturbances); neurological (peripheral neuropathy, cerebrovascular malformations, neuroimaging abnormalities, seizures, and migraine headaches); ocular (episcleritis, anterior uveitis, glaucoma, and xerophthalmia), and dental complications.[11] Extracutaneous disorders must be monitored carefully and treated aggressively.[4]

Diagnosis with *en coup de sabre* based on clinical symptoms and histopathological examination.[11] Impaired quality of life associated with this disorder can be measured using the CLDQI. If no extracutaneous manifestations and functional limitations are found, generally, *en coup de sabre* has a mild effect on the quality of life. Patients have a value of CDLQI 6 (moderate). This is because adolescence is the age when individuals are very concerned with appearance.

Histopathological examination of skin biopsy specimens provides an overview of the level of inflammation and tissue invasion so that it can assess the stage of the disease and is useful in making therapeutic decisions.[12,13] In the early inflammatory stage of morphea, a perivascular infiltrate dominated by lymphocytes, plasma cells, and eosinophils is seen in the dermis. Endothelial cells may swell and thicken in the collagen bundles. In the final stage of morphea, the inflammatory cell infiltration disappears, thickening of the collagen bundles occurs, eccrine glands atrophy due to being surrounded by hypertrophic collagen, and blood vessels are rarely found. Subcutaneous fat can be seen in the dermis due to the extension of collagen into the subcutaneous tissue.[14] In this patient, it appears stratified squamous keratinized epithelium, dense collagen bundles in the subcutis and periadnexal, as well as a light coating of lymphocytes and plasma cells and bleeding so that it is concluded as a linear morphea of the type with *en coup de sabre*.

Active morphea is generally treated with topical or systemic immunomodulating agents, depending on the subtype and extent of skin involvement.[15] Topical anti-inflammatory agents may be sufficient for superficial, well-defined cases of morphea that do not involve joints or cosmetically sensitive areas. Topical corticosteroids are the first line of treatment for superficial morphea, and the usual course of treatment is for 3-4 weeks. Topical tacrolimus 0.1% is an alternative option for superficially circumscribed morphea.[3] Previous studies stated that the combination of calcipotriol and UVA-1 phototherapy was proven to significantly improve clinical scores in patients with linear morphea.[14] In this patient, given therapy pimecrolimus cream 1% twice a day for 3 weeks on brown spots due to local abnormalities.

Pimecrolimus is used with *en coup de sabre* because it inhibits calcineurin.[15] Pimecrolimus will bind to FK506 binding protein (FKBP) and block the ability of calcineurin to dephosphorylate NFAT and effectively stop transcription of calcineurin-dependent genes for T-cell activation and production of cytokines such as interleukins 2, 4, and 10 and interferon-γ.[16,17] Systemic therapy is generally indicated for progressive lesions. Early and aggressive intervention is essential to prevent irreversible sequelae.

4. Conclusion

En coup de sabre is a rare autoimmune disorder and is a subtype of morphea. Early detection and

appropriate intervention will reduce disease progression.

5. References

1. Beigi PKM. The immunogenetics of morphea and lichen sclerosus. The Immunogenetics of Dermatologic Diseases. 2022; 155-72.
2. Abbas L, Joseph A, Kunzler E, Jacobe HT. Morphea: progress to date and the road ahead. Annals of Translational Medicine. 2021; 9(5).
3. Sapra A, Dix R, Bhandari P, Mohammed A, Ranjit E. A case of extensive debilitating generalized morphea. Cureus. 2020; 12(5).
4. Sink JR, Chiu YE. Pediatric morphea state-of-the-art literature review: Reframing morphea as a systemic disease. Pediatric Dermatology. 2021; 38(5): 1020-31.
5. Li SC, O'Neil KM, Higgins GC. Morbidity and disability in juvenile localized scleroderma: The case for early recognition and systemic immunosuppressive treatment. The Journal of Pediatrics. 2021; 234: 245-56.
6. George R, George A, Kumar TS. Update on management of morphea (localized scleroderma) in children. Indian Dermatology Online Journal. 2020; 11(2): 135.
7. Zhao M, Wu J, Wu H, Sawalha AH, Lu Q. Clinical treatment options in scleroderma: recommendations and comprehensive review. Clinical Reviews in Allergy & Immunology. 2022; 62(2): 273-91.
8. Albadr F, Alnasser HA, Alshathri RM. Ipsilateral hemispheric brain atrophy in an asymptomatic child with linear morphea: A case report. Cureus. 2022; 14(1).
9. Shmuylovich L, Paller AS, Kiguradze T, Anderson K, Sibbald C, Tollefson, et al. 385 Patterning of linear morphea on the face and neck: Blaschkoid or not?. Journal of Investigative Dermatology. 2019; 139(5): S66.
10. Kunzler E, Florez-Pollack S, Teske N, O'Brien J, Prasad S, Jacobe H. Linear morphea: clinical characteristics, disease course, and treatment of the morphea in adults and children cohort. Journal of the American Academy of Dermatology. 2019; 80(6): 1664-70.
11. Jacobe HT, Prasad S, Black SM, Sharma S. Clinical and demographic features of morphea patients with mucocutaneous involvement: A cross sectional study from the morphea of adults and children (MAC Cohort). Journal of Investigative Dermatology. 2020; 140(7): S74.
12. de Albuquerque JV, Andriolo BN, Vasconcellos MR, Civile VT, Lyddiatt A, Trevisani VF. Interventions for morphea. Cochrane Database of Systematic Reviews. 2019; 7.
13. Chiu YE, Abban CY, Konicke K, Segura A, Sokumbi O. Histopathologic spectrum of morphea. The American Journal of Dermatopathology. 2021; 43(1): 1-8.
14. Ulc E, Rudnicka L, Waśkiel-Burnat A, Warszawik-Hendzel O, Niemczyk A, Olszewska M. Therapeutic and reconstructive management options in scleroderma (Morphea) en coup de sabre in children and adults. A systematic literature review. Journal of Clinical Medicine. 2021; 10(19): 4517.
15. McGaugh S, Kallis P, De Benedetto A, Thomas RM. Janus kinase inhibitors for treatment of morphea and systemic sclerosis: A literature review. Dermatologic Therapy 2022; e15437.
16. El-Batawy MM, Bosseila MA, Mashaly HM, Hafez VS. Topical calcineurin inhibitors in atopic dermatitis: a systematic review and meta-analysis. J Dermatol Sci. 2009; 54(2): 76-87.
17. Murrell DF, Calvieri S, Ortonne JP, Ho VC, Weise-Riccardi S, Barbier N, et al. A randomized controlled trial of pimecrolimus cream 1% in adolescents and adults with head and neck atopic dermatitis and intolerant of, or dependent on, topical corticosteroids. Br J Dermatol. 2007; 157(5): 954-9.