Reflectance Confocal Microscopy Applied to Linear (en Coup de Sabre) Morphea

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Established Facts

- Band-like alopecia represents a condition related to linear scleroderma. The diagnosis of this rare disease is clinically supported.

Novel Insights

- The use of reflectance confocal microscopy in this scarring hair disorder could represent a helpful therapeutic and diagnostic tool, especially in the early stage of this condition.

Keywords

Reflectance confocal microscopy · Band-like alopecia · Linear scleroderma

Abstract

En coup de sabre morphea is a clinical variant of morphea, presenting as a linear depressed, atrophic area in the paramedian forehead or the frontoparietal scalp, resembling a stroke from a sword. It affects the skin and subcutaneous connective tissues, with possible extension to the underlying musculature, cartilage, and bone and variable association with neurologic symptoms. A 50-year-old woman pre-

sented to our clinic for evaluation of an atrophic lesion on her forehead and scalp appearing 1 year before, progressively extending over time. An alopecic atrophic area involving the skin and subcutaneous tissues of her right forehead and scalp arranged in a linear fashion with an “en coup de sabre” appearance was observed in relation with hair loss in the affected area. Reflectance confocal microscopy showed marked hyperreflective areas with severe eccrine gland atrophy. All sebaceous glands had disappeared, with sporadic follicular persistence reduction. Histopathological examination of a punch biopsy specimen taken from a central parietal alopecic area was consistent with a diagnosis of morphea. To our knowledge, this is the first report regarding the

use of reflectance confocal microscopy as an ancillary diagnostic technique in linear localized morphea of the scalp and face. This noninvasive technique may represent a useful tool in distinguishing between early stages of the disease, with prevalence of inflammatory lymphocytic infiltrate, and late stages characterized by more prominent sclerosis with mild or absent signs of inflammation.

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Introduction

En coup de sabre (ECDS) morphea is a clinical variant of morphea, presenting as a linear depressed, atrophic area in the paramedian forehead or the frontoparietal scalp, resembling a stroke from a sword [1]. It affects the skin and subcutaneous connective tissues, with possible extension to the underlying musculature, cartilage, and bone, and it has variable association with neurologic symptoms [1]. Loss of adnexal structure due to prominent sclerosis of the dermis is characteristic and is most typical of late stages [2]. The inflammatory, fibrosing process results in a nonspecific scarring alopecia. The condition is more common in women and is associated with a higher rate of autoimmune disorders; thus, a complete clinical assessment is recommended [1]. Diagnosis relies on clinicopathological examination. Histopathological features vary, ranging from pictures of sparse perivascular inflammatory infiltrate with mild or absent fibrosis to pictures characterized by prominent sclerosis with no inflammation [2].

Case Report/Case Presentation

A 50-year-old woman presented to our Clinic for evaluation of an alopecic patch on her scalp appearing 1 year before on the right side of the forehead, progressively expanding over time. Dermatological examination revealed an ivory-colored, atrophic, and indurated hairless area involving her scalp and forehead, arranged in a linear fashion with an “en coup de sabre” appearance. On her right forehead, atrophy of the subcutaneous tissues and bone recess with facial dysmorphism were observed. Personal history included positivity for antinuclear antibodies for many years. Family history reported a case of death for severe systemic sclerosis (Fig. 1–2).

We performed reflectance confocal microscopy (RCM) on an alopecic area of the scalp before performing a punch biopsy of the lesion.

Histopathological examination of a punch biopsy specimen taken from a central parietal alopecic area revealed thickened collagen bundles infiltrating the entire dermis extending into the subcutaneous fat with mild perivascular and focal interstitial lymphocytic infiltrate. Atrophy of sebaceous glands and follicular remnants consistent with an end-stage alopecia were also observed. A diagnosis of late ECDS morphea was made (Fig. 3).

RCM mosaic showed marked hyperreflective areas with severe eccrine gland atrophy. All sebaceous glands had disappeared, with the persistence of sporadic, reduced in size follicles (Fig. 4).

The patient underwent a complete panel of blood and instrumental tests, ruling out a systemic autoimmune condition. Treatment with methotrexate 7.5 mg/week was prescribed.

Discussion

Morphea is a fibrosing condition limited to the skin, subcutaneous tissues, and underlying bone. Involvement of the central nervous system is rare in relation to the face

Fig. 1. Clinical image of coup de sabre atrophy of patient's face.

Fig. 2. Clinical image of linear alopecia.

and head area localization [1]. Ocular involvement may be asymptomatic; thus, regular ophthalmologic examinations are recommended for children with head and neck morphea [3]. Young females (mean age 13.6 years) are most commonly affected, and more likely to have a family history of autoimmunity [4].

ECDS morphea indicates a peculiar clinical presentation, characterized by an atrophic, indurated area on the paramedian forehead or the frontoparietal scalp, resembling a stroke from a sword. Although very typical, a sim-

Fig. 3. Histopathological examination of a punch biopsy specimen taken from the central zone of the linear parietal alopecia revealed thickened collagen bundles infiltrating the entire dermis extending into the subcutaneous fat with mild perivascular and focal interstitial lymphocytic infiltrate. Atrophy of sebaceous glands and follicular remnants consistent with an end-stage alopecia were also observed. A diagnosis of late “en coup de sabre” morphea was made.

ilar clinical presentation could be observed in discoid lupus erythematosus, linear lichen sclerosus, and linear alopecia areata [5–8]. Diagnosis is based on histology; however, specific dermoscopic features have been identified, suggesting a role for noninvasive tool in diagnosis and management of morphea and lichen sclerosus [9]. No studies regarding the use of RCM in ECDS morphea have been published to date.

In our case, RCM described marked hyperreflective areas with severe eccrine gland atrophy. All sebaceous glands had disappeared, with sporadic follicular persistence reduction (Fig. 3).

These atrophic follicular remnants could be related to telogen follicles but were microscopically different. The morphology of these persistent follicles suggested an end-stage alopecia.

We believe that in cases of scarring alopecia, including morphea or connective tissue disorders, especially in early stages, RCM could represent a useful second-line diagnostic and therapeutic tool avoiding unnecessary biopsies.

Conclusion

To our knowledge, this is the first report regarding the use of RCM as an ancillary diagnostic technique in linear localized morphea of the scalp and face. This noninvasive tool may represent a useful method for distinguishing between early stages of the disease, with prevalence of inflammatory lymphocytic infiltrate, and late stages characterized by more prominent sclerosis with mild or absent signs of inflammation.

Further studies are needed to evaluate the usefulness of RCM in the diagnosis and therapy management of hair disorders.

Fig. 4. RCM mosaic shows marked hyperreflective areas with severe eccrine gland atrophy (blue circle). All sebaceous glands disappeared, with the persistence of sporadic, reduced in size follicles (red circles).

Statement of Ethics

The authors declare that the subject has given her written informed consent to publish her case (including publication of images).

Disclosure Statement

All authors have not conflict of interest.

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Author Contributions

Each author has (1) made substantial contributions to the conception or design of the work, or to the acquisition, analysis, or interpretation of data for the work; (2) participated in drafting the work or revising it critically for important intellectual content; (3) approved the final version to be published; (4) agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.