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Case Report

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Linear scleroderma *En coup de sabre* treated with a combination of soft-tissue fillers, microneedling and botulinum toxin. A case report

Abstract

Localized scleroderma (morphea) is a chronic inflammatory, immune-mediated disease. It is characterized by an area of fibrosis due to increased collagen production. *En coup de sabre* is a rare type of band-like scleroderma. The typical clinical finding is a linear lesion, often originating from one of the eyebrows and extending to the scalp, resembling a sword cut. Hair loss, skin dyschromia, and tissue atrophy may also be evident. There may be neurological and ophthalmological complications. Since this condition is more often seen in young patients and involves the visible face and scalp, linear scleroderma also benefits from aesthetic refinement, especially when standard therapy aims to stop disease progression rather than correct the cosmetic defect. We report a rare case of mixed morphea presenting with disseminated truncal plaques and a longstanding linear lesion *en coup de sabre*, successfully managed using a three-step aesthetic protocol combining radiofrequency microneedling, hyaluronic acid fillers, and botulinum toxin type A. No early or late adverse events occurred. Sustained clinical improvement in the tissue elasticity, volume restoration and facial symmetry was observed at 6 months follow-up. This case presents the role of minimally invasive aesthetic procedures as additional tools for structural improvement in stable morphea.

Keywords: Morphea, *en coup de sabre*, microneedling, filler, botulinum toxin, fibrosis

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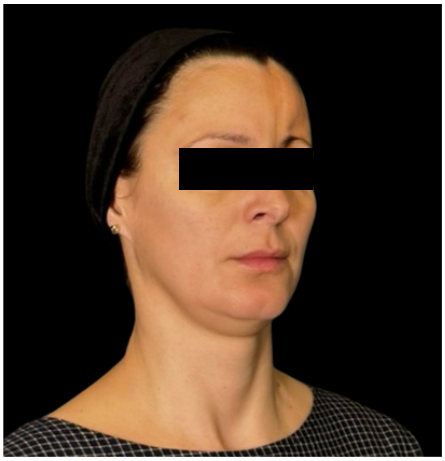
Introduction

Localized scleroderma (morphea, sclerodermia localisata, sclerodermia circumscripta) is a chronic, inflammatory skin disease, which manifests as a disorder of cellular immunity, microcirculation and abnormal collagen synthesis.[1] It can affect both adults and children. The exact aetiology still remains unknown. Many factors may contribute to disease development, including genetic predisposition, vascular dysregulation, Th1/Th2 imbalance with chemokines and cytokines associated with interferon-Y and profibrotic pathways as well as certain environmental factors.[2] There are different classifications regarding localized scleroderma, but the most frequent variants are: plaque-like morphea, guttate morphea, disseminated morphea, linear morphea (*En coup de sabre* and linear morphea of the limbs), erythematous localized scleroderma, nodular morphea, morphea profunda and others. Treatment options are selected based on disease severity and type. Standard therapy includes topicals (emollients, local corticosteroids, local tacrolimus, and imiquimod), phototherapy-based treatments (UVA1, PUVA, narrowband UVB, extracorporeal photopheresis), and systemic therapy with corticosteroids, methotrexate, and mycophenolate mofetil. Other possibilities are penicillin and antimalarials. Therapeutic decisions depend on the clinical subtype, disease activity, the depth of skin involvement, and the characteristics of tissue damage. Given its predominantly young age at presentation, particularly in females, and its location on exposed body parts, linear scleroderma warrants aesthetic correction. According to the standard guidelines therapeutic options tend to halt disease progression but do not address cosmetic defects. To date, various methods have been reported for the management of this deformity.[3] Reconstruction approaches include autologous tissue grafting, alloplastic materials, tissue expanders, and soft-tissue fillers.[4] There are also many reports on the usage of different energy-based devices such as fractional CO2 laser, pulsed dye laser, excimer laser, Q-switched laser, long – pulsed Nd: Yag laser.[5]

Case report

A 41-year-old Caucasian female patient presented with a longstanding band-like (linear depressed) lesion from the left eyebrow to the scalp. Clinical examination revealed a linear, ivory-colored depression causing periorbital asymmetry and upper facial disharmony (Figure 1A, B).



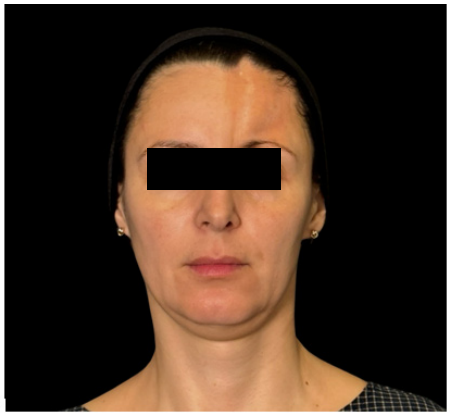


Figure 1 A deep loss of skin and subcutaneous tissue of the forehead: A - anterolateral image; B – anterior image).

Two additional indurated, plaque-like, tender lesions engaging the skin on her back and her left hip were observed (Figure 2).

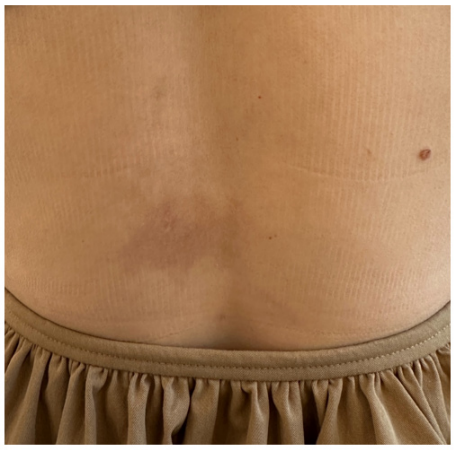


Figure 2 Plaque morphea lesion in the lumbar region.

According to the patient's medical history she was clinically diagnosed with localized scleroderma "*En coup de sabre*" at the age of six year. No previous trauma was reported. In addition, at the age of 36 she noticed two new lesions with circle shape, engaging the skin of her body (back and left hip) Histopathological biopsy, performed from the lesion on her back revealed sclerotic process, eosinophils, lymphocytic inflammatory infiltrate around the superficial and deep vascular plexi, which corresponds with morphea (Figure 3).

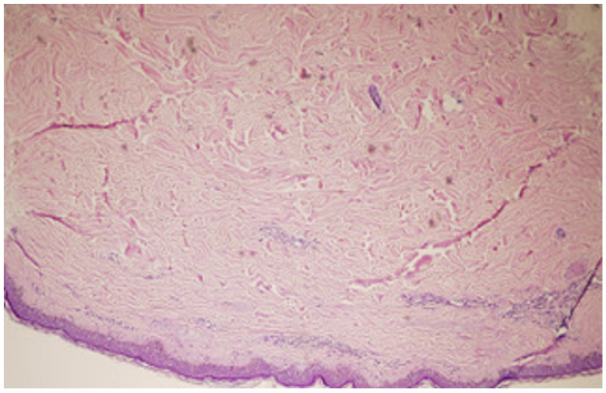


Figure 3 Biopsy result from skin of her lower back: sclerotic process, eosinophils, lymphocytic inflammatory infiltrate around the superficial and deep vascular plexi, which corresponds with morphea.

The laboratory results showed no abnormalities, the ANA and Scl-70 ELISA tests were negative. Brain MRI showed no cortical atrophy. The patient was diagnosed with mixed morphea- a rare form of localized scleroderma, which combines disseminated lesions and the single band pattern.

The patient underwent treatment in a dermatology department, which combined 14 sessions of UVA1 phototherapy, topical steroids and topical tacrolimus. A slight improvement was observed in the two plaque-like lesions on the trunk; however, the scar-like lesion on the forehead showed no response to the therapy. Since the lesion is located on a visible part of the face, the patient reported low self-esteem and impaired quality of life, and as there is a lack of progression and activity we decided to design a protocol to improve the cosmetic defect and restore harmony, balance, and symmetry of upper face.

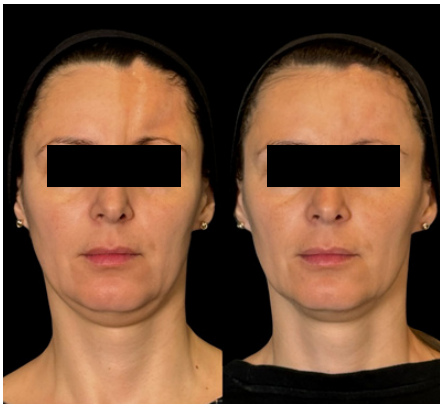
As a first step, the patient was treated with three sessions of RF Microneedling (Exion RF). The lowest possible energy setting (20%) was used, with a needle depth of 1.0mm. The whole affected area was covered uniformly with one pass. At the follow- up visit after the third session, the patient reported decreased tissue induration and improved skin mobility.

Approximately two weeks later, 1 ml cross-lined hyaluronic acid filler (12 mg/ml vycross technology (SKINVIVE by Juvederm) was injected using a micro-bolus technique with 32 needle. The atrophic area was injected slowly with micro boluses of about 0.01-0.05 ml.

One week later, an additional 1 ml of cross-linked HA 15mg/ml (VOLBELLA by Juvederm) was injected with 0.7 ml distributed in micro boluses, covering the whole atrophic area and 0.3 ml used to correction the A-frame deformity on the left side with 25G x 38 mm cannula. Slow injection with intermittent pauses was emphasized due to the area which is considered as one of the most dangerous zones on the face when it comes to injecting cross-linked HA because of the supraorbital and supratrochlear arteries and the risk of vascular complications. Gentle massage was applied right after the procedure. After the total volume of 2 ml hyaluronic acid, a visible improvement in tissue volume and contour was achieved.

Ten days later the patient was injected with a total amount of 32 units of Botulinum toxin type A (Vistabel) Twenty units were administered in the glabella region in 5 injection points, and 12 units were injected into the forehead only on the right side, the one with muscle activity.

At the 2-week follow-up, the patient showed significant improvement, especially in the symmetry of the interbrow region, as well as improved overall harmony and balanced upper face. The volume loss and tissue atrophy were less evident in both profile and frontal views (Figure 4A, B).



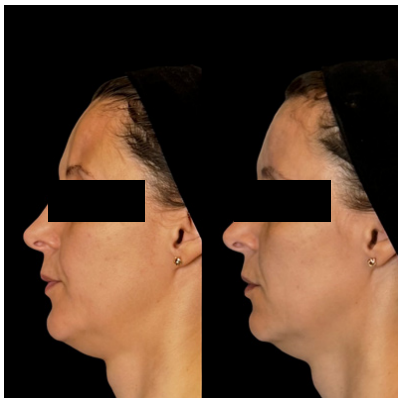


Figure 4 Effect of treatment before and after botulinum toxin and two treatments with hyaluronic acid products - at the 2-week follow-up (A - profile image; B – anterior image).

Discussion

In recent years, the rapid advancement of aesthetic medicine has led to the broader application of its techniques beyond their traditional roles in beautification and anti-aging. Increasingly, these methods are being utilized to address various skin diseases and conditions. Fibrosis is a hallmark of both scar formation and morphea, suggesting that devices and procedures commonly employed for scar treatment could also be effective modalities for managing morphea.

In agreement with Busch et al.,[6] who reported that microneedling seems to be a suitable therapy approach for treating atrophic burn scars, and relying on one more study conducted by El- Domyati et al.,[7] in which there was noticeable clinical improvement in atrophic post acne scars in response to skin microneedling, we also decided to start our treatment protocol with three sessions of microneedling. The efficiency of microneedling in morphea can be explained by the creation of micro punctures, which produce controlled skin injuries without disrupting the epidermis.[8] This process triggers a wound healing cascade, thus releasing various growth factors and resulting in increased dermal elastin and collagen, collagen remodeling and thickening of epidermis and dermis.[9] These effects may also help soften fibrotic tissue in morphea.

In this case, radiofrequency microneedling was selected over conventional microneedling to enhance remodelling effect. RF microneedling combines mechanical skin perforation with controlled thermal energy. This produces immediate collagen contraction followed by long-term collagen reorganization. Studies have shown that RF microneedling results in stronger dermal remodeling and improved tissue elasticity compared to microneedling alone.[10] Experimental data also demonstrate anti-fibrotic effects of RF energy, including reduced expression of extracellular matrix proteins and modulation of fibrotic pathways, supporting its potential benefit in morphea.[11] Although direct studies of RF microneedling in morphea remain limited, conventional microneedling has already shown significant clinical and histopathological improvement in morphea lesions with minimal adverse effects.[12] The additional RF is expected to enhance dermal remodeling and therapeutic effect.

HA filler was a choice in this case because hyaluronic acid fillers are preferred for facial reconstruction and volume loss. Collagen stimulators such as calcium hydroxyapatite (CaHA) and poly-L-lactic acid (PLLA) can induce prolonged inflammation, which, theoretically, may activate or exacerbate morphea in patients with sclerosing skin conditions, but this risk remains unproven.[13] Its use in the correction of scleroderma, a chronic disease, raises many concerns and can lead to inflammation and relapse. Collagen stimulants, such as hydroxyapatite derivatives (CaHA) and poly-L-lactic acid (PLLA), have been associated with inflammatory side effects that may trigger activation or exacerbation of scleroderma in individuals with scleroderma-related features, although this risk remains unconfirmed.[13] However, the literature reports cases of CaHA use in morphea, version.[14]

The ideal injectable agent should be non-immunogenic, biocompatible, and present at the implant site earlier, with a characteristic appearance. The hyaluronic acid used is innovative due to its reduced degradation rate and high water absorption, making it suitable for correcting even skin defects. Furthermore, HA acts as a scaffold for newly formed fibroblasts, stimulating proliferation of type I and II cells and angiogenesis.[15] The literature contains information on the correction of facial defects in scleroderma, particularly *En coup de saber* form.[15,16] Sharad analyzed 100 published cases after HA correction. It does not involve reactivation or exacerbation of the disease for up to 2 years after the procedure. However, it is important to inject fillers only in specific, localized cases to assess the course of the disease.[16]

Studies of the use of BTX-A in hypertrophic scars showed that the toxin inhibited proliferation and increased apoptosis of fibroblasts activated by TGF-β1 (transforming growth factor - β) in a dose-dependent manner. TGF-β is a profibrotic mediator in scleroderma, responsible for excessive collagen synthesis and for inhibiting extracellular matrix degradation, which leads to skin sclerosis and is considered a key factor in the pathogenesis of the disease.[17]

Injection of botulinum toxin type A is particularly beneficial in patients with morphea who develop unilateral reduced mobility due to sclerotic plaques. In addition, botulinum toxin does not induce a pro-fibrotic response, which makes it a safe option for both cosmetic and functional improvement in sclerosing skin disorders such as morphea.[13]

Conclusion

This clinical case demonstrates a multimodal approach for the treatment of stable morphea. Non-surgical techniques such as radiofrequency microneedling, hyaluronic acid fillers and botulinum toxin type A were used. Radiofrequency microneedling was the primary tool used to improve skin quality in the affected area. Hyaluronic acid fillers injections were used to restore volume and cutaneous fat loss and injection of botulinum toxin type A helped restore muscle disbalance in the upper face. The combination of these modalities resulted in visible aesthetic improvement and enhanced quality of life for the patient. Further studies and long-term follow-up are needed to evaluate the durability of the proposed treatments and therapeutic strategy, as well as larger patient cohorts with histological assessment.

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Informed consent

Written informed consent was obtained from all concerned patients.

Conflict of interest

The authors declare there is no conflict of interest.

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