

Enhancement of the aesthetic outcome of scleroderma en coup de sabre with botulinum toxin injection



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INTRODUCTION

Linear scleroderma is a rare form of localized scleroderma. The involvement of the frontal or frontoparietal region with linear scleroderma is called en coup de sabre (ECDS). ECDS presents as linear sclerotic depressed groove appearing on the frontoparietal region. It is usually unilateral and extends from the forehead into the frontal scalp. Its paramedian location is more common than its median one. The sclerotic changes affect the soft tissue and muscles as well as adjacent structures.¹ Linear scleroderma has diverse etiologies, and its pathogenic mechanism is still unclear. Botulinum toxin (BTX) is an exotoxin produced from *Clostridium botulinum*. It blocks the release of acetylcholine from the cholinergic nerve end plates, resulting in inactivity of the muscles or glands innervated. It has gained a great interest in cosmetic dermatology for its effects on hyperkinetic facial lines and focal hyperhidrosis.² Recently, BTX has also been used experimentally in many noncosmetic dermatologic conditions with agreeable results.² We present 2 patients with disfigurement-associated ECDS treated successfully with BTX injection.

Case 1

A 33-year-old man presented to our clinic seeking a BTX injection. He had ECDS on the median of his forehead for the past 7 years, during which he was treated with oral methotrexate and a topical combination of calcipotriol and betamethasone dipropionate. At the time of the injection, he had not received any treatment for >1 year because ECDS was inactive for that year. Photographs were taken at rest and during full frown in frontal view before injection and 10 days after treatment, with the same camera, lighting, and distance parameters.

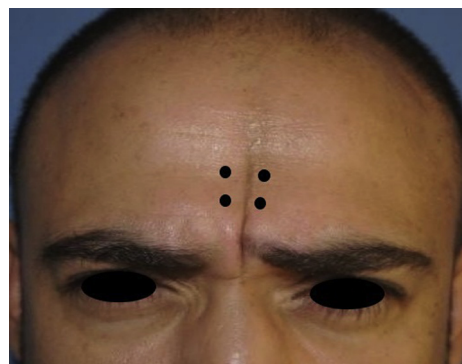


Fig 1. Sites of botulinum toxin injection.

BTX-A vials were reconstituted as follows: 2.5 mL of normal saline solution were added to 100 U vials for a final concentration of 40 U/mL. He was injected with BTX in the glabellar lines, crow's feet, and horizontal forehead lines with a total dosage of 40 U. To treat ECDS within itself, 2 injection points around the periphery of the lesion on each side, spaced 1.5 cm apart, were injected with 1 to 2 units of BTX in each point (total dosage of 2-4 units per side; Fig 1). The patient was instructed to report any side effects. The photographs were then evaluated by the author. On the follow-up visit (10 days postinjection) there was an improvement in the hyperfunctional facial lines and a remarkable decrease in sclerotic changes, contraction of the frontalis muscle, and firming of the skin with mild decrease of hyperpigmentation (Fig 2). No side effects were reported.

Case 2

A 29-year-old woman presented with a request for a BTX injection. She had a 3-year history of ECDS on her forehead. The patient had not received any

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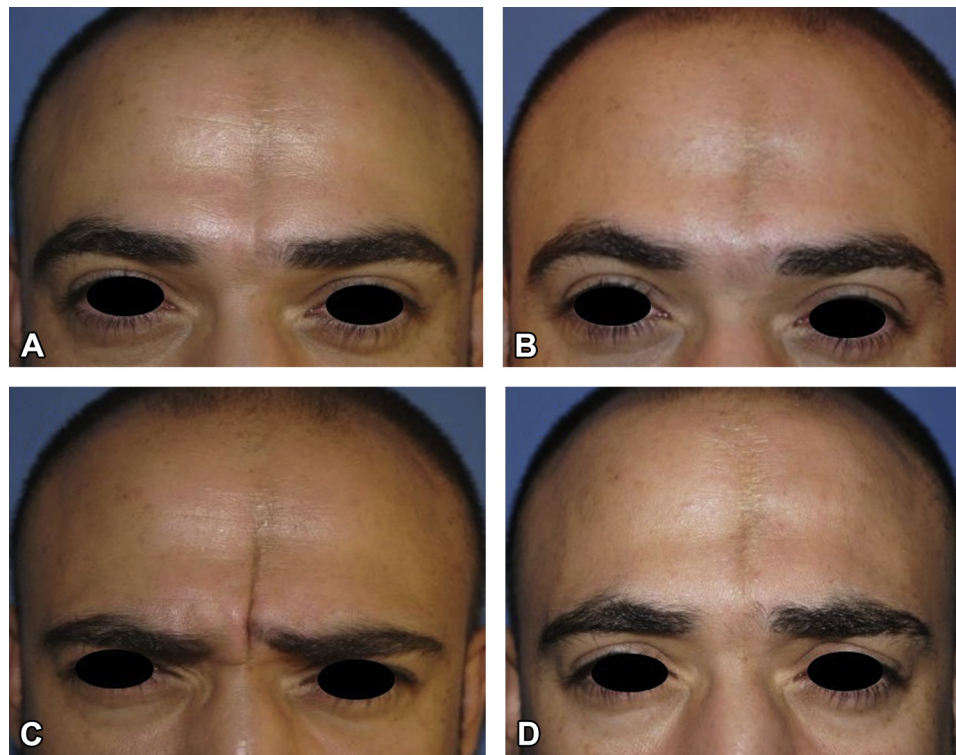


Fig 2. Case 1 before the botulinum toxin injection with the face at rest (A) and frowning (C), and 10 days postinjection with the face at rest (B) and frowning (D).

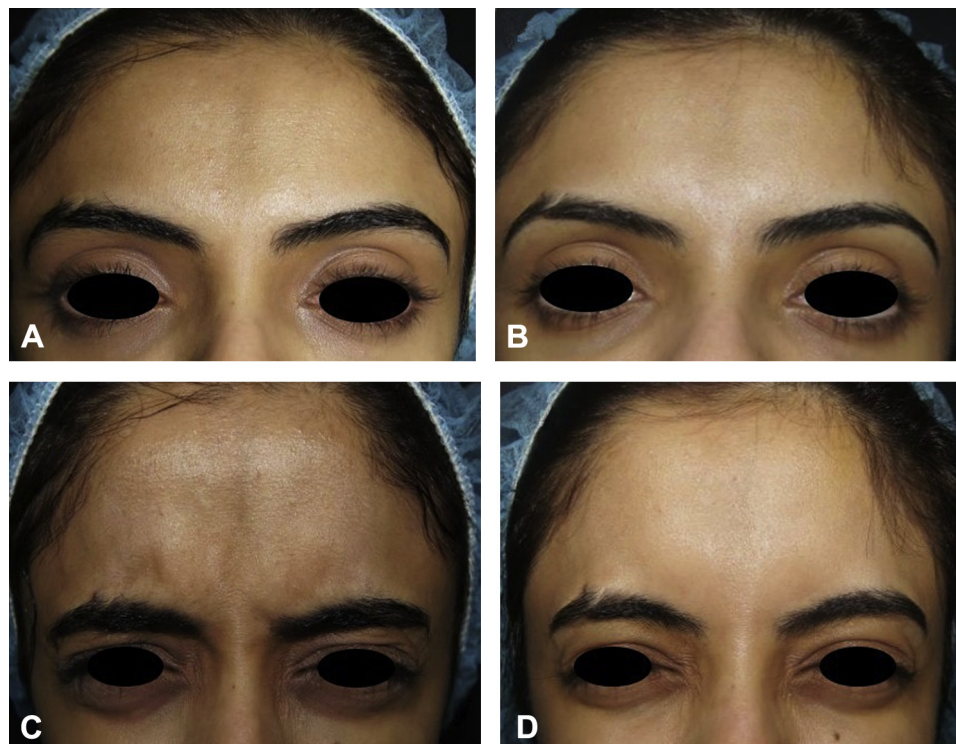


Fig 3. Case 2 before the botulinum toxin injection with the face at rest (A) and frowning (C), and 10 days postinjection with the face at rest (B) and frowning (D).

treatment for the previous 10 months because ECDS was inactive. Her ECDS was treated with BTX and she was given BTX for her hyperfunctional facial lines. She was injected in the same sites as the previous patients. The number of units was less (1 unit on each site), however, because her muscles were weaker in comparison to the first patient. On the follow-up visit (10 days postinjection) there was a remarkable aesthetic improvement for her lines and ECDS (Fig 3).

DISCUSSION

Scleroderma ECDS is a rare inflammatory condition of the skin and underlying tissues caused by dysfunctional collagen. The etiology of ECDS is unknown. However, some cases respond to immunosuppressive treatment, suggesting a possible autoimmune process.³

Although phototherapy and systemic immunomodulators, such as methotrexate and pulsed intravenous corticosteroids, have the highest level evidence for efficacy in improving the appearance of scleroderma, these modalities are not always feasible nor are they always successful.¹

The mechanism of BTX in the patients with scleroderma ECDS is still unclear. It may help relax the tense muscles by blocking the release of acetylcholine. It also demonstrates the efficacy of BTX in improving disfigurement associated with ECDS. The efficacy of BTX in facial aesthetics is well established; however, recent literature has highlighted its use in multiple noncosmetic medical and surgical conditions.⁴

We described 2 patients with disfigurement-associated linear scleroderma ECDS treated successfully with BTX injection. Both patients had excellent aesthetic outcomes, were satisfied with the cosmetic outcomes, and were willing to repeat the treatment in the future. The presence of ECDS on the middle of the forehead is aesthetically unpleasant. In these cases, we are not treating the ECDS itself, but are instead improving the appearance of the patients by reducing the groove induced by ECDS. The usage of BTX for treatment of ECDS is only effective when the disease is inactive.

New uses of BTX injection are being recognized, and indications for the use of BTX have expanded on a large scale.

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