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CASE REPORT

Treatment of Linear Morphea (en Coup de Sabre) with Micronized Acellular Dermal Matrix Filler: A Case Report

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En coup de sabre variant of linear morphea (LM) is a rare sclerotic skin disorder characterized by disfiguring linear depression of the frontal or frontoparietal forehead. Current attempts for cosmetic correction of atrophic lesions must be preceded by an evaluation of disease activity of LM, either by a sufficient clinical assessment or histologic evidence. Corrective procedures including corrective surgery, autologous fat grafting, hyaluronic acid filler injections were performed with varying degrees of success; still, there is a need for treatment options with non-invasive and long-term maintenance effects. Herein we report the use of micronized acellular dermal matrix filler as a novel and successful treatment for the atrophic defect of LM in a 24-year-old female. Molecular characteristics of the micronized acellular dermal matrix filler give enhanced durability and prolonged volume consistency, which results in a long-term extracellular matrix remodeling effect. **(Ann Dermatol 33(4) 373～376, 2021)**

-Keywords-
Dermal fillers, Morphea, Scleroderma, localized

INTRODUCTION

Linear morphea (LM) is a rare localized variant of scleroderma, and the term "en coup de sabre" (ECDS) refers to atrophic linear depression of the frontal or frontoparietal forehead extending to the scalp. The underlying etiology of LM is still poorly understood, although some have suggested that morphea represents a mosaic form of systemic sclerosis limited to the skin[1]. LM may have a self-limited course, but it frequently has a relapsing or chronic nature despite aggressive medical treatments during the active phase of the disease. Moreover, delayed diagnosis and subsequent permanent cosmetic sequelae are common, especially for adult-onset LM[2].

Cosmetic correction of LM with a residual atrophic scar involves the use of invasive techniques, such as corrective surgery, autologous fat grafting, and placement of synthetic tissue matrix, which has been performed with varying degrees of success[3]. However, considering the potential risk of re-triggering LM by surgical trauma, there is a need for minimally invasive techniques with appropriate timing of intervention. Herein, we describe the use of micronized acellular dermal matrix filler as a novel and successful treatment for the atrophic defect of LM. We received a consent form from the patient about publishing the photographic materials.

CASE REPORT

A 24-year-old female presented to our clinic with a linear vertical depression on the left medial forehead that extended to the scalp (Fig. 1A, C). The area was asymptomatic, and she did not remember the exact onset of the disease, but she reported that the lesion had been gradually progressing for about 1 year. There were no asso-

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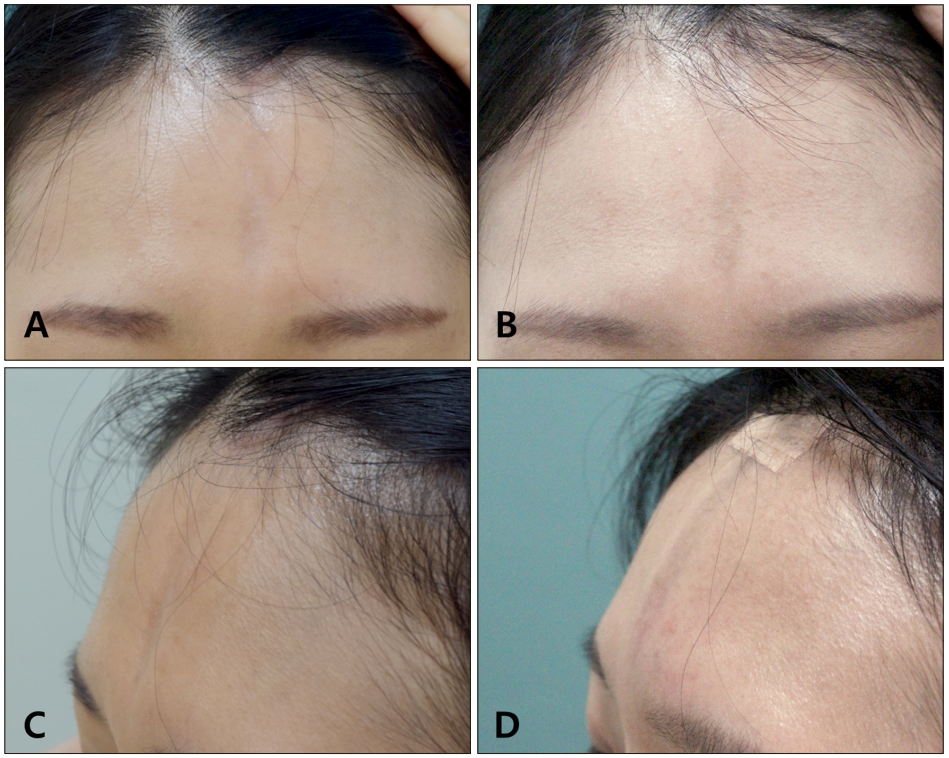

Fig. 1. (A) Linear vertical depression was present on the left medial forehead at the initial visit, (B) sustained improvement was achieved at 6 months post-injection. (C) Lateral view of patient at baseline, (D) 6 months post-injection.

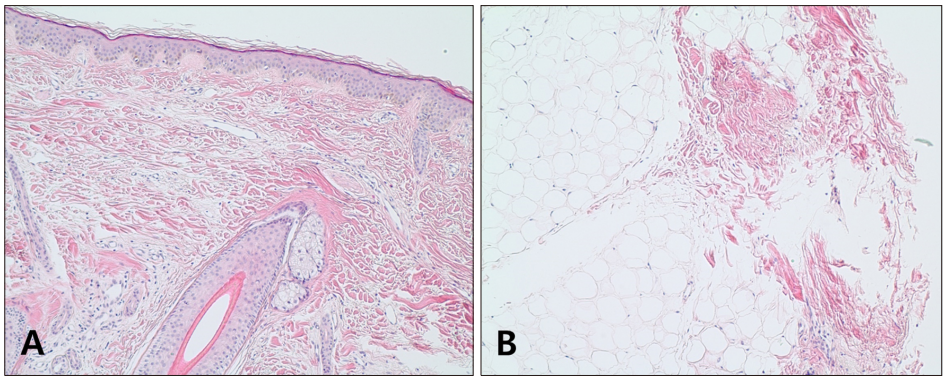

Fig. 2. (A) Irregularly thickened collagen bundles in the mid dermis and (B) subcutaneous fat. Note that inflammatory infiltrates were sparse, indicating the inactive phase of linear morphea (H&E, ×100).

ciated skull, ophthalmologic, or neurologic abnormalities, and she had normal laboratory values, including a negative antinuclear antibody screen result. With the initial diagnosis being the ECDS variant of LM, we prescribed oral prednisolone (5～10 mg/day) for 6 weeks followed by oral hydroxychloroquine (200 mg/day) for 6 months. After the medical treatments, the lesions remained stable without signs of inflammation or spreading. A skin biopsy of the forehead-scalp boarder revealed epidermal atrophy along with increased sclerosis of the dermis and subcutaneous fat, confirming the inactive phase of LM (Fig. 2).

Since the patient was concerned about the cosmetic disfiguration, we attempted volumetric correction of the depressions in the forehead region. With the patient under local anesthesia, we successfully injected 1 ml of a micronized acellular dermal matrix filler (Megafill®; L&C BIO, Seongnam, Korea) along two different planes: the periosteum and dermal-subcutaneous junction. Immediate improvement of the surface contour was noted, and no early or late complications developed postoperatively.

Six months following the injection, the depressed lesion was adequately augmented with a favorable aesthetic outcome (Fig. 1B, D). With the aid of a three-dimensional skin analysis camera system (Antera 3D; Miravex, Dublin, Ireland), we were able to perform a quantitative assessment of the skin defect. The depressed volume of the forehead remarkably decreased from 10.11 to 2.30 mm^3, which was about an 80% improvement (Fig. 3).

We received the patient's consent form about publishing all photographic materials.

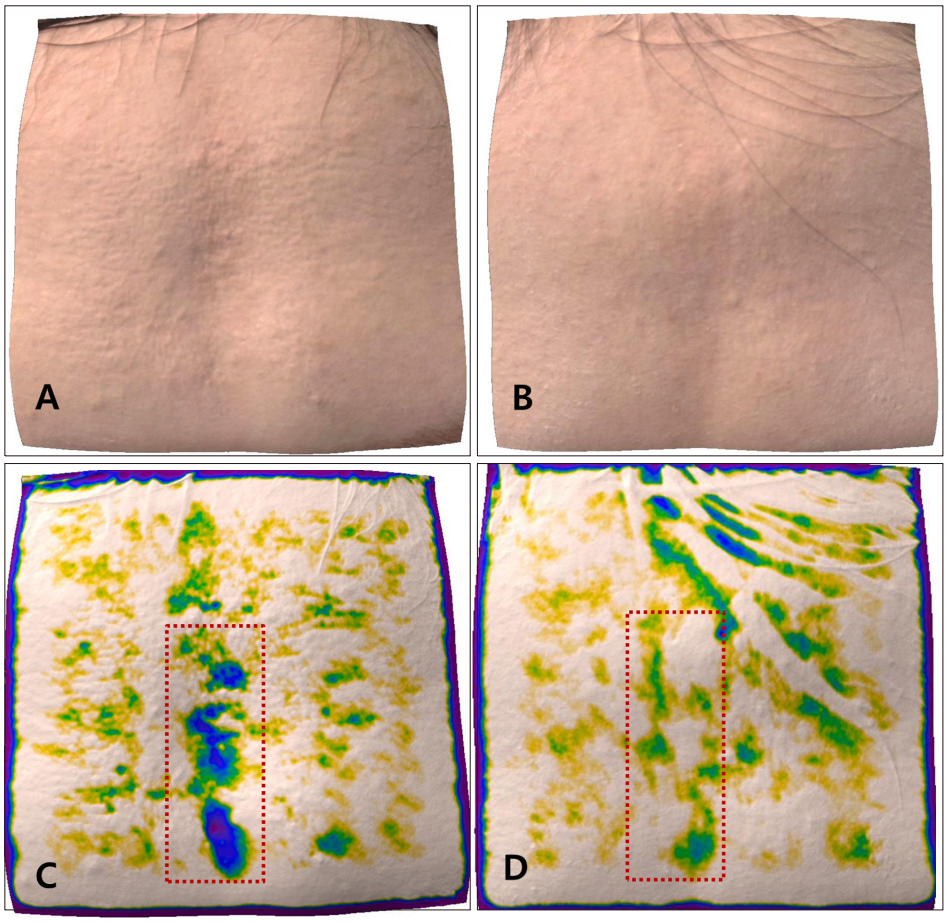

Fig. 3. (A) Baseline image was taken by the Antera camera (flat view) and (B) 6 months post-injection. The depressed volume within the area surrounded by the red dotted line was measured with depression mode in Antera camera (C) at the initial visit, (D) six months after the filler injection.

DISCUSSION

Soft tissue loss caused by the ECDS variant of LM results in disfiguring, permanent atrophy of the forehead and adjacent scalp. The lack of proper indicators of disease activity and prognosis cause difficulties in determining the optimal time for corrective procedures. Furthermore, even if the patient seems to be in a clinically quiescent disease phase, recurrence of the disease after corrective surgery is not uncommon[1]. Thus, treatment options with non-invasive and long-term maintenance effects are needed to minimize the risk of re-activation of LM by iatrogenic trauma.

Autologous fat grafting has long been used for soft tissue augmentation of facial LM, and favorable cosmetic results with a mean maintenance duration of 43 months were reported in the retrospective analysis by Roh et al[4]. However, they also noted that multiple treatment sessions were needed (2～11 sessions) owing to the high resorption rate of the injected fat. Besides, autologous fat grafting requires skilled practitioners to perform the procedure and a facility to store the extracted fat, which can cause increased medical costs.

Minimally invasive methods, such as hyaluronic acid (HA) filler, calcium hydroxyapatite filler, and poly-L-lactic acid filler, have been used in isolated case reports with varying degrees of success[3,5,6]. Among these options, the HA filler injection is considered to be the safest and relatively effective because of the stability of HA material *in vivo* and theoretical reversibility with hyaluronidase[5]. However, injection of HA filler into the frontal area could result in a persistently visible blue hue, which can cause cosmetic concern[7]. Additionally, although rarely reported, HA can result in foreign body granuloma formation, tissue necrosis by vessel compression, and allergic reaction[8].

The micronized acellular dermal matrix filler (Megafill®) is produced after pulverizing the cross-linked human acellular dermal matrix, and epidermal and dermal cellular components are removed to preclude immunologic reactions and rejection[9]. The manufacturing process involves the cross-linking of collagen by using electron beams, which gives augmented mechanical strength and structural integrity to the filler material. Moreover, implantation of acellular dermal matrix in an animal model revealed increased expressions of type I collagen, matrix metalloproteinases-1

(MMP-1), MMP-2, and transforming growth factor-ß for up to 6 months[10]. These molecular features result in enhanced durability and prolonged volume consistency of the micronized acellular dermal matrix filler not only by the volumizing effect but also by the long-term extracellular matrix remodeling effect. There was a case report of a LM patient who showed excellent cosmetic outcomes by acellular dermal matrix (AlloDerm®; LifeCell Corp., Branchburg, NJ, USA) implantation[11]. However, we expect that Megafill® filler has advantage over AlloDerm® because it is available in injectable formulation, allowing a minimally invasive technique.

In summary, we suggest that micronized acellular dermal matrix filler can be a less invasive and effective therapy for esthetic improvement of permanent atrophy caused by LM. Current attempts for cosmetic correction of atrophic lesions must be preceded by an evaluation of disease activity of LM, either by a sufficient clinical assessment or histologic evidence. During the treatments, the use of the image analyzer technique for objective measurement of the depressed volume is helpful for evaluating the treatment efficacy.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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REFERENCES

1. Christen-Zaech S, Hakim MD, Afsar FS, Paller AS. Pediatric morphea (localized scleroderma): review of 136 patients. J Am Acad Dermatol 2008;59:385-396.
2. Mazori DR, Wright NA, Patel M, Liu SW, Ramachandran SM, Franks AG Jr, et al. Characteristics and treatment of adult-onset linear morphea: a retrospective cohort study of 61 patients at 3 tertiary care centers. J Am Acad Dermatol 2016;74:577-579.
3. Thareja SK, Sadhwani D, Alan Fenske N. En coup de sabre morphea treated with hyaluronic acid filler. Report of a case and review of the literature. Int J Dermatol 2015;54:823-826.
4. Roh MR, Jung JY, Chung KY. Autologous fat transplantation for depressed linear scleroderma-induced facial atrophic scars. Dermatol Surg 2008;34:1659-1665.
5. Choksi AN, Orringer JS. Linear morphea-induced atrophy treated with hyaluronic acid filler injections. Dermatol Surg 2011;37:880-883.
6. Onesti MG, Troccola A, Scuderi N. Volumetric correction using poly-L-lactic acid in facial asymmetry: Parry Romberg syndrome and scleroderma. Dermatol Surg 2009;35:1368-1375.
7. Cho SB, Lee SJ, Kang JM, Kim YK, Ryu DJ, Lee JH. Effective treatment of a injected hyaluronic acid-induced Tyndall effect with a 1064-nm Q-switched Nd:YAG laser. Clin Exp Dermatol 2009;34:637-638.
8. Beasley KL, Weiss MA, Weiss RA. Hyaluronic acid fillers: a comprehensive review. Facial Plast Surg 2009;25:86-94.
9. Kim J, Lew DH, Roh TS, Lee WJ. Use of acellular allogenic dermal matrix (MegaDerm) in orbital wall reconstruction: a comparison with absorbable mesh plate and porous polyethylene. J Craniofac Surg 2017;28:e644-e649.
10. Lee JH, Kim HG, Lee WJ. Characterization and tissue incorporation of cross-linked human acellular dermal matrix. Biomaterials 2015;44:195-205.
11. Robitschek J, Wang D, Hall D. Treatment of linear scleroderma "en coup de sabre" with AlloDerm tissue matrix. Otolaryngol Head Neck Surg 2008;138:540-541.