Parry-Romberg Syndrome Augmented by Hyaluronic Acid Filler

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Parry Romberg Syndrome (PRS), also known as idiopathic progressive hemifacial atrophy, is a rare neurocutaneous disorder characterized by loss of skin and subcutaneous fat of face, muscles, and bones causing unilateral atrophy. Most patients require only soft tissue augmentation although syndrome has varying grades of severity. In the majority of reported cases, it has been treated with surgical flap or autologous fat transplantation. However, these treatments need complicated surgical skills which take a lot of time and cost. Herein we report the first case of PRS augmented by hyaluronic acid (HA) filler in a 42-year-old female patient to suggest that HA filler could be a safe, simple, and even rational economic alternative to surgical treatment. (Ann Dermatol 30(6) 704-707, 2018)

Keywords- Facial hemiatrophy, Filler, Hyaluronic acid

INTRODUCTION

Parry-Romberg Syndrome (PRS) was first described by Parry in 1825 and by Romberg in 1946.1 It is a rare neurocutaneous disorder that is characterized by loss of skin, subcutaneous fat, and muscles of the face resulting in unilateral atrophy. It can even involve bone, teeth, hair and eyes.2 Usually it starts in the second division of the trigeminal nerve, followed by the first and third division.3 Until now, the two prominent treatments for PRS were autologous fat graft and surgical flap.4-6 However, these treatments need complicated surgical skills which take a lot of time and cost. The objective of this study is to report that hyaluronic acid (HA) filler can be used as a safe, simple, and even rational economic alternative for the treatment of PRS.

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

CASE REPORT

A 42-year-old female patient came to dermatology clinic for the correction of facial hemiatrophy and hyperpigmentation on the right side of the face (Fig. 1A, 2A). She suffered from progressive symptoms since 20 years ago. She complained of intermittent unilateral migraine and trigeminal neuralgia, but there were no bony or vascular abnormalities on imaging study. She showed normal teeth arrangement and visual acuity, and had no other systemic or congenital disorder. She had been relieving her neurologic pain with painkiller for over ten years. She also received Nd-Yag laser toning (1064 nm, 1.5 J/cm²) for 8 times to treat hyperpigmented lesion. However, she did not receive surgical flap or fat graft for facial atrophy.

We choose a mixed form of filler (Restylane SubQ® [Q-Med, Uppsala, Sweden]: Restylane Vital® [Q-Med] = 2:1), composed of HA, for soft tissue augmentation. And we made two puncture sites with 22 gauge needle on the right cheek and mandibular area. After local anesthesia,
Fig. 1. Frontal views of the patient. At first visit, the patient shows overall atrophy of subcutaneous fat and hyperpigmentation on the right face (A). Immediately after injection by fanning technique. Total 7 ml of filler was injected (B). One month after the first filler injection, additional 1.5 ml hyaluronic acid (HA) filler was injected on the chin (C). One year after first injection, additional 3 ml HA filler was injected on the chin (D).

Fig. 2. Lateral views of the patient. At first visit, we planned to inject fillers in the direction of the arrows with fanning technique (A). After subcision with the cannula, total 7 ml of filler was injected via two holes on the cheek and chin (B). One month after the first filler injection (C). One year after first injection (D).

HA Filler was injected via 23 gauge cannula pulling the needle back. A total of 7 ml of filler was used: 4 ml of HA filler was injected into the cheek, and the remaining 3 ml was injected into the mandibular area (Fig. 1B, 2B). Because the adhesion between facial skin and subcutaneous fat was too tight to inject HA filler, we made a split space by subcision with a cannula before HA filler injection. A part of depressed area would need to be recontoured by second filler injection, however the patient was satisfied with cosmetic result and did not want to undergo another painful subcision. So we finished injecting further HA filler. It only took about 30 minutes for the procedure overall. One month after the first injection, additional 1.5 ml of HA filler was injected using fanning technique on the chin (Fig. 1C, 2C). After 1 year later, additional therapy was done with 3 ml of HA filler, significant improvement of contour of left chin was seen (Fig. 1D, 2D). Two months later, she did not show any signs of atrophy, and she was satisfied with the procedure. Also, there were no adverse events such as bruising, tenderness, overcorrection, chewing discomfort, Tyndall effect, the occlusion of the vessel or foreign body reaction.

DISCUSSION

PRS or idiopathic hemifacial atrophy could be an extreme...
form of localized scleroderma. PRS usually occurs on the trigeminal area with hyperpigmentation. However, because the PRS is rare, the etiology and pathogenesis are poorly understood. Besides, the patho-physiologic relevance between PRS and localized scleroderma remains unclear. However, Tollefson and Witman reported that 28% of 54 patients with en coup de sabre and PRS have both diseases simultaneously. Prevalence of seizure and epilepsy are especially high in both. This significant overlap may explain the similarity of neurologic mechanism in scleroderma and PRS.

There are numerous PRS treatment options available from medical treatments to surgical techniques. However, medications such as topical steroid or calcipotriol, oral steroid, methotrexate, D-penicillamine and antimalarial drug are effective only at the progressive phase and may not be adequate to correct depression. Most patients require only soft tissue augmentation although syndrome has varying grades of severity. In the majority of reported cases, PRS has been treated with surgical flap or autologous fat transplantation. However, there is no established surgical technique for treatment of PRS among free flap, latisimus dorsi flap, anteriolateral thigh adipofascial flap, pedicle flap, liposuction and fat graft. It would depend on the experience and preference of the physician. Furthermore, these treatments need complicated surgical skills, lots of instruments, several hours of surgery and long time for recovery. The physician should also consider the possibility of surgical complications such as graft failure, hematomata, infection, scar and the quality of life after flap or graft. Moreover, our patient was too thin (body weight, 45 kg) to obtain autologous donor fat. If the patient also had severe bony abnormality, we would definitely have considered surgical flap including bone augmentation and multiple stage treatment. But the patient has no bony abnormality, thus finding the simplest and safest method is best for her.

These days, numerous fillers are widely used in cosmetic fields such as the correction of nasolabial fold or tear trough, rhinoplasty, forehead augmentation and other depressed scar. These fillers can also be applied for depression of PRS. Until now, two PRS cases treated by fillers have been reported and one was with calcium hydroxylapatite (CaHA) and the other was with permanent polyacrylamide hydrogel (PAAG) filler. CaHA has a dramatic volumizing effect. After injection, the gel carrier is promptly spread out into adjacent tissue, and the remaining CaHA stimulates secondary collagen generation. A nonabsorbable volumetric PAAG filler is composed of 97.5% water and 2.5% cross-linked PAAG. Like CaHA, PAAG induces formation of internal scaffold vessel bearing connective tissue. It has longer lasting effect than HA. However, if the complications like unintended re-contour or compression of adjacent vessel or nerve occur, there are no eliminators available for CaHA or PAAG whereas hyaluronidase can be used to break down HA in case of side effects.

HA product is an absorbable, non-permanent filler and have replaced other implant materials as the standard filler. Because it shows a good safety profile and can be hydrolyzed by hyaluronidase, it had been used for facial fat atrophy in HIV-positive patients. HA has identical chemical composition in all species and tissues. Furthermore Restylane® (Q-Med) is an non-animal stabilized hyaluronic acid produced by fermentation of streptococcal bacteria. Thus it has less antigenic effect and is thought to induce less hypersensitivity reaction than animal source products. Because Restylane SubQ® (Q-Med) has larger gel particle size (approximately 1,000 particles/ml) than Restylane® (Q-Med), it is more suitable for contouring the cheeks and the chin. Also for the fine volumizing effect in upper dermal layer, we mixed Restylyne SubQ® (Q-Med) with Restylane Vital® (Q-Med). Generally, Restylane Vital® (Q-Med), which has the smallest particle size among all of the Restylane® (Q-Med) fillers, has been used for facial hydrofilling or hydrolifting in Korea.

In conclusion, HA filler has been widely used by dermatologists, which means that many dermatologists would prefer HA filler rather than flap, graft or other filler material. So, we suggest that HA filler will be the simplest and safest way for the correction of atrophy in PRS patient.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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