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SPECIAL TOPIC
Reconstructive

Dynamic Reconstruction Using Bilateral Lengthening Temporalis Myoplasty for Facial Palsies in Patients with Hereditary Skin Laxity

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Summary: Hereditary skin laxity is a rare condition, some cases of which are also referred to as cutis laxa, and those involving facial skin are considered a target for treatment by plastic surgery as patients present with an aged face, which can reduce their quality of life. In some of these patients, the facial nerve and muscles may be affected, and cause weakness of mimetic muscles. We performed one-stage bilateral lengthening temporalis myoplasty reanimation, followed by lower facial contouring with partial lower lip excision and hammock-shaped fascia grafting in two patients with hereditary facial skin laxity coexisting with facial palsy. The patient was a 63-year-old woman with hereditary gelsolin amyloidosis and a 64-year-old man who was diagnosed with oculopharyngeal muscular dystrophy. Postoperatively, a symmetrical facial contour was achieved in repose, and smiling with and without biting was possible. To our knowledge, there are no reports of dynamic smile reconstruction for facial weakness in patients with hereditary facial skin laxity. Although these patients may experience progressive loss of function of the trigeminal nerve and its innervating muscles, the static suspension effect of lengthening temporalis myoplasty can be expected to continue even if the temporal muscles lose their function in the future. We believe that, with careful patient selection, dynamic reconstruction is an option for progressive facial paralysis. In this article, we present the chronological history of two patients who underwent multiple plastic surgery procedures and discuss the importance of the role of plastic surgery in improving the quality of life under these conditions. *(Plast Reconstr Surg Glob Open 2024; 12:e5618; doi: 10.1097/GOX.0000000000005618; Published online 19 February 2024.)*

INTRODUCTION

Hereditary skin laxity is a rare condition, some cases of which are also referred to as cutis laxa, and those with facial skin laxity are considered a target for treatment by plastic surgery, such as blepharoplasty and face lift, because it presents with an aged face and can result in a reduction in quality of life.[1–6] In some of these patients, the facial nerve and facial muscles may be affected, and cause weakness in mimetic muscles.[7–10] Because neuropathy and muscle damage due to the original condition may progress, dynamic reconstruction is not recommended for facial palsy in these patients.[4] However, in cases where contraction of the temporalis muscle can be confirmed by occlusal movements, temporalis transfer surgery may not only improve the static balance of the midface, but may also achieve dynamic reconstruction of the smile. We performed one-stage bilateral lengthening temporalis myoplasty (LTM) reanimation, and secondarily lower facial contouring with partial lower lip excision and hammock-shaped fascia grafting in two patients with hereditary facial skin laxity coexisting with facial palsy. To our knowledge, there are no reports of dynamic smile reconstruction for facial weakness in patients with hereditary facial skin laxity. In this article, we present the chronological history of two patients who underwent multiple plastic surgery procedures and discuss the importance of

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Received for publication August 10, 2023; accepted January 9, 2024.

DOI: 10.1097/GOX.0000000000005618

Disclosure statements are at the end of this article, following the correspondence information.

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the role of plastic surgery in improving the quality of life under these conditions.

CASE REPORT

Case 1

A 63-year-old woman presented with bilateral facial nerve palsy, involuntary movements, and skin laxity that gradually progressed from her mid-50s (Fig. 1). She was also presented with symptoms of dysarthria, dysphagia, reduced chewing power, macroglossia, and bilateral blepharoptosis, and was diagnosed with hereditary gelsolin amyloidosis by genetic analysis at the age of 62 years (**See figure, Supplemental Digital Content 1,** which shows direct sequencing of a PCR-amplified region of genomic DNA spanning codon 187 of the gelsolin gene, showing a heterozygous c.654G>A mutation. **http://links.lww.com/PRSGO/D71**.) (**See table, Supplemental Digital Content 2,** which shows clinical and laboratory findings in the presenting cases. **http://links.lww.com/PRSGO/D72**.). Although the patient had no major problems in her daily life at home, such as eating and talking, when she went out, she found it difficult to eat and socialize with friends; so, she visited our department. The patient presented with an aged face with skin laxity on the mid and lower face, bilateral facial nerve impairment, and involuntary facial muscle movements, which greatly hindered communication during conversation and social activities. (**See Video 1 [online],** which shows preoperative facial expressions.) On preoperative examination, the musculature of the bilateral temporalis muscles and contractions during biting were palpable.

As a dynamic reconstruction for bilateral facial nerve palsy, modified LTM, using a lazy-S incision[11], was performed bilaterally in a single stage. Excess skin from the cheek to the mandible was excised simultaneously. (**See figure, Supplemental Digital Content 3,** which shows surgical procedures of bilateral modified LTM. **http://links.lww.com/PRSGO/D73**.) (**See Video 2 [online],** which shows 5 years postoperatively.) Postoperatively, a symmetrical upper and lower lip and mouth angle position has been achieved in repose, resulting in improvement of oral competence. Both smiling with biting and smile without biting are possible, and a balanced smile is achieved. This has led to a significant improvement in quality of life in everyday and social activities. However, the excess skin of the lower lip remained, and the symmetry was sometimes broken in the expression of saying "woo" (Fig. 2 and Video 2). Therefore, 5 years after bilateral LTM, the flaccid excess skin of the lower lip was excised, and a hammock-shaped fascia graft was placed over the bilateral masseter muscles (Fig. 3). After the surgery, the lower lip maintained moderate tension and good symmetry in repose. Good balance has been achieved in voluntary smiles associated with biting movements as well as in spontaneous smiles (Fig. 4). Fortunately, the progression of the primary disease was slow, and quality of life has improved dramatically by improving facial function and morphology through a series of operations. The patient also presented with blepharoptosis, but did not desire blepharoplasty, as she could achieve a double eyelid effect using eyelid tape.

Takeaways

Question: What are methods for performing hereditary facial skin laxity coexisting with facial palsy, which was difficult to cure?

Findings: We performed one-stage bilateral lengthening temporalis myoplasty reanimation and lower facial contouring with partial lower lip excision and hammock-shaped fascia grafting in two patients with cutis laxa combined with facial palsy.

Meaning: Plastic surgery for skin laxity and aging of the face with gelsolin amyloidosis or oculopharyngeal muscular dystrophy can be useful, and dynamic smile reconstruction with lengthening temporalis myoplasty should also be considered as an option when complicated by facial paralysis.

Case 2

A 64-year-old man, who was diagnosed with oculopharyngeal muscular dystrophy (OPMD) by a neurologist at our institution (Supplemental Digital Content 2; Table S1), had slow progressive blepharoptosis and dysphagia symptoms as well as facial skin laxity from 50 years of age, and underwent blepharoplasty at the age of 56 years by his previous doctor. By the time of his first visit to our department at the age of 64 years, bilateral facial paralysis had developed, and he was hardly able to make facial expressions, including smile and forehead wrinkling. At this time, the patient underwent bilateral ptosis correction by resection of the levator palpebrae aponeurosis and right brow elevation. However, facial laxity and drooping of the lower face had progressed further, so he requested facial reanimation and contouring at the age of 69 (Fig. 5).

Clinical examination revealed a normal bulk of the bilateral temporalis muscles and muscle contractions during biting. Therefore, bilateral modified LTM was performed, and excess skin from the cheek to the mandible was excised in a single stage. After surgery, restoration of resting commissural symmetry can be achieved, and oral competence can be improved. Drooping of the cheek and lower lip have been corrected, and creation of a balanced smile was possible (Fig. 6). However, because the lower lip ectropion remained, 1 year and 10 months later, further partial lower lip excision and hammock-shaped fascia grafting were performed. This enabled a tighter lower lip to be obtained (Fig. 7). Although there has been a slow progression of dysphagia due to the original disease since the mid-60s, the patient's quality of life has improved, and his attitude toward rehabilitation has become more positive as a result of the improvement in his facial function and morphology through a series of operations.

DISCUSSION

Hereditary skin laxity, some cases of which are also referred to as cutis laxa, is a rare elastolysis disorder,

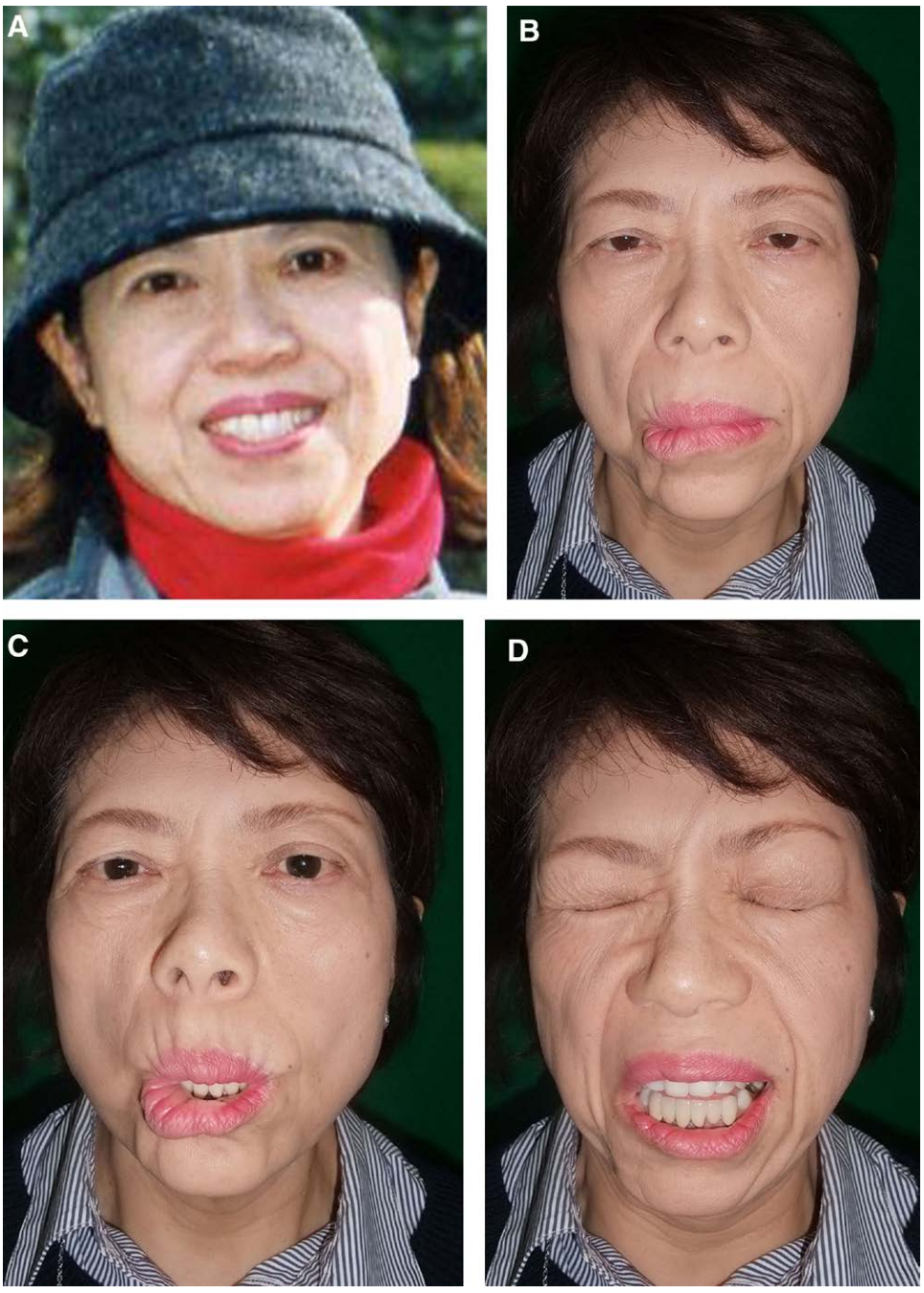

Fig. 1. A 63-year-old woman with bilateral facial nerve palsy, involuntary movements, and skin laxity that gradually progressed from her mid-50s, who was diagnosed with hereditary gelsolin amyloidosis by genetic analysis at 62 years of age. A, Before onset of the disease at 54 years of age. B, Preoperative view at 63 years of age in repose. C, Saying "woo." D, On smiling.

characterized by loose skin that hangs in folds. The congenital form of cutis laxa may be autosomal dominant, which is a benign form with less severely compromised internal organs. Patients with cutis laxa can expect a normal course of healing with no hyperelasticity syndromes; therefore, they are good candidates for surgical procedures.[1–3]

Gelsolin amyloidosis, or familial amyloid polyneuropathy type IV, is one of the diseases presenting with cutis laxa and is a dominantly inherited syndrome in which systemic amyloid deposition leads to early aging, bilateral progressive facial paralysis, and corneal lattice dystrophy.[12] The first symptoms of this progressive disease usually emerge during the third decade of life. In addition to the evident degradation of facial expressions due to facial paralysis and looseness of the skin, common complaints include dryness and ulcers of the eyes, hair

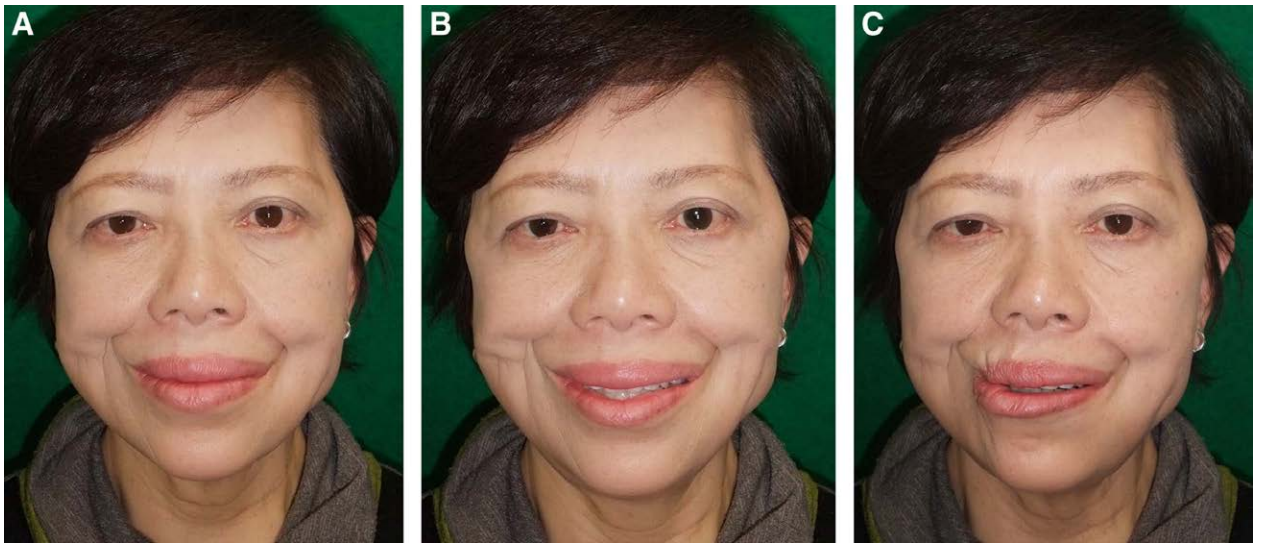

Fig. 2. Five years postoperatively. A, In repose, symmetrical upper and lower lip and mouth angle position has been achieved. B, Smile without biting is possible and a balanced smile is achieved. This has led to a significant improvement in quality of life in everyday and social activities. C, Excess skin of the lower lip has remained, and the symmetry was sometimes broken when saying "woo."

loss, dry itching skin, and diffuse functional gastrointestinal symptoms.[13] The symptoms of facial paralysis progress from the frontal facial nerve branches to the lower branches over the course of several years or decades.[4] A report analyzing the clinical symptoms of 29 patients with gelsolin amyloidosis revealed that involvement of the frontal, zygomatic, and lower buccal branches was 97%, 83%, and 52%, respectively, and bilateral paralysis was 72%, 14%, and 7%, respectively.[9] Clinically, patients have signs of involvement of other cranial nerves such as V, III, and XII.[4,9]

OPMD is a rare progressive, usually autosomal dominantly inherited, muscle disease that starts around the fifth decade of life[14,15] and is caused by an extended repeat mutation in polyadenylate binding protein nuclear 1 (PABPN1).[16,17] OPMD is characterized by blepharoptosis and dysphagia due to selective involvement of the muscles of the eyelids (levator palpebrae muscle) and pharynx, respectively. Two types of operations are used to correct the ptosis with overall good results: resection of the levator palpebral aponeurosis and frontal suspension of the eyelids.[8,16] In the present case, frontal suspension was predicted to be ineffective due to the dysfunction of the frontalis muscle, so correction of blepharoptosis with resection of the levator palpebral aponeurosis was performed. The upper and lower extremities can also be affected.[18] Other manifestations as the disease progresses include limitation of upward gaze, tongue atrophy and weakness, temporalis muscle atrophy, chewing difficulties, wet voice, and facial muscle weakness.[7,8,18]

As both gelsolin amyloidosis and OPMD progress slowly, plastic surgery, such as blepharoplasty and face lift, is recommended for ptosis and an aging face.[1–4,8] However, dynamic reconstruction is not recommended for facial palsy because neuropathy and muscle damage due to the original condition may progress[4] because preoperative bulging of the temporalis muscle and muscle contraction with occlusal movements were observed in both cases. Together with the fact that LTM requires less operative time than free muscle transfer because no microsurgery is required and that the temporalis muscle is not denervated in LTM,[11,19] reanimation by LTM was expected to be successful with high probability. In fact, both patients were able to achieve not only static facial balance improvement but also dynamic smile reconstruction, which was maintained for more than 4 years after surgery. The quality of life of the patients improved significantly after dynamic reconstruction for facial paralysis, and there were positive changes both mentally and behaviorally, such as greater social interactions with friends, active outings, and active engagement in rehabilitation for the primary disease. The patients also responded to subsequent improvement in lip morphology, which was further improved by partial excision of the lower lip and hammock fascia grafting. Moreover, reconstruction with LTM has also been reported to improve some aspects of swallowing function[20,21] and could provide supportive effects for oropharyngeal dysfunction associated with multiple neurological and muscular diseases.

To our knowledge, there are no reports of dynamic reconstruction of the smiles using bilateral LTM for facial weakness in patients with hereditary facial skin laxity. Rose et al reported a case of bilateral LTM dynamic reconstruction for immune dysregulation, polyendocrinopathy, enteropathy, X-linked (IPEX) syndrome, in which oral commissures were elevated at rest with improved oral competence, and indicated that with careful consideration and patient selection, successful surgical restoration of resting symmetry and dynamic commissural smile can be achieved in this complex cohort of patients.[22] Although both hereditary gelsolin amyloidosis and OPMD can lead to progressive loss of function of the trigeminal nerve and its innervating

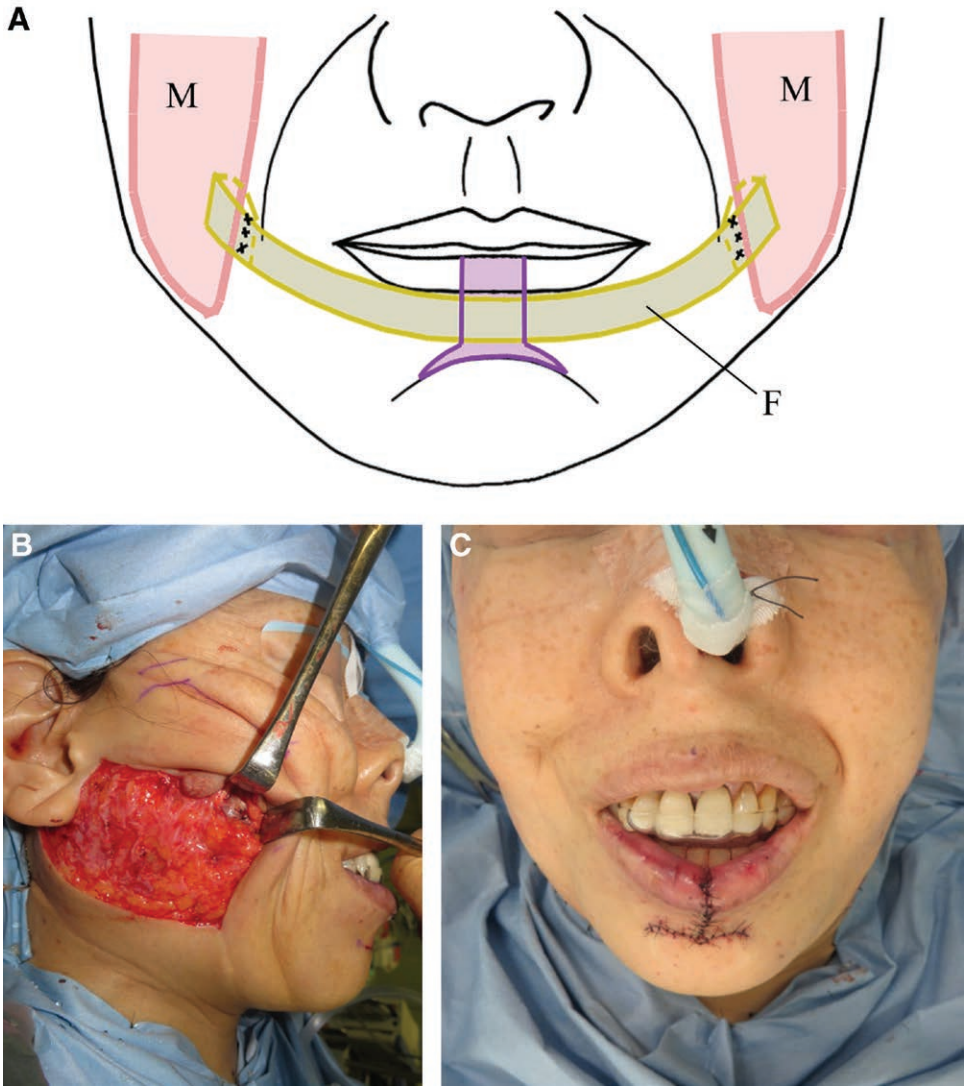

Fig. 3. Flaccid excess skin of the lower lip was excised, and a hammock-shaped fascia graft was also placed over the bilateral masseter muscles. A, Schema of the procedure. The lower lip was excised in all layers over a width of 1.5 cm in the midline (purple area). B, A slit was opened 1 cm posteriorly from the anterior margin of the masseter muscle, and a strip of fascia lata (1.5 × 22 cm) was passed under the lower lip skin and fixed through the slit bilaterally. C, Immediate postoperative frontal view; M, masseter muscle; F, fascia graft.

muscles, the static suspension effect of LTM can be expected to continue even if the temporal muscles lose their function in the future. We believe that, with careful patient selection, active dynamic reconstruction using LTM is an option for progressive facial paralysis.

Hummock-like fascia and tendon grafts to the lower lip have been reported to improve oral competence in cases of facial nerve palsy and to maintain the position of the skin flap after lower lip reconstruction.[23–27] In these methods, the fascia and tendons are suture-fixed to the zygomatic arch, deep temporal fascia, or hanging end of the temporalis flap. We fixed the ends of the fascia lata through slits in the masseter muscle, which is essentially the same fixation method as the muscle bow traction method for unilateral facial nerve palsy.[28] The hammock-like fascia fixed to the masseter muscles bilaterally is expected to be pulled to the left and right in biting, thereby increasing tension and lifting the lower lip. However, no obvious dynamic effect was observed in the cases we dealt with, and the effect was considered to be mainly static.

In the two cases presented here, lower lip tightening was performed secondarily at a later date because the patient first wanted to improve facial balance and dynamic reconstruction, and because it was difficult to predict how the results of dynamic reconstruction would affect the lower lip morphology. If the opportunity arises to treat similar cases in the future, dynamic reconstruction and lower lip tightening at the same time could be an option.

Fig. 4. Six months following revisional surgery of the lower lip (5 years and 7 months following bilateral lengthening temporalis myoplasty). A, The lower lip maintains moderate tension and good symmetry in repose. B, Good balance has been achieved in voluntary smile associated with biting movements.

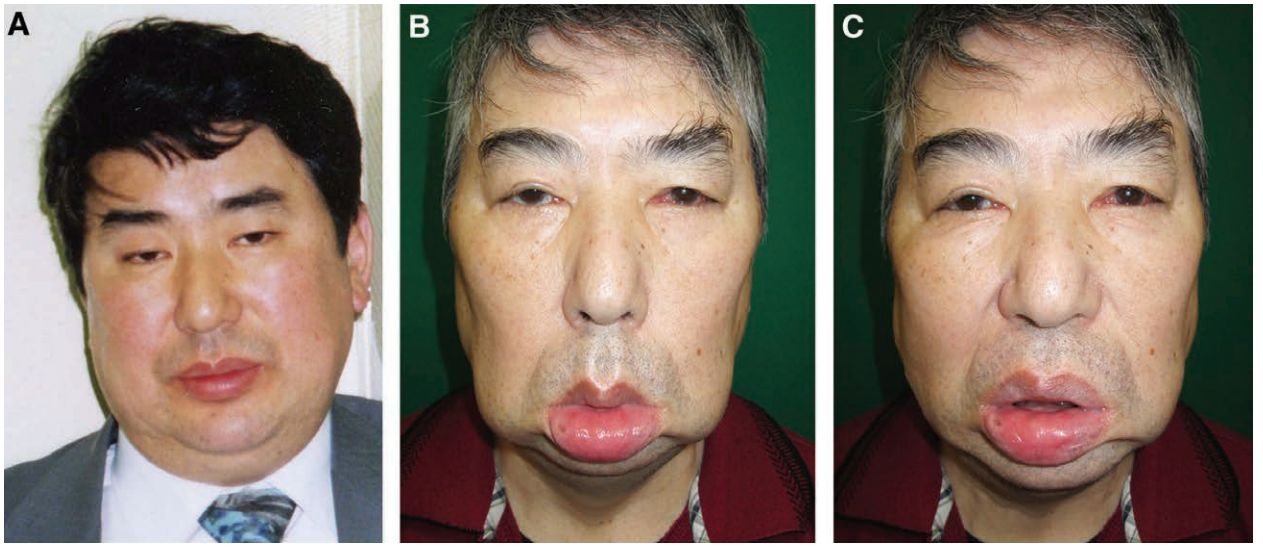

Fig. 5. 64-year-old man with oculopharyngeal muscular dystrophy. A, Before onset of the disease at 46 years of age. B, C, At the age of 69 before lengthening temporalis myoplasty, in repose and voluntary smile, respectively.

CONCLUSIONS

Plastic surgery for skin laxity and aging of the face with gelsolin amyloidosis or OPMD can be useful, and dynamic smile reconstruction with LTM should also be considered as an option when complicated by facial paralysis. In addition to other facial plastic surgery procedures, reanimation with LTM may further improve patients' social activity and quality of life.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

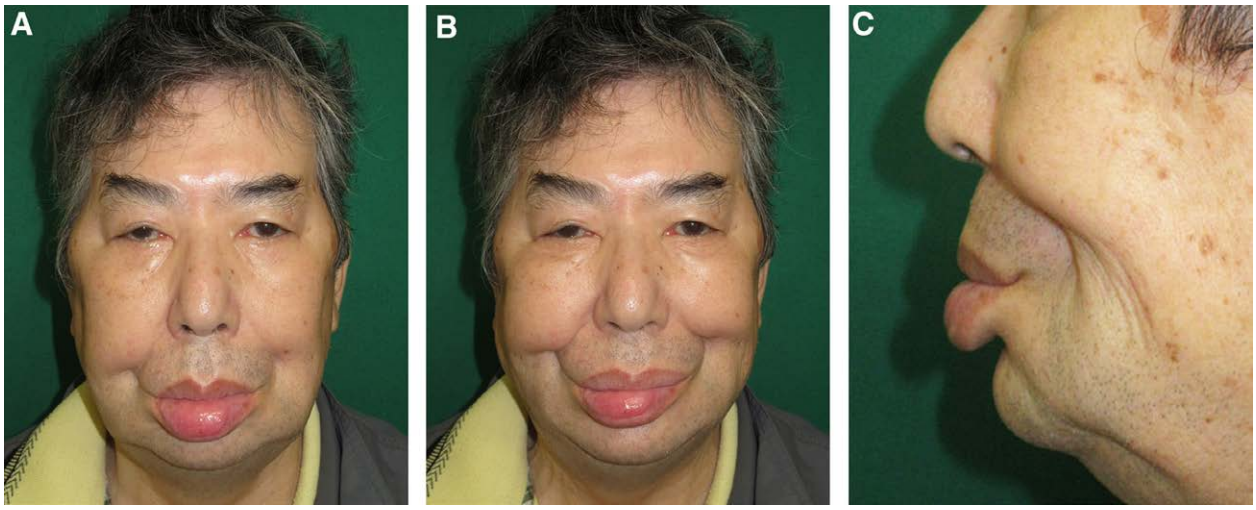

Fig. 6. Two years and 8 months after bilateral modified lengthening temporalis myoplasty. The drooping of the cheek and lower lip has been corrected. Although the patient is capable of generating a balanced smile, lower lip ectropion has remained. A, In repose. B, Voluntary smile with biting. C, Lateral view in repose.

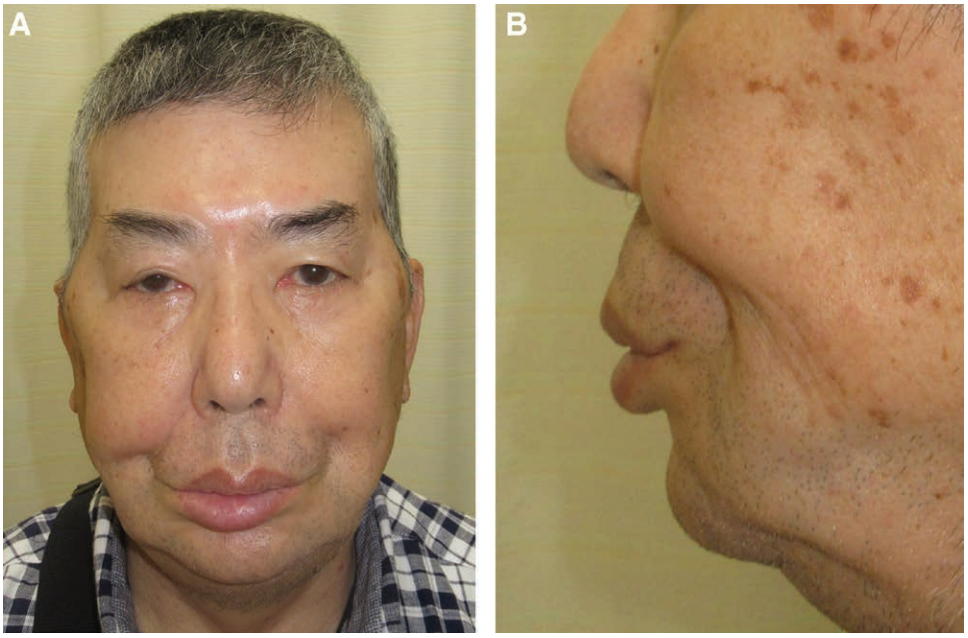

Fig. 7. One year and 10 months following revisional surgery of the lower lip (4 years and 6 months following bilateral lengthening temporalis myoplasty). A, The lower lip maintains moderate tension and good symmetry in repose. B, Lateral view in repose.

ETHICAL APPROVAL

The study was approved by the Ethics Committee of Toho University Sakura Medical Center (Permission no.: S16058).

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