INTRODUCTION

First described by Meckel in 1822,1 congenital hemifacial myohyperplasia (HMH), also known as hemihyperplasia or hemimacrosomia, is a rare condition of unknown etiology, uniformly identified by the presence of one or more facial organs displaying progressive hyperplasia. Due to the amorphous definition of the condition, it may cover a wide spectrum of distinct etiologies. Nonetheless, classification based on the number and types of tissues involved offers some structure for diagnosis and treatment.2,3 True or total HMH is characterized by unilateral enlargement of all tissues on one side of the face (ie, skin, bone, nervous, vascular, and fat). Partial or limited HMH is characterized by enlargement of one or more but not all organ systems of the face. The vast majority of cases of HMH described in the literature are classified as “true” or “total” hyperplasia.4,5

While the etiology, rate of recurrence in subsequent offspring, and pathogenesis of HMH remain unknown,6–9 Pereira-Perdomo et al10 postulated that this condition arises during the fourth week of gestation. Further classification of the condition applies to the implied clinical significance: (1) combined involvement of the muscles of facial expression and the muscles of mastication (derived from the first pharyngeal arch); and (2) isolated involvement of the muscles of facial expression (derived from the second pharyngeal arch). Facial asymmetry becomes more pronounced as development progresses, and treatment to guide tissue growth is necessary to improve both functional and aesthetic outcomes.

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Summary: Hemifacial myohyperplasia (HMH) is a rare congenital disorder characterized by the unilateral enlargement of facial muscles and unilateral hypoplasia of the skeletal structures. The causes, risk of recurrence in subsequent offspring, and pathogenesis of HMH remain unclear, and the condition can involve a number of features. Among them are pronounced facial asymmetry and changes to both hard and soft tissue structures, as well as facial hemiparesis. We describe the long-term surgical management of HMH in a 3-year-old girl who presented to our department with no other systemic manifestation. We describe the treatment options as well as our approach, which included the use of botulinum toxin injections, and our considerations when choosing to employ careful preservation of the facial nerve and facial mimetic muscles (rather than facial paralysis and facial reanimation) in 2 stages using muscle and nerve grafts. We found that sequential debulking procedures undertaken at significant intervals have offered our patient improved aesthetic and functional results in comparison with the use of nonsurgical techniques, comparable to the more complex grafting technique used in facial reanimation surgery. Due, in part, to the rarity of HMH, there is currently no consensus regarding the optimal treatment approach to the condition. Our use of serial debulking rather than the more complex and problematic microsurgical approach of facial reanimation surgery offers a feasible surgical solution with both aesthetic and functional improvement for these patients. (Plast Reconstr Surg Glob Open 2020;8:e2724; doi: 10.1097/GOX.0000000000002724; Published online 21 July 2020.)
CASE REPORT

A 3-year-old girl was referred to the Department of Reconstructive and Plastic Surgery at the Sourasky Medical Center (Tel Aviv, Israel) for a marked left-sided facial asymmetry and spasm. Her past medical history was significant for congenital HMH. The patient was the fourth child of young, healthy, and non-consanguineous parents with no notable history of teratogenic exposure. Pregnancy and delivery were uneventful. Left palpebral ptosis and muscular hyperplasia were noted immediately at birth. Anthropometric measures and psychomotor development have been normal.

Physical Examination

Upon physical examination, gross asymmetry and spasm of the left face was apparent. Skin dimpling was evident over the left facial muscles, namely perioral (depressor labii inferioris, depressor anguli oris, orbicularis oris), cheek (major and minor zygomaticus, masseter), and periocular (orbicularis oculi). Enophthalmos of the left eye was present with a substantially diminished palpebral fissure. The left lateral canthus was shifted inferiorly with minimal narrowing of the palpebral fissure. The left brow was considerably ptotic. The left nasal vestibule, the left pinna, and external auditory canal were smaller than the right. The left malar process of the maxilla was flattened. There was posterior displacement of the left auricle. The chin deviated to the left. Mandibular hypoplasia was evident on the left side. Cranial nerve examination was normal. The remainder of the physical examination was unremarkable.

Imaging

A craniofacial magnetic resonance imaging (January 2015) revealed a mass—partly linear and partly spindle-like in shape—starting at the level of the left temporalis muscle, anterior to the left zygoma, occupying the left malar area of the maxilla with a constituent extending into the naso-orbital fold (Fig. 1). The mass, muscular hyperplasia continued inferiorly along the external aspect of the jaw with a clear spindle-like segment anterior to the mandibular body, surrounding it inferiorly and protruding into the submandibular space. Left facial skeleton deformation was evident, namely (1) thinning of the mandible; (2) thinning and minor shortening of the ramus and condyle; and (3) underdevelopment and dysmorphism of the zygoma. Vascular malformation and plexiform neurofibroma were ruled out.

Management

The patient began receiving botulinum toxin injections at the age of 3 years roughly once every 6 months until the age of 7. The patient’s response was suboptimal; however, the decision was made to delay surgical intervention until facial development progressed further (Figs. 2, 3) (see Video 1 [online], which displays the preoperative stage of the patient: facial muscle hyperactivity, blepharospasm and periauricular spasm, and platysma hypertrophy).

Eventually, the patient underwent 5 operations:

- On July 2015 (when the patient was 8 years old), we performed selective partial paralysis of left facial nerve branch and partial debulking of the platysma to correct facial spasm.
- In December 2015 (at age 8 years), we performed further debulking of the left lower facial muscles due to blepharospasm and hyperactivity of the left cheek muscles, which caused severe functional limitations.
- In December 2016 (at age 9 years), the patient underwent further debulking of the left facial muscles due to refractory blepharospasm and hyperactivity of the left cheek muscles. Specifically, aberrant muscle tissue was debulked from the cheek area up to the midline (Fig. 4), nose dorsum, superior frontolateral facial area, orbicularis oculi, upper and lower eyelid, and postauricular area.

Fig. 1. Preoperative magnetic resonance imaging: left muscular hyperplasia.
In October 2017 (at age 10 years), despite significant improvement in relaxation of the facial spasm, further debulking was necessary. The operation included debulking of refractory hypertropic muscles at the cheek, nostril, lower eyelid, chin, and auricular muscles. In addition, we shortened the upper eyelid muscles to increase the eye aperture.

In July 2018 (at age 11 years), additional improvements were deemed necessary. The operation included further debulking of aberrant muscles in the left retroauricular area, as well as the left nasolabial fold and nostril. Left otoplasty was performed to improve auricular symmetry. Myotomy of the right depressors and injection of 13 ml fat into the left cheek were performed as well for improved contouring (Fig. 5) (see Video 2 [online], which displays postoperative results of the patient).

**DISCUSSION**

HMH is a condition that creates both functional and aesthetic challenges for the patient. Treatment strategies to date have been limited to nonsurgical reconstruction with minimal functional improvement and surgical reconstruction, including facelifts, osteotomy, or orthognathic procedures. We offer a multifaceted approach to this complex condition with a surgical solution that offers both cosmetic correction and functional improvements in HMH patients.
with a prominent facial spasm. While the microsurgical approach of facial reanimation may be a promising route, resection of aberrant hypertrophic muscles in our patient allowed for a simpler surgical approach with comparable results, without induction of total facial paralysis or functional nerve injury.14

CONCLUSION

HMH is a complex congenital malformation that may be treated using maximal resection of the hypertrophic muscles with careful preservation of the facial nerve and healthy muscles with excellent functional and aesthetic results.

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PATIENT CONSENT

The patient provided written consent for the use of her image.

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