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Möbius Syndrome: Surgical Treatment for Eyelid Dysfunction

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Key Words
Möbius syndrome · Eyelid dysfunction · Inferior recurrent keratitis · Scleral show

Abstract
Introduction: Möbius syndrome is a heterogeneous congenital disorder that is linked to bilateral palsies of the cranial nerves VI and VII, resulting in congenital facial paralysis sometimes associated with impaired ocular abduction. ***Case Report:*** We present the case of a 44-year-old woman with Möbius syndrome and inferior recurrent keratitis secondary to scleral show in both eyes. We decided to use a cartilage graft from the ear in the inferior eyelid to avoid eyelid retraction and scleral show. ***Discussion:*** Patients with Möbius syndrome have a severe dysfunction of their facial mimic. Their treatment must be individualized, depending on their age, clinical examination and symptoms.

Introduction

Möbius syndrome is a heterogeneous congenital disorder that is linked to bilateral palsies of the cranial nerves VI and VII [1, 2]. From an ophthalmological perspective, these patients present symmetrical facial diplegia with variable patterns of horizontal gaze paresis and facial paralysis [3, 4], which affect palpebral dynamics in adult life.

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Case Report

We present the case of a 44-year-old woman with Möbius syndrome and congenital craniofacial cleft. During her childhood, she had undergone multiple surgeries in the Departments of Traumatology, Plastic Surgery and Maxillofacial Surgery.

The patient was being monitored due to her eye problems derived from the facial paralysis. Four years ago, she had undergone a permanent partial bilateral tarsorrhaphy that improved her eyelid closure problems. However, at the last checkups, the lower eyelids had retracted again, and she presented recurrent lower keratitis caused by a scleral exposure of 1–2 mm in both eyes, in spite of a good Bell's phenomenon and an intensive treatment with lubricants (fig. 1).

We decided to introduce a graft for the lower eyelid in order to alleviate the retraction and reduce the scleral exposure. In view of the fact that the patient had undergone surgery of the palatal area on several occasions due to her craniofacial malformations (fig. 2), we used an auricular cartilage instead of a hard palate.

We performed an incision at the end of the lower conjunctival sac, we separated the retractor muscles from the tarsal plate, and we disinserted the capsulopalpebral ligament, so that the tarsal plate was left free and completely exposed. A 6 × 20 mm section of auricular cartilage was extracted and its upper border was sewn into the lower edge of the lower tarsal plate with 7-0 vicryl. The lower edge of the graft was sutured to the retractors, and the incision in the conjunctival sac was also sutured with 7-0 vicryl.

Results

Two weeks after the procedure, we observed that the lower eyelid retraction had improved notably. The patient did not present scleral exposure, and the eyelid closure had also improved (fig. 3).

Before this procedure, the patient presented recurrent type III keratitis despite intensive treatment with lubricants; however, after the graft had been inserted, we only observed type I keratitis sporadically and always when she had forgotten to apply tear supplements.

Discussion

Möbius syndrome is a rare complex of malformations which was first described by Von Graefe in 1880 [5]. Its etiology is unknown, although some components have been described, either genetic, vascular or related with the intake of some drugs like misoprostol during pregnancy. It is characterized by bilateral congenital paralysis of the cranial nerve pairs VI and VII, and it is accompanied by symmetrical facial diplegia with variable patterns of horizontal gaze paresis.

Facial paralysis and lagophthalmos have been documented in as many as 80% of cases with Möbius syndrome. During childhood, the phenomena derived from facial paralysis are not very noticeable. Lagophthalmos or other alterations of the eyelid dynamics are rare before 20 years of age, but facial amimia of the patients is common in the patients, especially in expressions like crying or smiles. In patients younger than 7 years of age, a certain function of the facial muscles can be achieved with the transference of the temporal muscle or the latissimus dorsi, or the gracilis muscle, which can be reinnervated with the mandibular branch of the trigeminal nerve or with a ramification of the spinal nerve [6–8].

However, patients are usually admitted as adults with no previous treatment, when the effects of facial paralysis are more evident. The main reasons for consultation are problems caused by the appearance of exposure keratitis due to poor eyelid closure during the night. In this case, the main therapeutic measure is a medical approach with lubricants and hygienic measures in order to prevent the evaporation of tears.

When the alterations in the eyelid dynamics increase, patients suffer a retraction of the lower eyelid with scleral exposure. This aggravates the recurrence and severity of keratitis, which in some cases causes extremely severe corneal ulcers that increase the risk of neurotrophy and can lead to corneal perforation and endophthalmitis. It is also common to see ectropion of the lower eyelid and eyebrow ptosis. In these cases, surgery is required, and the approach will depend on the severity of the facial paralysis, the exploration of the dynamic and static condition of the eyelids and the symptoms of the patient.

We can divide surgical treatments into static and dynamic approaches. Static approaches narrow the opening of the eyelid either horizontally (tarsorrhaphy) or vertically (elevation of the lower eyelid via desinsertion of the retractors, placing of spacers and/or a facial lifting). Dynamic approaches improve the eyelid closure, and the most common procedure is a gold weight implant [9]. To transfer a muscle, either vascularized or nonvascularized, such as the platysma muscle, towards the free edge of the eyelid and to combine it with a gold weight implant is usually enough for the correction of lagophthalmos [10].

Disclosure Statement

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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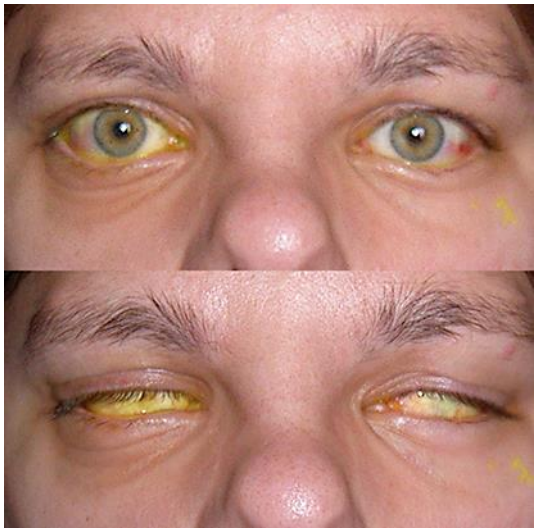

Fig. 1. Patient before surgery with 2 mm of sclera exposure.

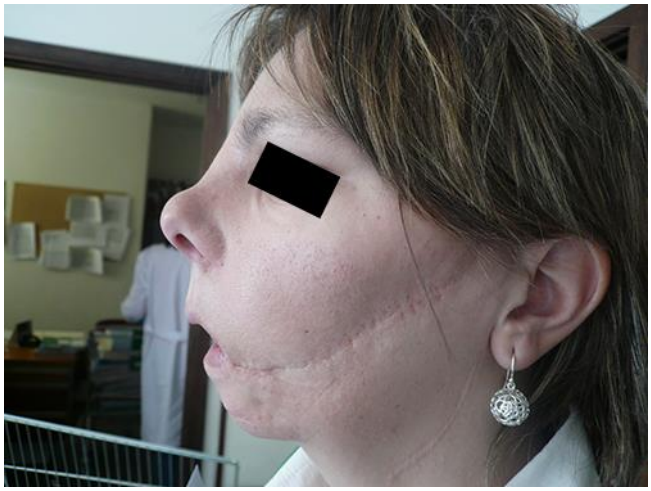

Fig. 2. Scars of previous maxillofacial surgeries.

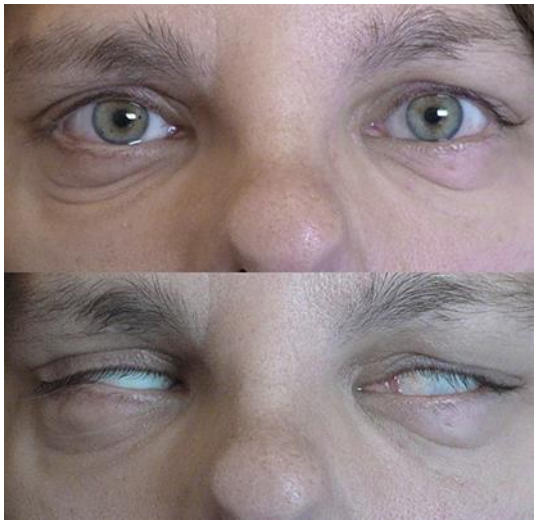

Fig. 3. Patient after ophthalmic surgery. Improvement in lower eyelid retraction.