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Case Report

Successful Outcome of Two-Stage Strabismus Surgery in a Rare Case of Unilateral Möbius Syndrome: A Case Report

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Abstract

Möbius syndrome (MBS) is a rare congenital disorder characterized by facial and abducens palsy which is usually bilateral and may involve multiple cranial nerves. Additional features, such as craniofacial anomalies, tongue malformations, and limb defects, may also be present. The etiology of this syndrome is not well established but the ischemic necrosis of the brainstem leading to hypoplasia of the cranial nerve nuclei is the supposed to be most likely one. MBS leads to a large angle esotropia along with bilateral limitation of abduction with delayed diagnosis, which is very challenging to correct with a single procedure. There are only a few reports illustrating the surgical outcome of strabismus in MBS. In this case report, we describe an unusual case of unilateral MBS who presented with large esotropia since childhood and was managed by two-stage strabismus surgery, including unilateral medial rectus (MR) muscle recession followed by vertical recti transposition. The final primary position alignment was satisfactory. Therefore, MR muscle recession alone is an effective first procedure of choice for mild-to-moderate esotropia in patients with MBS, whereas severe cases may further require the transposition of vertical recti. As it is a congenital condition, early surgical intervention may prevent amblyopia and provide an opportunity for the development of binocular single vision. Hence, prompt diagnosis and early surgical intervention are of utmost importance in these cases.

Keywords: Medial rectus recession, Möbius syndrome, strabismus surgery, vertical rectus transposition.

Introduction

Möbius syndrome (MBS) is a rare non-progressive neurological disorder, characterized by weakness or palsy of multiple cranial nerves, most often the abducens and facial nerves (1). It was first described by Von Graefe (1880) and later reviewed by Paul Julius Möbius (1888), a German neurologist after whom the syndrome was named (2).

The estimated prevalence of MBS is 1/250,000 live births having equal incidence in both sexes. There are about 300 cases reported in the literature, most of which are sporadic, with only 2% of all affected individuals being familial (2,3). The most common symptom of this syndrome is loss of facial expression. Ocular presentations include strabismus, restricted lateral eye movements, nystagmus, inability to close eyelids, conjunctivitis, and ptosis (4). In addition

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to these features, limb deformities (syndactyly, brachydactyly or absent digits, and talipes), orofacial anomalies (bifid uvula, micrognathia, and ear deformities), swallowing and speech difficulties, hypoplasia of tongue, musculoskeletal system abnormalities (rib defects and absence of pectoralis major muscle), and mental retardation may also be presented (5).

In 2007, diagnostic criteria were formulated by the International Group of Experts at the MS Foundation for diagnostic consistency, which included (1) Congenital facial diplegia or uniplegia of lower motor neuron (LMN) type, and (2) Paralysis of lateral movements of eyes and strabismus due to sixth cranial nerve palsy (1).

Although it is a congenital disorder having symptoms since birth, diagnosis is often delayed. These cases are often managed by a multidisciplinary approach, involving various specialists. Strabismus in MBS is characterized by large esotropia (of up to 100 prism dioptres [PD]) with medial rectus (MR) contracture and complete lateral rectus (LR) palsy poses a challenge for the strabismus surgeon. Numerous reports have outlined the various symptoms of MBS, but only a few have discussed the outcomes of strabismus surgery in these cases. We report a rare case of unilateral MBS in a 29-year-old male, with a favorable surgical outcome after two-staged strabismus surgery.

Case Report

A 29-year-old male patient presented with a constant inward deviation of his left eye (LE) since birth, along with a diminution of vision. There was no significant insult during birth and no history of delayed developmental milestones. The patient did not have any history of past ocular trauma, surgical intervention, or systemic illness. None of his family members had any similar complaints.

General and systemic examination revealed features of left-sided facial nerve palsy (LMN), which included the following: Absence of wrinkling on the left side of the forehead, lagophthalmos of LE, deviation of the mouth toward the right, and inability to puff cheek of the left side. Besides this, hypoplasia of the tongue was also noted (Fig. 1). The patient had an abnormal head posture with a head turn toward the left side. On ocular examination, the best corrected visual acuity in the right eye (RE) was 6/6 and LE was counting fingers at 4 m. There was an esotropia of 45° on the Hirschberg test with a -4 limitation of abduction, and inferior oblique overaction in LE (Fig. 2). The prism bar reflex test revealed a primary position deviation of 95 PD. Cycloplegic retinoscopy was performed under homatropine 2% at a working distance of one meter, and the best corrected visual acuity was 6/6 with −0.50 DS in RE and 6/60 with −7.50 DS/−1.00 DC @180° in LE. The Worth 4 dot

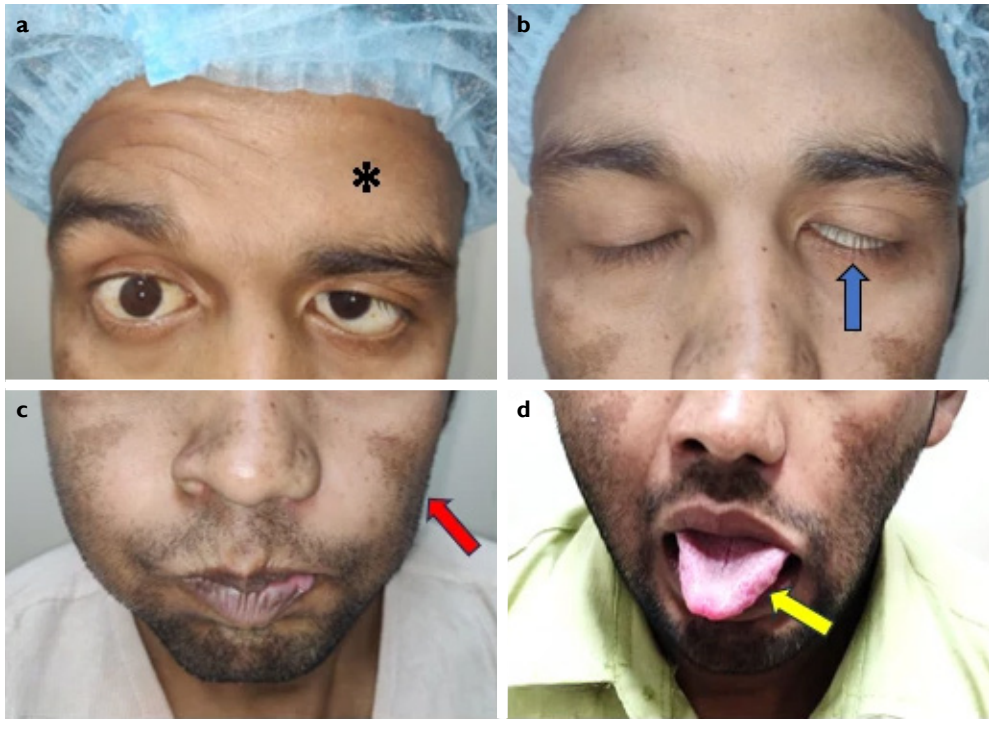

Figure 1. Clinical photographs showing features of 7th **(a-c)** and 12th **(d)** nerve palsy. **(a)** Absence of wrinkling on the left side of the forehead (black asterisk). **(b)** Lagophthalmos of the left eye (blue arrow). **(c)** Inability to puff the cheek of the left side (red arrow). **(d)** Tongue hypoplasia (yellow arrow).

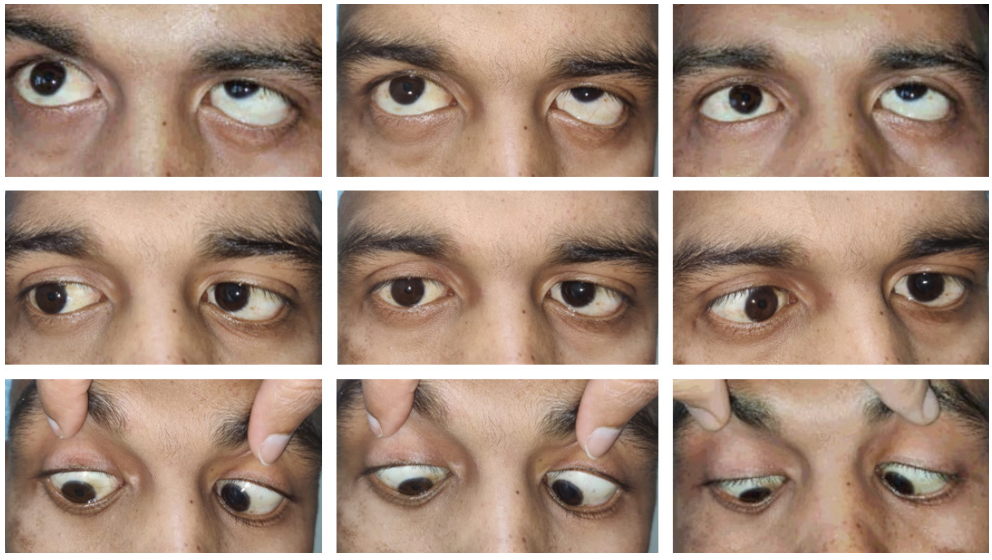

Figure 2. Clinical photographs of preoperative nine gaze showing limitation of abduction of the left eye, corresponding to left 6th nerve palsy.

test showed LE suppression and nil stereopsis on the Titmus fly test. Anterior segment and posterior segment examination were within normal limits. No other abnormality was detected on thorough systemic examination. Neuroimaging and genetic analysis could not be performed as the patient refused it due to financial constraints. Based on the above-mentioned history and examination, a working diagnosis of unilateral MBS on the left side was reached.

Forced duction testing was positive for LE MR muscle. The patient underwent a two-stage surgical approach. In the first sitting, LE MR was recessed to 7 mm with conjunctival recession. There was a residual deviation of 30 PD on the first post-operative day, with no significant improvement in LE abduction (Fig. 3). On further follow-up, the residual deviation increased to 50 PD with over-elevation in adduction. After 3 months of the first procedure, the patient was planned for LE full tendon vertical recti (superior and inferior recti) transposition by Hummelsheim technique due to large residual deviation. A limbal-based incision was made superiorly and inferiorly, and the superior and inferior recti were isolated and dissected. The recti were disinserted after securing with 6-0 vicryl sutures, transposed, and sutured to the sclera within 1 mm of the insertion of the LR following the spiral of Tillaux (6). The patient was orthophoric with marked improvement in LE abduction, and residual overelevation in adduction at 2nd

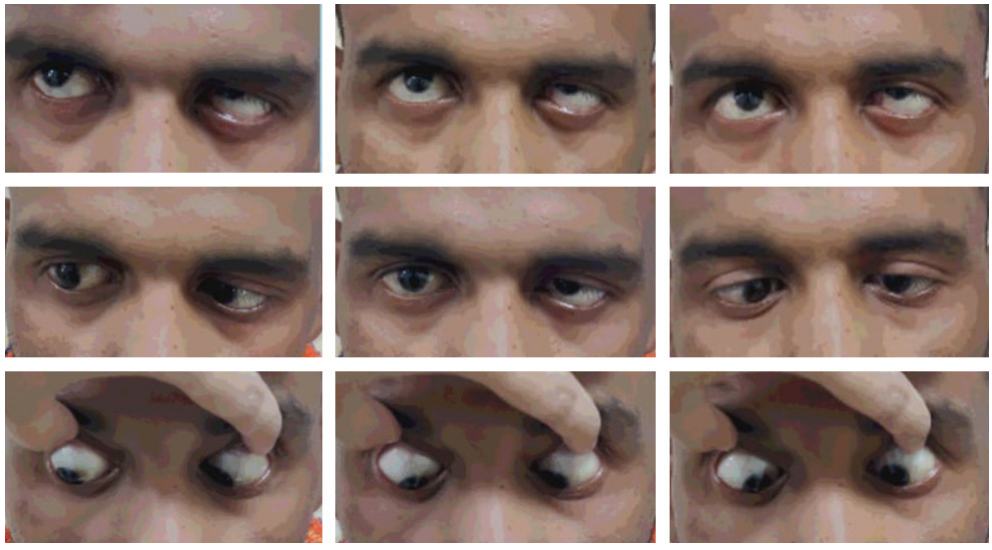

Figure 3. Clinical photographs showing post-operative nine gaze position after left eye medial rectus muscle recession along with the conjunctival recession. Residual esotropia of 30 prism dioptres and marked limitation of abduction in left eye were noted.

week post-operatively (Figs. 4 and 5). The satisfactory primary position alignment was maintained on subsequent follow-up visits for 2 years without any adverse outcome or any sign of symptom of anterior segment ischemia, but there was no improvement in the visual acuity of LE.

Discussion

MBS is a rarely reported congenital disorder of facial diplegia associated with lateral gaze palsy and is mostly bilateral. It may also involve other cranial nerve palsies, skeletal malformations, and craniofacial anomalies (1-5).

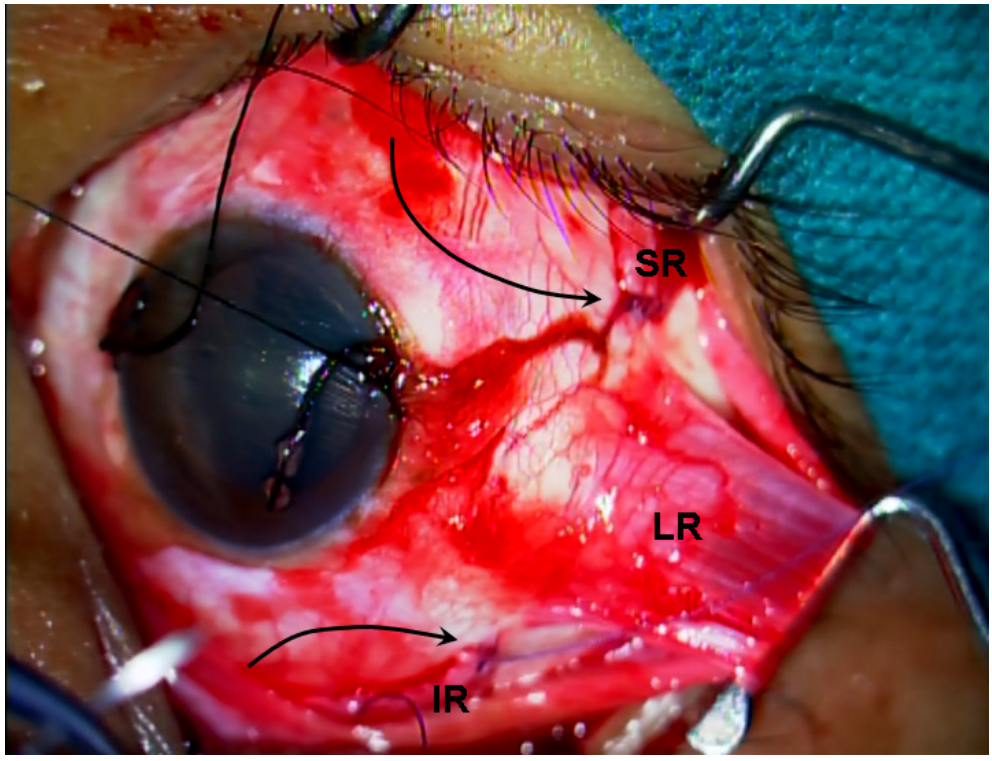

Figure 4. Intraoperative photograph of the second surgical procedure showing full tendon transposition of superior and inferior recti in left eye.

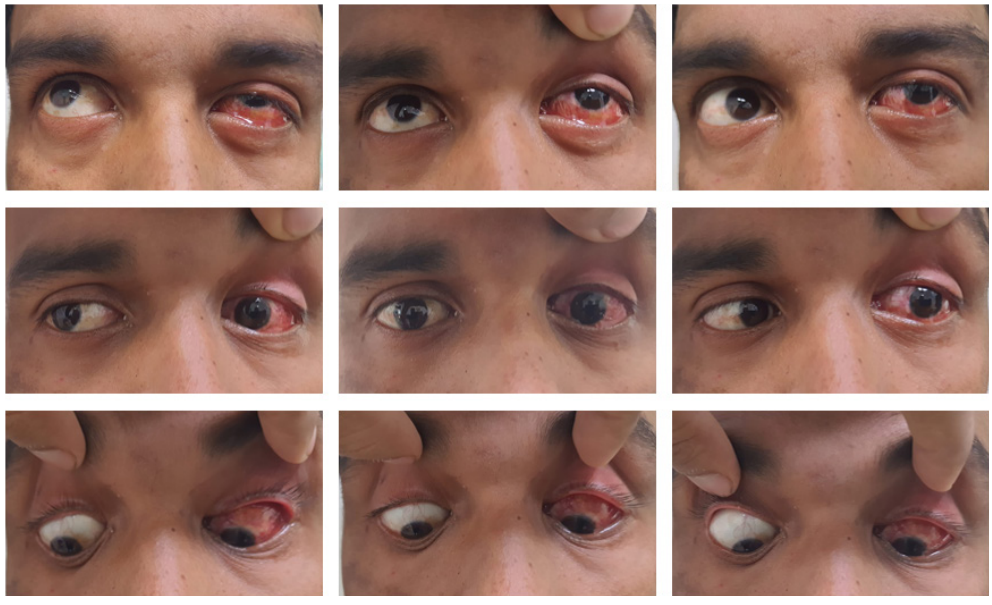

Figure 5. Clinical photographs showing post-operative nine gaze position at 2 weeks of post-operative day after full muscle vertical recti transposition. The patient was orthophoria and good improvement in left eye abduction was noted.

The etiopathogenesis of MBS appears to be multifactorial. Most authors believe that there is congenital hypoplasia of cranial nerve nuclei due to ischemia of the brainstem during fetal development due to any environmental, mechanical, or genetic cause (7-9). Exposure to infections, alcohol, cocaine, thalidomide, or misoprostol during pregnancy relates to a higher incidence of MBS (10). Another group consists of patients with hereditary congenital facial palsy in which agenesis or hypoplasia of the cranial nerve nuclei is pathologically presented and its penetrance is due to genetic or non-genetic factors. Most of the cases are sporadic in nature, but other modes of inheritance have also been described (4).

MBS presents more challenges to surgical correction of strabismus as these patients frequently have bilateral LR palsies along with MR contractures. Different surgical approaches have been described for treating esotropia in MBS. According to some authors, bilateral MR recession alone was insufficient to correct such a large angle esotropia, therefore combined MR recession and LR resection were recommended. In more severe cases, vertical muscle transposition was required for satisfactory results (11-13). Three patients having moderate-to-large angle esotropia (ranging from 40 to 80 PD) along with bilateral limitation in the abduction were reported by Sun and Gole, in which one patient underwent a vertical rectus muscle transposition with augmentation sutures alone, whereas two patients required two procedures, including MR recession and vertical rectus muscle transpositions with augmentation sutures. A satisfactory post-operative primary position alignment was seen in all three but there was no improvement in abduction limitation (11). Laby et al. described the surgical outcome in five patients with esotropia ranging from 30 PD to >80 PD. They underwent one or more procedures, including vertical rectus muscle transposition and MR weakening (recessions in four patients and botulinum toxin chemo denervation in one patient), in which four patients were overcorrected postoperatively and all four showed only a slight improvement in abduction (12). A case report by Zheng and Donahue showed good primary position alignment and mild improvement in abduction post-operatively in a patient with esotropia of 25 PD who underwent combined MR recession and superior rectus muscle transposition (13).

While some advocate that MR recession alone provides satisfactory primary position alignment postoperatively as seen in a report by Spierer and Barak, where the conventional approach of bilateral MR recession was found to be sufficient for primary motor alignment (14). According to Ventura et al., ten patients with esotropia of 15 to 85 PD were treated with MR recession only, out of which seven patients had ≤10 PD residual deviation even after 2 years of follow-up (15). Another study by Lueder and Galli, including five children having esotropia ranging from 20 to 30 PD, showed similar results after bilateral MR recession alone with primary position alignment <8 PD postoperatively (15). MR recession has some advantages as it is simpler than vertical recti transposition, has a lesser chance of causing vertical strabismus, and also has a lesser risk of anterior segment ischemial (16).

Our patient had a unique presentation of unilateral MBS which is even more unusual as most of the reported cases had bilateral involvement. The successful primary position alignment within 8 PD with good improvement in the abduction was achieved by a two-stage procedure without any improvement in visual acuity and binocularity. Moreover, large esotropia along with MR contracture is difficult to manage with a single procedure. Therefore, we opted for a two-stage surgical approach for our patient, including unilateral 7-mm MR recession followed by full muscle vertical recti transposition, and a favorable primary position alignment was achieved postoperatively and was maintained till 2 years of follow-up. Both the surgeon and the patient were satisfied with the final alignment.

Conclusion

MR recession alone may be a good first choice of surgery for mild-to-moderate esotropia in patients with MBS, but severe cases may further require the transposition of vertical rectus muscles to restore good primary position alignment. Furthermore, early diagnosis and timely intervention can prevent amblyopia and provide scope for binocular vision to develop.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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