

Congenital Bilateral Hypoplasia of Medial Rectus Muscle: A Case Report and Its Management

Mohammad Sharifi, MD¹ • Masoud Shafiee, MD²

Abstract

Purpose: To describe the clinical findings, preoperative radiologic findings and results of surgery in a patient with congenital bilateral hypoplasia of medial rectus muscle

Case report: A 50-year-old man presented with large angle incomitant horizontal deviation with marked deficit of adduction of both eyes. MRI finding defined very thin medial rectus muscle. Intraoperatively medial rectus muscles were not found. Lateral rectus recess combined with partial vertical rectus muscle tendon transposition was carried out on both eyes. The patient had orthotropia in primary position and adduction improved in final follow-up.

Conclusion: This case is the first case report of the Iranian population who had congenital hypoplasia of medial rectus muscle. MRI allowed effective surgical planning to correct congenital abnormality.

Keywords: Congenital, Hypoplasia, Medial Rectus Muscle

Iranian Journal of Ophthalmology 2014;26(2):108-10 © 2014 by the Iranian Society of Ophthalmology

Introduction

Ocular motility problems related to congenital abnormalities of the extraocular muscles are rare. Such abnormalities may present in a spectrum from accessory additional rectus muscles to the absence of extraocular muscles.¹

Congenital absence of rectus muscles are a very rare disorders usually seen in association with craniofacial syndromes. The range from hypoplasia to aplasia of rectus muscle and patients show large angle strabismus with limited ductions abnormalities. Radiologic studies including MRI must be performed before surgical plan. The surgical management of these disorders is difficult requiring various surgical procedures. Result of surgery is disappointing in most patients.^{2,3}

We report a rare case of congenital bilateral hypoplasia of medial rectus muscle in an adult man in which surgery had excellent results.

Case report

A fifty-year-old man was referred to strabismus clinic of our teaching hospital for squint since childhood. He had no history of trauma, medical disorders, previous surgery and family history of strabismus. He had large angle alternating exotropia about 100 prism diopter in primary position. Adduction was limited and eyes could not pass the midline (Figure 1). Other ductions were full. Visual acuity was $20/20$ in both eyes.

-
1. Assistant Professor of Ophthalmology, Eye Research Center, Mashhad University of Medical Sciences, Mashhad, Iran
 2. Resident in Ophthalmology, Eye Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

Received: February 15, 2014

Accepted: July 6, 2014

Correspondence to: Mohammad Sharifi, MD

Assistant Professor of Ophthalmology, Eye Research Center, Mashhad University of Medical Sciences, Mashhad, Iran

Email: sharifim@mums.ac.ir

Biomicroscopic and fundoscopic exam were unremarkable. At a planned recess/resect procedure for both eyes, patient had free forced duction test and 10 mm recess of lateral rectus muscle was performed for him. After opening of conjunctiva no medial rectus had been hooked and only fine insertion line was found after further evaluation (Figure 2) in both sides. Due to lack of radiologic study preoperatively we decide to perform orbital MRI postoperatively for rule out absence of

rectus muscle. After surgery patient had remained unchanged horizontal deviation.

Magnetic resonance imaging showed presence of medial rectus posteriorly but it was thin in both sides in anterior part (Figure 3). In second surgery patient underwent partial (half tendon) transposition and resection (4 mm) of superior and inferior rectus muscle toward medial rectus without scleral fixation suture in both sides. The patient had orthotropia with improved dduction after six months (Figure 4).



Figure 1. Large angle exotropia and limited adduction at first visit



Figure 2. Intraoperative view of eye showed absence of medial rectus muscle.

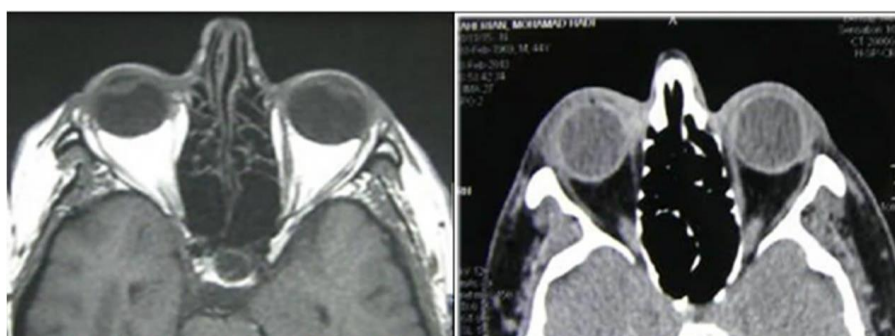


Figure 3. Magnetic resonance imaging showed thin medial rectus muscle.



Figure 4. Six months after augmented partial vertical transposition and lateral rectus recession showed orthotropia in primary position and improved adduction in both eyes.

Discussion

Agensis or hypoplasia of extraocular muscles has been reported since the eighteenth century.¹

These disorders have been grouped as congenital cranial dysinnervations disorders with absent muscle development or absent innervations of target muscle.² Genetic susceptibility is frequently seen.² There are various associated non-ocular defects and phenotypical diversity is common. Magnetic resonance imaging may show normal, hypoplastic or absent cranial nerves and extraocular muscle.^{3,4} For the first time Girard and Neely in 1958 reported unilateral genesis of medial rectus muscle and another case presented by Houtman in 2009. He reported congenital absent of medial rectus muscle in 3-months' infant presented with large angle exotropia. Maximum recess coupled with partial transposition and botulinum injection was performed. Patient had 15 prism diopter alternate exotropia in last follow-up.⁴ Anirudh et al reported familial occurrence of this anomaly. They reported congenital aplasia of medial rectus in 50-year-old man and his two sons. They had less than 20 prism diopter exotropia and improved adduction after vertical transposition of rectus muscle at last follow-up.⁵ Our case is a rare case of congenital hypoplasia of medial rectus muscle in both eyes. Lee et al reported congenital absence of rectus muscle in 8-year-old girl. After full tendon vertical transposition she had 15 prism diopter exotropia in primary position

and improved adduction.⁶ We performed two stage surgeries and have excellent results.

Conclusion

In conclusion we must be aware that in patient with large angle horizontal deviation and marked duction deficit, orbital MRI must be performed. If thin or absent extraocular muscle was found, transposition procedure combined with maximum recess of antagonist muscle are logical surgical options.

References

1. Duke-Elders S. Congenital deformities. In: Duke-Elders S. System of Ophthalmology. Vol III Part 2. Henry Kimpton: London, 1964:980.
2. Gutowski N, Ellard S. The congenital cranial dysinnervation disorders. *Adv Clin Neurosci Rehabil* 2005;5(3):8-10.
3. Demer JL, Ortube MC, Engle EC, Thacker N. High-resolution magnetic resonance imaging demonstrates abnormalities of motor nerves and extraocular muscles in patients with neuropathic strabismus. *J AAPOS* 2006;10(2):135-42.
4. Houtman A, Hocking G, Lloyd C, Biswas S. Large-angle congenital exotropia due to 'absent' medial recti: a case of congenital cranial dysinnervation disorder. *Eye (Lond)* 2009;23(6):1489-90.
5. Singh A, Sharma A, Sharma P. Congenital bilateral aplasia of medial recti in a family. *JAMA Ophthalmol* 2013;131(6):798-800.
6. Lee DW, Lee S, Ahn M. Congenital bilateral medial rectus aplasia. *J Pediatr Ophthalmol Strabismus* 2013;50(3):134-5.