Case Report

Successful Surgical Outcome in a Case of Monocular Elevation Deficiency after Knapp's Procedure with Foster Augmentation- A Case Report

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Abstract

Monocular elevation deficiency has multifactorial etiology and different surgical techniques have been described for its management involving inferior rectus recession, knapp's or modified knapp's transposition or full tendon knapp's transposition with foster augmentation in a single or multi staged surgical intervention. In this case report, we present a case of MED with large angle hypotropia and pseudoptosis managed surgically and documenting the ocular alignment stability at 6 months follow up.

Keywords: Knapp's procedure, Foster augmentation, Monocular elevation deficiency.

Introduction

Monocular elevation deficiency (MED) is a rare ocular condition characterised by restricted elevation in one eye in all positions of upgaze, sometimes associated with hypotropia, ptosis &/or pseudoptosis.[1] It can be congenital or acquired. There are three forms of this disorder.[2]

- Type 1: Inferior rectus fibrosis -Force duction test (FDT) positive for inferior rectus, poor bell's phenomenon & normal upward saccades till midline
- Type 2: Superior rectus palsy-FDT negative, absent bell's phenomenon, slow upward saccades above & below midline
- Type 3: Supra nuclear palsy-FDT negative, present bell's phenomenon, slow upward saccades below midline & absent above midline

Management of MED depends upon the degree of hypotropia and FDT

Case presentation

An 18 years old female presented in strabismus OPD with complaints of drooping of right eye upper lid since birth. It was non-progressive and there was no diurnal variation. Patient had come due to cosmetic concerns regarding the same. She also complained of diminution of vision in right eye since childhood. There was no history of trauma or other systemic illness. There was no H/o similar complaints in the family.

Ocular examination

Her best corrected visual acuity in right eye (RE) was 4/60 and in left eye(LE) was 6/6. Orthoptic evaluation was done and right eye showed severe ptosis (Figure 1A) with 20 degree hypotropia and small angle exotropia on hirschberg test. Ocular deviation was measured using loose prisms. RE had hypotropia of 45PD with modified krimsky test and exotropia of 10PD. RE extra ocular movements were severely restricted to -4 (Figure 1C) in upgaze in both adduction and abduction position. Bell's phenomenon was poor in RE. On cover test, RE ptosis improved when LE was covered, demonstrating pseudoptosis in RE. Worth's 4 dot test revealed no diplopia. Jaw winking phenomenon was absent. Rest anterior segment and fundus examination was normal in both eyes. FDT was negative in both eyes. RE was amblyopic due to long standing ocular deviation and associated ptosis. CT head and orbit was normal.

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Figure 1: A) Pre op image depicting severe ptosis in Right eye. B) Right eye hypotropia with exotropia. C) Right eye severe upgaze restriction to -4

Management

Patient was explained regarding the need for multiple staged surgical procedures and guarded prognosis. After informed consent and physical fitness, patient was planned for Right eye Knapp's procedure with foster augmentation (Figure 2) under local anaesthesia. FDT was repeated on table and was documented as negative. Using limbal approach, full tendon medial rectus (MR) and lateral rectus (LR) muscles were transposed to the insertion of the superior rectus (SR) muscle using 6-0 vicryl sutures. Foster augmentation was done by suturing 1/3rd fibres of MR and LR muscle with SR 6mm behind superior rectus insertion using non- absorbable 5-0 ethibond sutures. Conjunctiva was closed using 8-0 vicryl sutures.

Result

At 1 week follow up, RE hypotropia had significantly reduced from 45PD to 16PD and elevation improved from -4 restriction to -2. As the hypotropia reduced significantly, RE ptosis also improved after strabismus surgery. At 3 months and 6 months follow up, the improved ocular alignment was stable with significant improvement in ptosis (Figure 5).

Discussion

MED should be differentiated from progressive external ophthmoplegia, myaesthenia gravis, congenital fibrosis of extra ocular muscles, 3rd nerve palsy or brown's syndrome. In MED, when the patient fixates with the nonparetic eye, the paretic eye will take a hypotropic position and the upper lid may be ptotic. Fixation with the paretic eye will cause a hypertropia of the nonparetic eye and improvement of ptosis in paretic eye, demonstrating pseudoptosis. Restriction in upgaze is present in both addiction and abduction. The goal of surgery in MED associated with ptosis or pseudoptosis is the combined management of hypotropia and ptosis. The procedure of choice for surgical correction of MED is determined firstly by FDT, to rule out any inferior rectus (IR) restriction and secondly by degree of hypotropia. In the presence of IR restriction, IR recession is done primarily followed by knapp's transposition as a secondary procedure

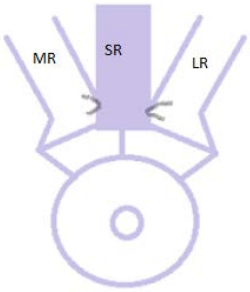

Figure 2: Knapp's procedure with foster augmentation

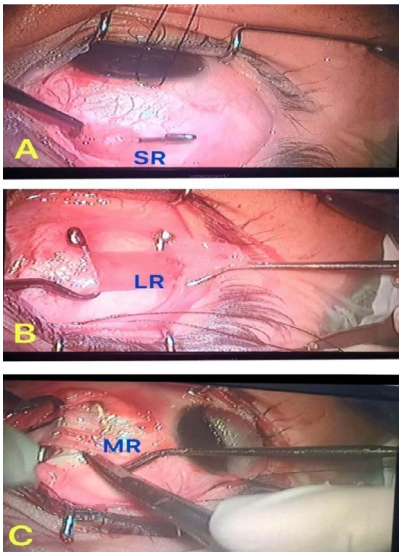

Figure 3: Using limbal approach, medial rectus (MR), lateral rectus (LR) and superior rectus (SR) muscle were secured

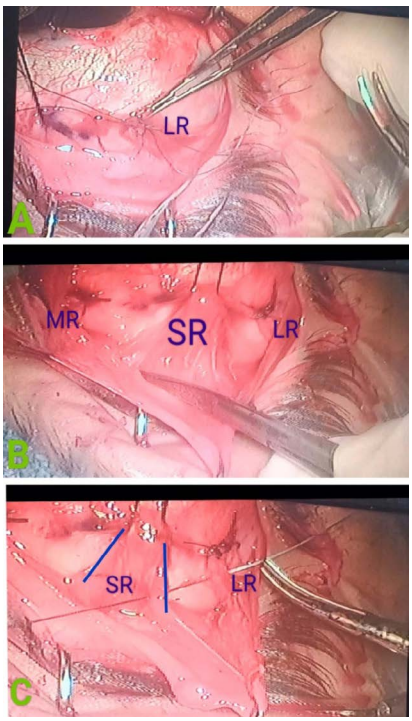

Figure 4: Full tendon MR and LR muscles were transposed to the insertion of SR muscle using 6-0 vicryl sutures. Foster augmentation was done by suturing 1/3rd fibres of MR and LR muscle with SR 6mm behind SR insertion using non- absorbable 5-0 ethibond sutures

Figure 5: Significant reduction in Right eye Ocular deviation and improvement in ptosis

to avoid risk of anterior segment ischemia. In the presence of SR palsy (paretic form), Knapp transposition procedure is preferred.

For hypotropia <25 PD, modified knapp's is preferred in which superior half of equally divided (up to 15 mm) tendon of MR and LR are transposed to the insertion of the SR muscle.[3] The remaining of the horizontal muscles can be used for surgical correction of any horizontal deviation as a combined procedure. For hypotropia 25–35 PD, full tendon knapp's transposition is done. For hypotropia >35 PD, knapp's with foster augmentation is preferred.

Improvement in upgaze after Knapp transposition, is due to change in the point of tangency of the muscle with the globe, thus changing the point of mechanical action.[1] Transposition results in a new muscle plane with a new axis of rotation.[4] Improvement in upgaze following IR recession occurs due to weakening of the restricted muscle, in the presence of normal SR function. In cases associated with Marcus Gunn jaw winking phenomenon, levator disinsertion and sling surgery may be required as secondary procedure. MED may be associated with dissociated vertical deviation (DVD).[5] But was not found in this case.

Conclusion

MED is etiologically multifactorial and may require multiple staged surgeries. Satisfactory surgical results can be achieved by judicious selection of patient, selecting surgical technique based on the results of the forced duction test and intervening for any associated true ptosis after squint correction if required.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflict of Interest

There are no conflicts of interest.

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