Lateral Rectus Superior Compartment Palsy

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ABSTRACT

Lateral rectus palsy with hypotropia constitutes a portion of sixth nerve palsy cases in which only the superior part of the lateral rectus is affected. We present such a case in a 10-year-old young Pakistani lady who presented with a peculiar appearing right esotropia and hypotropia due to acquired lateral rectus palsy along with apparent ipsilateral superior rectus underaction. Neuroimaging confirmed atrophy of the superior part of lateral rectus as compared to the inferior half. The superior rectus muscle was normal, along with other extraocular muscles. This confirmed our suspicion of superior compartment LR palsy. Augmented superior rectus transposition to the lateral rectus along with adjustable bimedial recessions and bilateral inferior oblique myectomies were done to restore her cosmetic appearance.

Key Words: Lateral rectus palsy, sixth nerve palsy, esotropia, hypotropia, transposition, strabismus.

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INTRODUCTION

Compartmentalization of extraocular muscles has been studied extensively by Demer1 and Clark2, who put forth evidence that individual muscles have different functions corresponding to different fiber groups. The lateral rectus (LR) muscle is believed to have a dual embryonic origin and the abducent nerve (cranial nerve VI) is believed to innervate the lateral rectus by two or more trunks, with separation seen from as far as the cavernous sinus to the muscle itself, in autopsies. This is believed to divide the lateral rectus into two separate compartments: superior and inferior; and a sixth nerve palsy, either complete or partial, may affect any one of these. This holds true for other extraocular muscles as well.

Physiological behaviour of extraocular muscles studied through special MRI techniques also favors neuromuscular compartmentalization. Selective pathology of the different compartments can yield peculiar strabismus patterns, leading to erroneous diagnosis. The majority of sixth nerve palsies affect the superior compartment more than the inferior compartment of the lateral rectus, characterized by an esotropia with coexisting ipsilateral hypotropia. This may pose the diagnostic dilemma of a vertical muscle palsy in addition to the LR palsy1-3.

We present a case of LR superior compartment syndrome in a 10-year-old girl who presented with a right sided acquired esotropia with hypotropia, accentuated upon abduction.

CASE PRESENTATION

A 10-year-old girl presented to the Eye OPD of Fauji Foundation Hospital, Rawalpindi; which is a tertiary care teaching hospital affiliated with the Foundation University Medical College; with a right sided esotropia and inability to turn the right eye outwards of 1½ year duration, consequent to a severe febrile illness. Old hospital records were unavailable. Previous photographs showed that she was orthotropic...
prior to the event. On examination, her unaided visual acuity was 6/6 bilaterally. On prism cover testing, she had a right esotropia (RET) of 75 prism diopters (PD) along with a right Hypotropia (RHoT) of 5 PD in primary distance gaze, RET of 62 PD and RHoT of 2 PD in upgaze, RET of 80 PD and RHoT of 2 PD in
downgaze, RET of 60 PD and RHoT of 10 PD in right gaze and RET of 70 PD and RHoT of 9 PD in left gaze. The esotropia had a ‘V’-pattern of 18 PD. At near fixation, she had a RET of 80 PD and no hypotropia. She had a right LR underaction of -3 to -4, a right medial rectus (MR) overaction of +2, right superior rectus (SR) underaction of -2.5, bilateral inferior oblique overaction of +3, left LR underaction of -2 and bilateral superior oblique underaction of -1.5 (Figure 1).

She had a right face turn. She was thus diagnosed with a right partial abducent nerve palsy and there was suspicion of a right superior rectus palsy. Titmus fly test showed stereopsis at 160 seconds of arc. Worth 4 dots testing showed alternating suppression. Anterior segment was normal. Her fundus examination although normal showed bilateral fundus extorsion. We presumed that the cause of the sixth nerve palsy was infectious, possibly meningitis. Systemically she did not have any co-morbid conditions.

A thorough investigative workup was done. Complete blood picture was normal, ESR was 24, plasma glucose and thyroid profile was normal. Urine routine examination, liver function tests and renal function tests were normal. Neurological examination was normal. A thin section MRI Orbit & Brain with contrast (2.5 mm) was ordered, which showed normal Pons and failed to reveal a lesion along the path of the abducent nerve. The right LR muscle was atrophied as compared to the left LR; and its superior half was smaller as compared to the inferior half on coronal sections (Figure 2). The superior rectus muscle was normal, along with other extraocular muscles. This confirmed our suspicion of superior compartment LR palsy.

We performed a bimedial recession of 7 mm (right eye on an adjustable suture with a final adjustment to 5 mm). An augmented transposition of the right SR to the LR with a non-absorbable Ethibond 5/0 augmentation suture, 12 mm behind the insertions was done, incorporating 1/4th of both the muscle bellies. Bilateral inferior oblique myectomies were also performed in a single setting. At 1 year post-operatively, she was well aligned, very happy although an elevation deficit in upgaze was persistent which we attributed to SR transposition, and inferior oblique myectomy (Figure 3).

DISCUSSION

Abducent nerve is the sixth cranial nerve and solely innervates the lateral rectus muscle. It primarily functions to abduct the eye. Its nucleus lies within the pons, ventral to the floor of the fourth ventricle, and its fasciculus leaves the brainstem at the ponto-medullary junction. The basilar part passes upwards near the skull base and is crossed by the anterior inferior cerebellar artery. It pierces dura below the posterior clinoids and passes the petrous tips, through the Dorello canal to enter the cavernous sinus, lying in close proximity to the cranial nerves III, IV and V1, and the internal carotid artery. It enters the orbit via the superior orbital fissure within the annulus of Zinn. A sixth nerve palsy can be caused by any insult along its long course. Causes of acquired sixth nerve palsy in children are trauma, neoplasms, infections,
vasculopathies, aneurysms, arteriovenous malformations, raised intracranial pressure, demyelination or iatrogenic; to name a few.\textsuperscript{4,5}

The abducent nerve has a bifid innervation structure to divide the lateral rectus into two functional neuromuscular compartments; superior and inferior. This compartmentalization allows the LR to have additional vertical and torsional actions in addition to abduction, due to differential contraction of these compartments during ocular counter-rolling, vertical ductions and vertical vergence. Thus, a lesion along the abducent pathway may affect only one compartment of the LR to cause a compartmental palsy. The lateral rectus superior compartment palsy is a newer subtype of abducent palsy, exhibiting asymmetric atrophy of the superior compartment only, resulting in vertical and torsional abnormalities concurrent to the abduction limitation. This results in paralytic esotropia coexistent with ipsilateral hypotropia and excyclotropia; with the hypotropia increasing in abduction. This was clearly seen in our case. This may occur in both complete and partial palsies of the sixth nerve. Surface coil thin section coronal MRI studies have confirmed the existence of such palsies, and have shown significant reduction in the maximum cross-sections of the superior compartment\textsuperscript{1,2,5,6}.

Treatment of an acute LR palsy is alternate occlusion, botox to the medial rectus and requires observation for six months to one year, to allow for spontaneous resolution to occur. Surgery for non-resolving abducent palsies depends upon the degree of deviation and whether the palsy is complete or partial; it involves either recession of the contralateral synergistic muscle (MR), recession of the direct antagonist (ipsilateral MR), LR resection, or contralateral antagonist resection (LR). In cases of complete palsies, temporal transposition procedures of the vertical rectii may be done, like Hummelsheim or Jensen. Operating on multiple muscles especially the vertical rectii in conjunction with the horizontal rectii may pose a risk of anterior segment ischemia. A recent therapy for LR palsy is augmented transposition of the SR to the LR with a non-absorbable suture\textsuperscript{7-10}.

We thus performed a bimedial recession with an augmented SR transposition, as we have found this to be safer and effective, and because LR function has completely returned, this also supports the hypothesis of a superior compartment LR paresis. Inferior oblique overaction was treated by myectomies, which improved her bothersome up shoots on adduction.

**Conflict of Interest**
Authors declared no conflict of interest.

**Author’s Designation and Contribution**
Dr. Sana Nadeem; Assistant Professor: Concept, data collection, patient management, manuscript writing.

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