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A Case Report of Congenital Left Eye Monocular Elevation Deficiency With Pseudoptosis

KEYWORDS: Monocular elevation deficiency, Double elevator palsy, pseudoptosis, hypotropia.

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ABSTRACT *A monocular elevation deficiency is a rare condition which is also known as Double elevator palsy. It has been documented less frequently. It suggests that both elevator muscles (the superior rectus and the inferior oblique) of one eye is weak, with resultant reduced ability to elevate the eye in all fields of the gaze and a hypotropia in the primary gaze. It may be congenital or acquired. 40 % of the cases present with pseudoptosis. It is occurred due to the presence of fascial attachments between the superior rectus and the levator palpebral superioris muscle. Pseudoptosis resolves when paretic eye gets fixation unlike true ptosis. I am presenting a case of 14 yrs old female patient having left eye congenital monocular elevation deficiency with pseudoptosis.*

Introduction:
Double elevator palsy currently known as Monocular elevation deficiency is a rare condition which was first described by White in1942 and later by Dunlap. [1] It can be congenital or acquired. In MED, both the elevators (the superior rectus and the inferior oblique) are weak or there may primary restriction of the inferior rectus muscle. Classically, congenital MED presents with a unilateral limitation of elevation in both adduction and abduction. There is also hypotropia of the affected eye. Rarely, a patient may fixate with the hypotropic eye manifesting a large secondary hypertropia of the normal eye. [2] It is also associated with pseudoptosis which resolves after taking fixation. [2].

Case history:
A 14 yrs old female brought to our department by her parents with the complaints of drooping of the left upper eyelid and downward displacement of the left eye since birth. She was not complaining about doubling of the images. Her birth history was normal. Developmental milestones were normal. There were no histories of any systemic diseases or association with any congenital syndrome. Family history was negative. General examination was normal. Other systemic examinations were normal including neurological also.

On ocular examination, Visual acuity in both eyes were noted to be 20/40 with her spectacle n unaided were 20/80 with pinhole 20/40in her right eye n 20/120 with pin hole 20/40 in her left eye. She is presented with slightly chin up head position. Left eye pseudoptosis was present which resolves after taking fixation with the left eye and occluding the right eye (Fig 1, 2).

Extraocular muscles movement revealed a restriction of the movement of left eye in up gaze (supraduction, dextroelevation, levoelevation) (Fig-3) pointing towards elevator deficit (left superior rectus and left inferior oblique). All other movements were normal. Cover uncover test revealed left eye hypotropia in primary gaze. When left eye took the fixation, right eye showed hypertropia. Hirschberg test was done which showed deviation of 30 degree. Worth's four dot test was done revealed no diplopia. Visual fields were normal by confrontation test. Bell's phenomenon was normal. Jaw winking phenomenon was absent. Pupil was regular, round and reactive to light. There were no restrictions of the right eye movements. Anterior segment were normal with normal fundus evaluation. Intraocular pressures were normal. FDT was negative on elevation. Cycloplegic refraction revealed 20/40 with -0.75 cyn at 180 degree in right eye n -1.25 cyn at 180 degree in the left eye. CT was done and was normal.

Discussion:-
In MED, there is unilateral limitation of elevation in all horizontal orientations of the eye. When the patient fixates with the nonparetic eye (Right), the paretic eye (left) will take a hypotropic position and the upper lid may be slightly ptotic. Fixation with the paretic eye (Left) will cause a hypertropia of the nonparetic eye (Right), and ptosis may disappear, due to some fascial attachment between superior rectus and levator palpebral superioris provided the levator palpebral is not involved unlike true ptosis. Anatomical improbability that both superior rectus and inferior oblique are involved by single lesion suggests long standing SR palsy as the primary event with later spread of comitance leading to inferior oblique involvement. FDT is done to confirm any mechanical restriction of movement and was negative. Often there is chin up position to achieve binocular single vision. Bell's phenomenon is usually preserved and differentiates between supra nuclear and infra nuclear lesions. No risk of post-operative corneal exposure.

There are mainly two forms; Congenital and Acquired. [3,4] Congenital causes are superior rectus and inferior oblique palsy, primary inferior rectus muscle restriction, defect in supranuclear pathway for upgaze located in midbrain pretectum. Possible differential diagnosis of DEP include blow-out fracture of the orbital floor, congenital or acquired fibrosis, endocrine myopathy, myasthenia gravis, skew deviation, Parinaud's syndrome, Browns syndrome, heavy eye syndrome, pineocytoma and 3rd nerve palsy.

There are different surgical treatments for DEP which reflects the various causative mechanisms. [5]

Treatment is generally based on carrying out the forced duction test (FDT).

If FDT is positive, inferior rectus recession is carried out

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and most patients recover satisfactorily. In the absence of IR restriction, (forced duction test negative) surgical treatment comprises of the KNAPP PROCEDURE. [6, 7] Here the entire tendon of both the medial and lateral rectus muscle is transferred to the side of superior muscle insertion. In transposition, as the 4 anterior ciliary arteries are sacrificed, a 6 month adaption time should be given before operating the third rectus to prevent anterior segment ischemia. In the absence of IR restriction, results are good. If the IR is restricted, the muscle has to recess before or after transposition.

If vertical deviation in primary position is large, then IR recession with SR resection is done.[8] Recently, tucking of IR of normal eye is also done to remove residual hypotropia and pseudoptosis that may persist after the Knapp Procedure .[9]

Therefore, the surgical procedure should be individualized for each case based on the etiology and mechanisms of involvement.

Fig no 1:- Left eye pseudoptosis.

Fig no 2:- Pseudoptosis is relieved after taking fixation with same eye.

Fig no 3- left eye elevation palsy showing restriction in elevation, levoelevation dextroelevation.

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