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Case Report

Surgical Dilemma in Combined Anomaly of Squint and Ptosis with Jaw Winking-A Case Report

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Abstract

Ptosis and Pseudoptosis (secondary to hypotropia): If a difference in width of two lid fissure exists, the possibility must be entertained that there is a ptosis or pseudoptosis of the upper lid of the eye with the narrow lid fissure. The two must be differentiated. Here we are presenting an unusual case of uniocular double elevator palsy with component of pseudoptosis along with jaw winking phenomenon and true ptosis. When affected eye fixates, part of ptosis improves (referred as pseudoptosis) while residual true ptosis remains. Surgical management was undertaken -with inferior rectus recession of 5 mm. Patient was kept under regular follow up and residual ptosis of 2 mm was present. Since patient has jaw winking and BCVA of 6/6 in both eyes, there is no urgency in performing ptosis surgery for residual ptosis.

Keywords: *Ptosis; Pseudoptosis; Hypotropia; Marcus-Gunn*

Introduction

Ptosis and pseudoptosis (secondary to hypotropia)

- If a difference in width of two lid fissure exists, the possibility must be entertained that there is a ptosis or pseudoptosis of the upper lid of the eye with the narrow lid fissure. The two must be differentiated.
- When a patient has weakness of the elevation of one eye, the lid fissure of that eye is narrower than that of the unaffected eye. The patient may have a true ptosis of the upper lid, especially if the superior rectus is involved.
- However, the ptosis may be only apparent, with the narrowness of the lid fissure due to hypotropic position of the globe. There is then a pseudoptosis.
- Pseudoptosis can be established by having the patient fixates with affected eye. If there is pseudoptosis, the lids will open to their normal width. The lids of the unaffected eye will widen abnormally as the globe receives an excessive impulse and goes into a hypertropic position.

- For correction of pseudoptosis, all that is required is to bring the eyes to the same level by operating on the appropriate extra-ocular muscle.
- In double elevator palsy, elevation of paretic eye from any position is severely restricted.
- Bell's phenomenon is usually preserved but may be absent.

Case Report

- Here we are presenting an unusual case of uniocular double elevator palsy with component of pseudoptosis along with jaw winking phenomenon and true ptosis.
- When affected eye fixates, part of ptosis improves (referred as pseudoptosis) while residual true ptosis remains.
- A 29 year old male attended our eye opd with chief complaints of:
    - Drooping of right eye upper lid,
    - Intermittent diplopia since birth,
    - Diminution of vision.
- The patient was born of normal vaginal delivery with no other systemic illness.
- There was no history of similar or other ocular problems in the siblings of family.
- On detailed examination the visual acuity in the right eye was 6/18 and in left eye was 6/6 improving with BCVA 6/6 in both eye. In primary position there was eye ptosis with hypotropia of 15 degree (Hirschberg test).
- Rest of the slit lamp findings were within normal limits.
- Jaw winking phenomenon was present.

Squint workup

- On Prism bar cover test, right eye hypotropia was present of 32 prism diopters.
- On worth four dot test, three green circles were seen suggestive of left eye dominance with right eye suppression.
- Ocular movement were normal in left eye while in right eye the movements were restricted in elevation, levoelevation and dextroelevation.
- Central fixation was present in both eyes with no abnormal retinal correspondence.
- No diplopia was present.

Ptosis work up

Variables	Right Eye	Left Eye
Palpebral Fissure Height	6 mm	9 mm
Margin Reflex Distance I	0 mm	3 mm
Upper Lid Crease	5 mm	7 mm
Levator Function Test	6 mm	15 mm
Jaw Winking Reflex	Present	Absent
Bell's Phenomenon	Absent	Absent
Superior Rectus Function	Absent	Intact

Management

- Refractive correction was made:
    - RE-0. 75D sph with -0. 5cyl 90 degree-6/6
    - LE-0. 5D cyl 90 degree-6/6.
- Surgical management was undertaken -with inferior rectus recession of 5 mm.
- Patient was kept under regular follow up and residual ptosis of 2 mm was present.
- Since patient has jaw winking and BCVA of 6/6 in both eyes, there is no urgency in performing ptosis surgery for residual ptosis.

Discussion

Marcus Gunn jaw winking ptosis is a congenital ptosis associated with synkinetic movements of upper lid on masticating movements of the jaw. It is usually unilateral but rarely presents bilaterally. Affects male and females in equal proportion [1].

Our patient presented with uniocular double elevator palsy with component of pseudoptosis along with jaw winking phenomenon and true ptosis. When affected eye fixates, part of ptosis improves (referred as pseudoptosis) while residual true ptosis remains. For managing the case, Refractive correction was made- RE-0. 75D sph with -0. 5cyl 90 degree-6/6 and LE-0. 5D cyl 90 degree-6/6.

Surgical management was undertaken -with inferior rectus recession of 5 mm. Patient was kept under regular follow up and residual ptosis of 2 mm was present. Since patient has jaw winking and BCVA of 6/6 in both eyes, there is no urgency in performing ptosis surgery for residual ptosis.

Similar conclusions have been drawn in other studies, Dillman and Anderson stated that removal of a portion of the levator muscle above the Whitnall's ligament (i.e. myectomy) is adequate to obliterate its function without extensive dissection and damage to eyelid structures [2].

Beard and others have advocated bilateral excision of the levator muscle and bilateral frontalis suspension. This approach almost completely eliminates the wink and results in better symmetry, it is often difficult to persuade the parents and the patient to perform surgery on the contralateral normal eye.

Kersten., *et al*. advocate unilateral levator muscle excision and frontalis sling only on the affected side [3].

Conclusion

Differentiation of ptosis and pseudoptosis is of utmost importance:

- Element of amblyopia should be ruled out.
- Surgery should be planned in stages and explained to the patient beforehand.
- Pseudoptosis associated with hypotropia, should be corrected by operating on appropriate extra ocular muscle. Then the residual ptosis, if present should be corrected surgically.
- Jaw winking phenomenon is usually not associated with amblyopia.

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