Congenital Oculomotor Synkinesis - A Case Study
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Citation

Abstract
A case of oculomotor synkinesis was presented with deviation of the right eye and intermittent total ptosis associated with abnormal lid movements since birth. Her mother has been suffering from tuberculosis with irregular usage of anti-tubercular treatment during pregnancy. The strabismus was corrected with maximal unilateral recession and resection with acceptable post-op results.

CASE REPORT
A 19-year-old female reported to our hospital with complaints of deviation of right eye and restriction of movements. She also complained of diminution of vision and abnormal movements of the eyelid in the right eye since childhood. Her parents revealed that the presence of abnormal eyelid movements and deviation of right eye were there since birth. She complained of intermittent and involuntary closure and opening of the right eyelid associated with facial expressions like smiling and during sleep. Her mother had a history of tuberculosis for which she had taken anti-tubercular drugs irregularly for 6 months during her pregnancy (no medical records were available). Her mother does not recall any use of isotretinoin or Vitamin A supplements. There were no complaints of diplopia, tearing or any asthenopic symptoms. There was no history of spectacle wear, ocular trauma or surgery as well as any systemic illness. Her birth was a normal vaginal delivery at full term. Family and sibling history were also not significant.

On examination, her vision in her right eye (RE) was counting finger 2 metres, not improving with pinhole and in the left eye (LE) was 6/6. Head posture, forehead and eyebrows were normal. There was complete, but intermittent, blepharoptosis of the right eye with involuntary opening of the eyelid on levoversion, with sideways jaw movements and sometimes with smiling. A faint lid crease was present 5mm above the lid margin. Bell’s phenomenon was absent. (Fig. 1) It does not seem to be a case of typical Marcus Gunn jaw winking phenomenon or Duane’s retraction syndrome. There was no evidence of any other cranial nerve abnormality on examination.

Figure 1
Figure 1: Lid elevation on smiling and lateral movement of jaw

On primary gaze, the left eye was fixating and there was right eye exotropia of 65 prism dioptries (PD) with slight hypotropia. Extraocular movements of the right eye were restricted in all gazes. (Fig. 2)
On slit-lamp examination the conjunctiva, cornea and anterior chamber of both eyes were within normal limits. The right eye pupil was mid-dilated and showed aberrant response to light stimulus, sometimes reacting and sometimes not showing any response to direct as well as consensual stimuli. It also showed no response on convergence. Fundus examination of both eyes revealed no abnormality, thus suggesting amblyopia. MRI brain and orbit was unremarkable for the brain, but showed hypogenesis of right levator palpebrae superioris muscle (LPS) and abnormal contour and course of right medial rectus. (Fig. 3)

For cosmetic purposes, correction of the strabismus was planned. On the operation table, a forced duction test before and after conjunctival resection, revealed a restrictive component of the right lateral rectus. A maximal 10 mm recession of the right lateral rectus and an 8mm resection of the right medial rectus were done. The temporal conjunctiva was also resected by 5mm.

Surgical intervention to correct the Blepharoptosis was not done in view of absent Bell’s phenomenon. Lid crutches were advised for Blepharoptosis.

On post-op day 1, the patient was orthophoric in primary gaze. Slight adduction (15 prism dioptres) of the right eye was present on dextroversion. Upon rest, all movements were absent. There were no complaints of diplopia. (Fig. 4)
Figure 4
Figure 4: The patient was orthophoric in primary gaze on post-op day 1.

On 1 month post-op, 10 prism dioptres exodeviation with slight hypotropia was seen. (Fig. 5)

Figure 5
Figure 5: 1 month post-op, 10 PD exodeviation with slight hypotropia was seen.

DISCUSSION
The most likely explanation for abnormal movements (oculomotor synkinesis) is prenatal aberrant innervations of eyelid and extraocular muscle. It is well known that the common congenital synkinetic aberrant innervations syndromes are Marcus Gunn jaw winking and Duane’s Retraction syndrome. The present case does not appear to fit into these two syndromes. Besides these conventional syndromes, very few cases of unconventional aberrant regeneration have been described in literature. There have been reports of Isotretinoin (RA) embryopathy and oculomotor synkinesis. In these case reports, RA, which is a known teratogen, was used for treatment of cystic acne. In the present case, to the best of patient’s mother’s knowledge, neither vitamin A nor multivitamin were given during pregnancy. Though the patient’s mother’s medical history is not well known, it is known that she was given antitubercular drugs, namely, rifampicin, isoniazid, ethambutol and pyrizinamide. Thus, it may be suggested that there may be a possible correlation between antitubercular drugs and oculomotor misdirection. Though certain second line drugs, particularly, aminoglycosides poses a very high risk of fetal deafness, no such case of oculomotor synkinesis appears to have been reported in literature, due to antenatal exposure to anti tubercular drugs. Hence, it is suggested that in case of oculomotor synkinesis, exposure to antitubercular drugs in the ante-natal period should be investigated.

References
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