ORIGINAL RESEARCH PAPER
Volume - 11 | Issue - 10 | October - 2022 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

INVERSE MARCUS GUNN SYNDROME – A RARE FACIAL SYNKINESIS

Ophthalmology

Dr. Anant Prakash Tripathi* DO, MS, PGDHA *Corresponding Author

Dr Ekta Sain MS

ABSTRACT

Marcus gunn syndrome is one of the most common variety of oculofacial synkinesis. It is seen in 4-6% of congenital ptosis patients. An aberrant neural misdirection appears to exist between motor branch of trigeminal (CN V3) nerve innervating the external pterygoid muscle and fibers of the superior division of oculomotor nerve (CN 3) that innervate the levator palpebrae superioris. Inverse Marcus Gunn syndrome, a very rare variant of synkinesis characterized by lid drooping on masticatory movement and opening of mouth due to the misfiring between 3rd and 5th cranial nerve. An 8-year-old male child presented with congenital ptosis with anomalous drooping of upper lid while eating or opening of mouth.

KEYWORDS

inverse Marcus gunn syndrome, jaw-winking, ptosis, congenital, orbit, synkinesis, misdirection

INTRODUCTION

Marcus Gunn first described the syndrome in 1883 in a patient of congenital ptosis which had paradoxical retraction of upper eyelid on movement of the jaw, which later came to be known as "jaw-winking" syndrome. [1] It accounts for 5% of all congenital ptosis.

Inverse Marcus gunn is a rare congenital synkinesis presenting as a drooping of eyelid on masticatory movement. This is most commonly seen as a congenital anomaly rather than acquired. [2]

Case Report

An 8 year old male child presented with right eye upper eyelid drooping which was noticed by mother since birth. It was associated with aberrant increase in drooping of upper eyelid while eating or opening of mouth since birth. The child was otherwise well, no history of any other ocular problems, and there was no family history of similar complaints. On examination, the best corrected visual acuity was 6/6 , N6 both eyes (OU). The intraocular pressure was 14mmHg OU. The anterior segment was unremarkable. Fundoscopically , both eyes were normal. Chin elevation was noted with a right eye (OD) ptosis of 3mm with frontalis overaction. Margin reflex distance (MRD) 1 was +1mm in right and 3.5mm in left eye with good LPS action of 12mm in right and 15mm in left eye, and normal bell's phenomena. Ocular motility was full and free in all directions of gaze. The right eye ptosis drooping increased on opening of mouth. There was no evidence of facial paralysis or sensory changes. There was an ear abnormality - non-bifid conchal arch and absence of fossa triangularis and umbilical hernia.

DISCUSSION

Also known as Marcus Gunn "jaw-winking" syndrome, is an autosomal dominant condition with incomplete penetrance. [3] No known racial predilection exists.

Reports showed jaw-winking ptosis to be more prevalent in females than in males; however, larger case series have shown an equal prevalence among males and females. [4]

Marcus Gunn jaw-winking syndrome is usually evident at birth. The winking phenomenon is often first noted by the parents when the infant is feeding.

There are multiple hypothesis regarding the exact mechanism of the syndrome. The electromyographic studies done at different timelines and individuals showed different results:

Most commonly accepted hypothesis is the inhibition or inactivation of the levator palpebrae superioris muscle with jaw opening or mastication without any impulses in orbicularis oculi , thus denoting a misfire of the oculomotor (CN III) with trigeminal activity of lateral pterygoid – a trigemino-occulomotor (3^{rd} – 5^{th} CN) synkinesis. [5]

The co-contraction of the orbicularis oculi with the lateral pterygoid when patient opened her mouth , showing this phenomena is caused by the synkinesis of the trigeminal and facial nerves (5^{th} – 7^{th} CN) . [6]

In a patient recovering from facial nerve paresis (acquired cases) the mechanism is ascending proprioceptive impulses from the stretch during the full mouth opening which are carried by facial nerves could trigger contraction of orbicularis oculi after a traumatic nerve injury. probably caused by aberrant regeneration of nerve fibers, some axons growing into the wrong sheath in the anatomical disorder which may accompany regeneration, showing an Interfacial association. [7,8,10]

Oldest hypothesis :Atavistic reversion, as suggested by Sano on basis of electrophysiological study, that the old intimate association of the orbicularis and the external pterygoid muscles, although separated in the course of development, may be re-united as a release-phenomenon in presence of supra-nuclear lesion affecting the cortical connections of the trigeminal system explaining Wartenberg's hypothesis of 'release-phenomena' which represent failure of inhibition of primitive reflexes. [7]

Marcus Gunn jaw-winking syndrome may occur in isolation or association with other ocular movement disorders such as, strabismus is found in 50-60% of cases. Superior rectus palsy is found in 25% of cases, and double elevator palsy is found in another 25% of cases. Amblyopia occurs in 30-60% of patients with Marcus Gunn jaw-winking syndrome and is almost always secondary to strabismus or anisometropia, and, only rarely, is due to occlusion by a ptotic eyelid. . Incidence of anisometropia among patients with Marcus Gunn jaw-winking syndrome is reported to be 5-25%. In double elevator palsy, a deficiency in elevation of the globe occurs in all positions of gaze, secondary to an apparent weakness of the superior rectus and inferior oblique muscles. On rare occasions, horizontal strabismus in the absence of a vertical motility disturbance may occur. Some systemic anomalies such as cleft lip or palate are found in rare cases, while CHARGE syndrome association is seen in bilateral cases. [2]

Marin-Amat syndrome, having similar pathophysiology, also needs to be differentiated from Inverse Marcus gunn syndrome. Marin Amat syndrome is associated with a blepharospasm on mouth opening due to orbicularis oculi contraction. Generally following a facial nerve palsy- an acquired lesion, with a hypothesis of 'aberrant facial nerve' regeneration. Mainly treated medically with Botulinum toxin injections. [4]

While inverse Marcus Gunn is a congenital disorder due to the misinnervation between 3^{rd} and 5^{th} (trigemino-occulomotor synkinesis) with misfiring of the 3^{rd} nerve. [5]

Treatment may not be necessary for patients with a slight degree of deformity. For severe ptosis cases, bilateral frontalis sling with disinsertion of the levator has been suggested to abolish the synkinesis and correct the subsequent ptosis. Once the diagnosis is made based on clinical signs, it is important to investigate other body systems that may be involved. A multidisciplinary approach is important to diagnose and treat the systemic associations.

CONCLUSION

Inverse Marcus Gunn is an extremely rare phenomena of orbit. The major challenge is its diagnosis and differentiation from Marin Amat syndrome, Management should always be multidisciplinary due to multiple systemic associations. Electro physiological studies is helpful in making a confirmatory diagnosis after appreciating the clinical picture.

Declaration by the patient

Authors certify that they have obtained all appropriate patient consent forms. They have given his/her/their consent for display of the images and the clinical information to be reported in a journal. The patient understands that the name will not be published and due efforts will be made to conceal their identity.

Financial support and sponsorship

NIL

Figure 1: Normal Eyelid Position In Primary Gaze

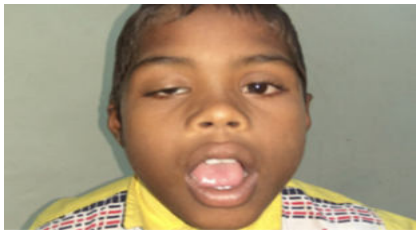

Figure 2 : Drooping Of Upper Eyelid On Opening Of Mouth

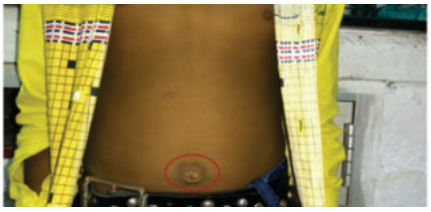

Figure 3 : Associated Umblical Hernia

Figure 4 : Absence Of Fossa Triangularis

REFERENCES :

1. Gunn RM. Congenital ptosis with peculiar associated movements of the affected lid. Trans Ophthal Soc UK. 1883;3:283–7.
2. Davis, G., Chen, C. & Selva, D. Marcus Gunn syndrome. *Eye* **18,** 88–90 (2004). https://doi.org/10.1038/sj.eye.6700509
3. Kirkham TH. Familial Marcus Gunn phenomenon. *Br J Ophthalmol*. 1969;53(4):282. 283. doi:10.1136/bjo.53.4.282
4. Jethani J. Marin-Amat syndrome: A rare facial synkinesis. Indian J Ophthalmol 2007;55:402-3
5. Lubkin V. The Inverse Marcus Gunn Phenomenon: An Electromyographic Contribution. *Arch Neurol.* 1978;35(4):249. doi:10.1001/ archneur. 1978.00500280 067015
6. Oh JY, Kim JE, Kim YJ*, et al.* A case of familial inverse Marcus Gunn phenomenon *Journal of Neurology, Neurosurgery & Psychiatry* 2003;**74:**278-277.
7. Prakash M V, Radhakrishnan M, Yogeshwari A, Nazir W, Maragatham K, Natarajan K. Inverse marcus gunn phenomenon. Indian J Ophthalmol 2002;50:142-4
8. Shukla I M. Inverse marcus gunn phenomenon.Indian J Ophthalmol 1963;11:108-109
9. Pratt SG, Beyer CK, Johnson CC. The Marcus Gunn phenomenon. A review of 71 cases. Ophthalmology. 1984;91:27–30.
10. Rana PV, Wadia RS. The Marin-Amat syndrome: an unusual facial synkinesia. *Journal of Neurology, Neurosurgery & Psychiatry* 1985;**48:**939-941.