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CASE REPORT

Marcus Gunn jaw-winking syndrome: A case report

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Marcus Gunn jaw wink phenomenon or Trigeminal oculomotor synkinesis, is a congenital disorder in which the upper lid moves synkinetically in response to jaw movement during chewing. The term synkinesis describes the simultaneous movement or a coordinated sequence of movements of muscles, which are supplied by different nerves or by separate peripheral branches of the same nerve. Although it rarely manifests bilaterally, it is typically unilateral. In 1883, Dr Robert Marcus Gunn, a Scottish Ophthalmologist described a 15-year-old girl with a peculiar type of congenital ptosis that included an associated winking motion of the affected eyelid on the movement of the jaw. It is known to affect both men and women equally. This phenomenon has been reported to be a similar phenomenon affecting 2-13% of all cases of congenital ptosis. It can be congenital or acquired, for example through trauma. The Congenital Marcus Gunn jaw wink phenomenon is thought to arise from the connection between the branch of the trigeminal nerve (responsible for chewing) supplying the middle or lateral pterygoid muscle and the branch of the oculomotor nerve supplying the upper superior levator ocular defect. Here we present a case report of Marcus Gunn's Jaw-Winking Synkinesis in congenital ptosis.

Keywords: Marcus Gunn jaw wink phenomenon, trigeminal nerve, oculomotor nerve

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Introduction

Marcus-Gunn jaw wink phenomenon (MGJWP) or Trigeminal oculomotor synkinesis, is a congenital disorder in which the upper lid moves synkinetically in response to jaw movement during chewing. Synkinesis is the term used to describe the simultaneous movement or coordinated series of movements of muscles that are supplied by distinct peripheral branches of the same nerve or by different nerves. Although it rarely manifests bilaterally, it is typically unilateral. Scottish ophthalmologist Dr. Robert Marcus Gunn documented a 15-year-old girl with an unusual congenital ptosis in 1883. The girl also had an accompanying blinking motion of her eyelid on the movement of her jaw [1]. MGJWP has been reported to be a similar phenomenon affecting 2-13% of all cases of congenital ptosis. It can be congenital or acquired, for example through trauma. Congenital MGJWP is thought to arise from the connection between the branch of the trigeminal nerve (responsible for chewing) supplying the middle or lateral pterygoid muscle and the branch of the oculomotor nerve supplying the upper superior levator ocular defect. It is postulated that acquired cases following trauma are due to defective regeneration of the injured trigeminal nerve, which makes abnormal connections with the branches of the oculomotor nerve during recovery [2]. Here we present a case report of Marcus Gunn's Jaw-Winking Synkinesis (MGJWS) in congenital ptosis.

Case details

A 21-year-old female patient was presented with a complaint of abnormal eye movements and jaw clenching that had been noticeable since childhood. She reported that the symptoms have been stable and unchanged over the years, but have recently become a cause of concern. History of the presenting illness revealed that the patient had experienced a condition which was characterized by a congenital abnormality of the jaw and eye movements. She had noticed this abnormality during early childhood, but the severity and frequency have remained relatively consistent throughout her life. The patient's primary concern was the impact of these symptoms on her overall appearance and self-confidence.

The patient's medical history was unremarkable, with no significant illnesses or surgeries. There was a positive family history of similar symptoms of the mother present.

On ocular examination, the patient's visual acuity was within the normal range, with no abnormalities detected during the examination. During the examination, it was observed that the patient had unilateral ptosis (drooping of the upper eyelid) that worsened upon jaw movement (figure1a & 1b). The pupillary examination revealed no abnormalities in size, shape, or reactivity. Extra ocular movements were intact, except for the jaw-induced movement of the ptotic eyelid (Marcus Gunn phenomenon or jaw-winking syndrome). On oral examination, the patient had no difficulties with speech or swallowing. There was no evidence of any structural abnormalities or asymmetry in the face or jaw. Jaw movement was normal, except for the abnormal elevation of the ptotic eyelid, which occurred in response to jaw movements.

Based on the clinical findings and characteristic presentation, the patient was diagnosed with Marcus Gunn Syndrome, also known as jaw-winking syndrome. This

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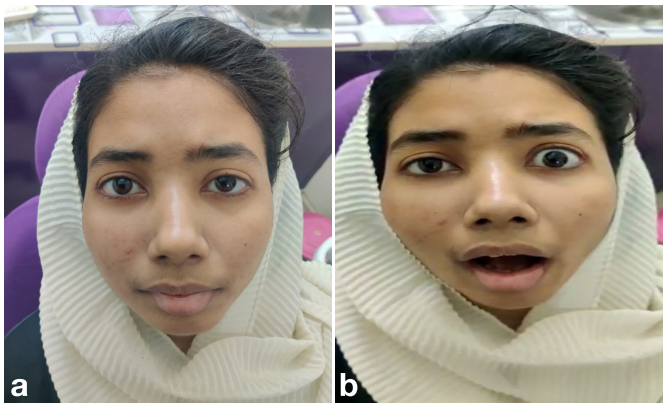

Fig. 1. a. Clinical image of a young female with evidence of unilateral ptosis in primary gaze. b. Showing unilateral ptosis that worsened upon jaw movement

syndrome is a congenital condition characterized by a miswiring of the trigeminal and oculomotor nerves, resulting in aberrant eyelid movement in response to jaw motion. As the patient's symptoms were stable and not causing functional impairments, a conservative approach with regular follow-up and monitoring was recommended.

Discussion

Marcus Gunn jaw winking syndrome is a rare condition that causes the eyelid to droop when the mouth is open. It usually occurs on its own, but in a few instances, it was coexisting with other ocular or systemic disorders, including pseudo inferior oblique overactivity, Duane's syndrome, upper rectus muscle paralysis, and iris hetrochromia. Mutations in the TUBB3 gene, which codes for beta-tubulin isotype III, have been found in a variety of conditions including MGJWS, which are characterized by anomalies of the central nervous system and oculomotor function [3] . Other names of MGJWS include Marcus Gunn ptosis, Marcus-Gunn jaw winking phenomenon (MGP), Maxillopalpebral synkinesis, Pterygoid-levator synkinesis and congenital cranial dysinnervation disorders (CCDD) [4].

According to aetiology, an abnormal connection between the motor branches of the fifth cranial nerve and the ipsilateral superior division of the third cranial nerve causes this particular type of congenital blepharoptosis. The ipsilateral levator palpebrae superioris muscle and the external pterygoid muscle, which moves the jaw towards the opposing side, are both involved in "jaw-winking" synkinesis. After ophthalmic surgery, syphilis, trauma, pontine tumours, etc., it may manifest. The congenital type endures, whereas the acquired form may spontaneously remit. Although they have been described in the literature, autosomal dominant familial instances are quite uncommon [5]. Our patient reported here with the proper familial history.

Pathophysiology is explained by the hypotheses of aberrant connection, functional interference, and atavistic reversal. The proprioceptive receptors of the external pterygoid muscle, according to electromyography studies, are the source of the impulses that cause this synkinetic movement in Marcus Gunn's syndrome. The sternocleidomastoid muscle contraction, tongue protrusion, chewing, sucking, smiling, mandibular lateral movement, Valsalva maneuver, and even breathing can cause the movement. Eyelid elevation during the closing of the mouth and clenching of the teeth has been observed in certain people with the synkinetic action between the internal pterygoid and levator palpebrae superioris (LPS) muscles [6].

Differential diagnosis for Marcus Gunn jaw winking syndrome is inverted Marcus Gunn Phenomenon, Marin-Amat syndrome, chronic progressive external ophthalmoplegia, congenital fibrosis syndrome, and myasthenia gravis. Eyelid closure on jaw opening is a sign of the inverse Marcus Gunn phenomenon, which is the opposite of the Marcus Gunn phenomenon. When the jaw fully opens or the jaw moves laterally, the eyelids close. This synkinesis is known as Marin Amat syndrome [7]. Usually, when the child is breast or bottle-fed, the mother will notice this synkinetic movement of the eyelid initially [8].

The following list of diagnostic indicators includes unilateral blepharoptosis, upper eyelid movement upon opening the mouth, jaw movement to the opposite side, chewing, sucking, and swallowing, as well as teeth clenching and protrusion. The cover test may reveal hypotropia on the affected side because of the concomitant superior rectus palsy.

The clinical examination criteria include visual acuity (it is crucial to rule out amblyopia in children and newborns), pupillary examination, cycloplegic refraction is used to detect anisometropia and rule it out. To rule out superior rectus or double elevator palsy, perform fundus examinations, extraocular motility tests, and cover tests. Determine whether the Bell phenomenon is adequate because these conditions can make it less severe. Ptosis may cause the child's chin to raise for better eyesight. It is crucial to rule out amblyopia if a child has moderate-to-severe ptosis but is not striving to lift their chin. Measurement of eyelid position in downgaze and the presence of any lid lag. Vertical palpebral fissure, which is the widest distance between the upper and lower lid [9]. The upper lid excursion measured in millimetres with the mouth open is used to grade the jaw winking reflex [10]. Mild (no more than 2 mm), Moderate (between 2 and 5 mm), and Severe (greater than 5 mm).

The management of Marcus Gunn Syndrome involves addressing the cosmetic concerns associated with abnormal eyelid movement. Treatment options may include observation and surgical intervention. As the patient's symptoms are stable and not causing functional impairments, a conservative approach with regular follow-up and monitoring is recommended. In severe cases or if the patient experiences significant social or psychological distress, surgical correction may be considered. Surgical procedures aim to improve eyelid function and reduce the Marcus Gunn phenomenon.

The prognosis for individuals with Marcus Gunn Syndrome is generally favorable. Although the condition is

lifelong, it does not typically progress or cause additional complications. The patient should be reassured that the syndrome is benign and would not have any significant impact on her overall health or life expectancy.

Author's contribution

SS: Manuscript preparation and review, Direct supervision of patient care
SM: Data Curation, Manuscript formatting
PP :Manuscript draft preparation
NH: Patient care, Manuscript formatting
PA: Supervision, Writing – original draft, Writing – review & editing

Conflicts of interest

There are no conflicts of interest.

Ethical statement

The patient has given written permission to publish this case report.

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