CASE REPORT

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Marcus Gunn Jaw-Winking Syndrome: a Case Report

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ABSTRACT

Introduction: Marcus Gunn syndrome is a rare phenomenon with very less number of cases reported in literature. It may be congenital or acquired. **Aim:** The aim of this case report was to report the clinical characteristics of Marcus Gunn patient from our Clinic. **Case report:** A comprehensive opthalmologic examination, CDVA (corrected distance visual acuity), fundus examination and photography, was conducted in Marcus Gunn patient. Clinical findings of patient presented as - chin positioned slightly upwards, extraocular motility normal on both eyes, cover test with normal findings, pupillary examination normal on both eyes. Left upper eyelid was in a lower position than the right one. On right eye, rima interpalpebrarum was 9 mm with upgaze of 13mm. On the left eye, rima interpalpebrarum was 5 mm with upgaze of 6 mm, and with open mouth, left rima interpalpebrarum was 10 mm. Visual acuity on both eyes was 1.0. Cycloplegic refraction on both eyes was +0,75 diopters (D), and Lang test was normal. In the differential diagnosis of patients with ptosis, Marcus Gunn jaw winking syndrome should be considered especially if it improves during feeding, sucking, chewing, smiling or any kind of mouth movement. In case of ptosis always do the jaw test. Have the infant bottle-feed. An older child can chew gum. Have the patient open the mouth, move the jaw from side to side, or protrude the jaw forward. **Conclusion:** Address first to treatment of any amblyopia if present - eyeglasses, patching etc., or strabismus. Think twice before deciding to operate.

Keywords: Marcus Gunn, ptosis, jaw winking.

1. INTRODUCTION

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 (1). Marcus Gunn phenomenon is type of neurogenic congenital ptosis (rare genetic disorder that is usually present at birth). It is also known as Marcus Gunn jaw winking trigemino-oculomotor synkinesis or pterygoid-levator synkinesis, and it has been observed in 2-13% of patients with congenital ptosis. It is thought to occur because of congenital mis wiring of a branch of the fifth cranial nerve into the branch of the third cranial nerve supplying the levator muscle (2). It is characterized by the movement of one upper eyelid in a rapid rising motion each time the jaw moves. In rare cases it can be present bilaterally, but jaw-winking ptosis is almost always sporadic. Familial cases with an irregular autosomal dominant inheritance pattern have been reported (3). Marcus Gunn jaw-winking syndrome is associated with strabismus in 50-60% of cases. Incidence of anisometropia 5-25% . 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. Equal prevalence among males and females. Amblyopia usually is defined as a decrease in vision of 2 or more lines on the Snellen chart (4). Treatment of Marcus Gunn phenomenon is usually not needed because the condition tends to grow less noticeable with age. Any amblyopia and strabismus should first be addressed. In severe cases, surgery with unilateral levator excision and frontalis brow suspension may be used. Unilateral suspension may lead to asymmetry, therefore some authors recommend bilateral frontalis suspension.

2. AIM

The aim of this case report is to report the clinical characteristics of Marcus Gunn patient from our Clinic

3. CASE REPORT

A 11-year old patient presented first time at our Clinic. Since early childhood, parents noticed that his left upper eyelid was in a lower posi-

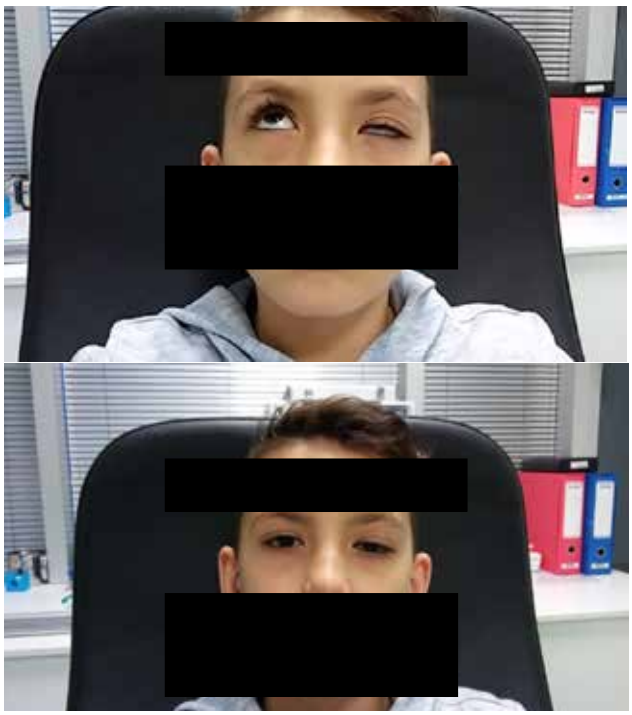

Figure 1. Ptosis of the left eye while looking straight, with worsening with upgaze

tion than the right one (Figure 1 and 2). Mother also noticed that his left upper eyelid went „up and down" while she was breastfeeding him. He was born at 40 weeks of pregnancy, with normal delivery, Apgar score was 10/10. There was no history of trauma, ptosis or any other abnormalities of extraocular movements. Ophthalmological examinations showed normal external, anterior segment and posterior segment structures. Clinically we noticed that chin was positioned slightly upwards, extraocular motility was normal on both eyes and cover test showed normal findings. Pupillary examination was normal on both eyes. Visual acuity on both eyes was 1.0 on a Snellen chart. Cycloplegic refraction on both eyes was +0,75 diopter (D). Lang test was normal. Patient´s left upper eyelid was in a lower position than his right upper eyelid. On the right eye rima interpalpebrarum was 9 mm with upgaze of 13mm. On the left eye rima interpalpebrarum was 5 mm with upgaze of 6 mm. When the patient opened the mouth, left rima interpalpebrarum was 10 mm – moderate form of the syndrome (ptosis). When ptosis is moderate and esthetically acceptable, surgery is not recommended. He presented as a healthy boy, with no medical history, no diminished sweating on the left side or anisocoria. Laboratory investigations revealed that complete blood count, blood urea nitrogen, serum creatinine, liver function tests, fasting blood sugar, serum electrolytes, serum uric acid, lactate, pyruvate, ammonia, erythrocyte sedimentation rate, C reactive protein, thyroid function tests and urinalysis were within normal limits. Brain MRI of the patient showed normal anatomy.

4. DISCUSSION

Gunn phenomenon is a rare condition that causes the eyelid to fall upon opening of the mouth. In Marcus Gunn jaw winking syndrome (MGJWS), elevation and even retraction of the affected eyelid is triggered by chewing, suction, lateral mandible movement, smiling, sternocleidomastoid contraction, protruding tongue, Valsalva manoeuvre and even by breathing.

Etiopathogenesis of Marcus Gunn jaw winking syndrome is not well defined. It is thought to occur from an abnormal branch of the trigeminal nerve, which has been congenitally misdirected in the oculomotor nerve supplying the levator muscle (5, 6). The gestational age of the infant is not a predisposing factor for MGJWS. Koelsch and Harrington (7) reported a full-term baby with MGJWS as in our case. However, premature patients have also been reported (6, 8). The necessity for correction of the jaw wink is determined by the parent, patient and surgeon. In many children, the ptosis improves with time, but there is no scientific proof that this really does take place. It is believed that, as time passes, the affected individual will come to recognize which movements are responsible for the synkinesis and learn to control or avoid them and thus minimize or mask the syndrome (9, 10). The diagnosis is generally made earlier by the child's parents or guardians who observe the synkinetic movement when feeding during infancy and some goes unnoticed till they attain teenage. However there is a hypothesis by some authors that it can be caused due to trauma of facial nerve which gives aberrant growth branches to the mandibular branch of trigeminal nerve while some authors believe that on stimulation of trigeminal nerve innervating the pterygoid muscles causes excitation of oculomotor nerve innervating the levator palpebrae superioris on the ipsilateral side causing rhythmic jerking of upper eyelid (7, 8, 10) .The attending specialist focuses the treatment, mainly in relation to correct congenital major lesions/repair of facial nerve. Treatment is usually not required but it is noteworthy to point out that the correction of the palpebral ptosis and when severe, surgery can be carried out with bilateral levator excision and frontalis brow suspension in order to avoid exaggerated permanence of the levator palpebral superioris synkinesis (11).

Figure 2. With open mouth, left rima interpalpebrarum windens and ptosis resolves

5. CONCLUSION

This report was carried out with the objective of alerting physicians to the existence of the Marcus Gunn phenomenon, which remains little known. In the differential diagnosis of patients with ptosis, Marcus Gun jaw

winking syndrome should be considered especially if it improves during feeding, sucking, chewing, smiling or any kind of mouth movement. In case of ptosis always do the jaw test. Think twice before operating. Also, authors would like to emphasize the importance of routine examinations of all children in Bosnia and Herzegovina.

- **Author's contribution**: N.Z., A.B. gave substantial contributions to the conception or design of the work in acquisition, analysis, or interpretation of data for the work. M.A.P., A.B and A.P. had a part in article preparing for drafting or revising it critically for important intellectual content, and A.P and N.Z. gave final approval of the version to be published and agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.
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