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Case Report

Marcus Gunn Jaw-Winking Syndrome: A Case Report

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Abstract: Marcus Gunn Jaw-Winking Syndrome (MGJWS) is due to aberrant innervation of the levator muscle of the upper eyelid by a branch of the motor division of the trigeminal nerve that supplies the muscles of mastication. MGJWS is often congenital in origin and usually presents unilaterally. Here we describe a case of unilateral MGJWS in a 6-year-old female patient with unilateral ptosis.

Keywords: Marcus-Gunn syndrome, pathogenesis, clinic, treatment.

INTRODUCTION

Marcus Gunn syndrome is a rare phenomenon with a very small number of cases reported in the literature. It was first described in 1883 as a unilateral ptosis associated with upper eyelid contraction and contraction of the external or internal pterygoid muscle [1]. It has been observed in 2-13% of patients with congenital ptosis [2]. Males and females are affected equally, however whether there is a predominant side affected is not clearly established [3].

PURPOSE

The purpose of this work is describing the clinical features and discussing the therapeutic options of our patient's Marcus Gunn syndrome.

OBSERVATION

A 6-year-old female patient with no previous history presented to the consultation with left upper eyelid ptosis. She was born at 40 weeks of pregnancy, with a normal delivery and to non-consanguineous parents. She had no medical or surgical history, particularly of orbital or facial trauma, but the mother noted that the left upper eyelid would rise and fall during chewing. On examination, the best-corrected visual acuity was 10/10 on both eyes. The patient had a low left upper eyelid position (Fig. 1), associated with rhythmic elevation that occurs consistently during chewing and mouth opening (Fig. 2). Upper eyelid levator function was good on both eyes with a marginal reflex distance-1 (MRD-1) at 4 mm in the right eye and a slight ptosis in the left eye with a MRD-1 at 2.5 mm. Extraocular motility was normal. The unilateral screen test was normal with centered and symmetrical corneal reflections. Anterior and posterior segment examination was unremarkable with normal *pupillary light reflex* (PLR) on both eyes.

The pediatric examination was unremarkable.

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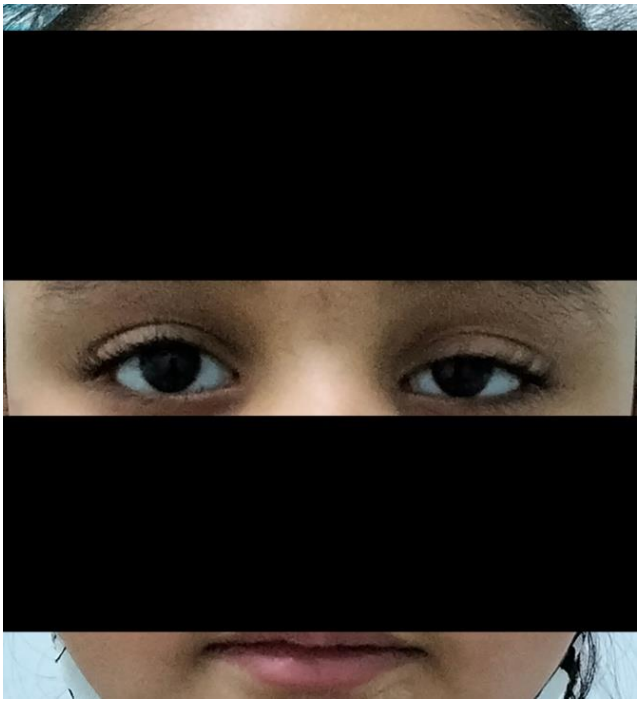

Fig. 1: Minimal left Ptosis

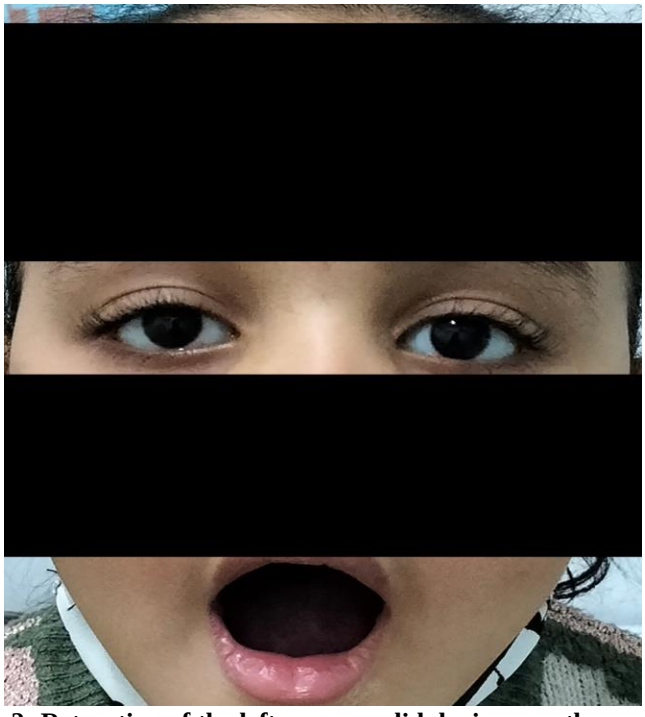

Fig. 2: Retraction of the left upper eyelid during mouth opening

DISCUSSION

The phenomenon of jaw-winking ptosis is a rare condition. It is generally unilateral and manifests as a ptotic eyelid elevates with opening of the mouth and movement of the jaw to contralateral side [4]. The Marcus Gunn jaw winking syndrome's etiopathogenesis is unclear. which has been congenitally misdirected in the oculomotor nerve supplying the levator muscle. However, some authors propose that trauma to the facial nerve may be the cause, leading to aberrant growth branches in the trigeminal nerve's mandibular branch [2]. Familial cases with an irregular autosomal dominant mode of transmission have been reported [5].

In our child, questioning revealed no history of facial trauma or similar cases in the family.

Although there were reports of bilateral cases, the majority were unilateral and occurred more frequently on the left side than the right [1], as was the case with our patient. It is associated with anisometropia

in 5-25% of cases and strabismus in 50%–60% of cases. Amblyopia occurs in 30-60% of Marcus Gunn jaw-winking syndrome patients and is almost always the result of strabismus or anisometropia, with the rare exception of ptotic eyelid occlusion [2]. Most of the time, MGJWS is unilateral and occurs alone. It is less frequently associated with ocular abnormalities like superior rectus palsy, Duane's syndrome, pseudo inferior oblique over action and iris heterochromia [6]. No ocular or extraocular anomalies were found in our case.

Surgery for MGJWS is currently a contentious topic of discussion. An aggravation of the aberrant eyelid movement, which may be cosmetically disfiguring for the patient, may result from any surgical correction that only addresses the ptosis and does not address synkinetic jaw winking. Surgery should be avoided if the ptosis is mild or the jaw-winking is cosmetically insignificant. If a correction is desired in mild ptosis, then procedures of choice include Muller muscle and conjunctival resection, a Fasanella– Servat procedure, or a standard external levator resection [7]. A unilateral levator denervation with frontalis suspension was required for patients with moderate or severe retraction [8]. This is a common surgical technique that achieves better synkinesis elimination and bilateral symmetry results [7]. Abstention from treatment in our child is justified because syncinesis and ptosis are minimal in the primary position of gaze.

CONCLUSION

In any child with congenital ptosis, Marcus Gunn's syndrome, a phenomenon that is still poorly understood, should be routinely investigated. It is probably due to aberrant innervation of the levator muscle of the upper eyelid by the trigeminal branches. This phenomenon does not usually affect the patient's quality of life when the ptosis is mild and surgical abstention is advised.

Conflicts of Interest: There are no conflicts of interest.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate consent forms from the patient's parents. In this form, the parents gave their consent for the images and other clinical information to be reported in the journal.

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