UDC: 617.7-056.7

MARCUS GUNN SYNDROME-CASE REPORT

Markovska Cheleva Vesna, Blazeska S
Eye Clinic, University "Ss. Cyril and Methodius", Skopje, Republic of Macedonia

ABSTRACT

The aim of this paper is to make a Clinical presentation of a rare case of Markus Gunn Syndrome.

A 10 month old female baby, was examined at the Department of Strabology at the University Eye Clinic in Skopje, by the ophthalmologist after the pediatric examination. On the physical examination it was noticed that rhythmic elevation of the right mild ptosis constantly occurred when the child sucked on a pacifier. The eyes were in orthophoria in the primary position. All other ophthalmological-orthoptics, neuropediatrics and laboratory investigation were in normal range.

Establish differential diagnosis from other, more severe forms of ptosis, requiring more aggressive treatment, especially if the ptosis is connected with the jaw movement and the ptosis improve with sucking, chewing or other mouth movement.

***Key words**:* Markus Gunn Syndrome, congenital ptosis, neural misdirection, ptosis, jaw winking syndrome

INTRODUCTION

Marcus Gunn syndrome or jaw winking synkinesis was first described in 1883 as a congenital unilateral ptosis with associated contraction of upper eyelid and contraction or either the external or the internal pterygoid muscle (1,2,3). In the case of chewing movements, the more or less pronounced ptosis of the affected lid decreases or even upper eyelid retraction occurs. The Synkinesis is particularly noticeable when the mandibula is pushed to the opposite side or forward.

Some patients succeed in training the position of the defective lid under control. The type of eventual treatment is dependent on the ausmas of the ptosis of the levator function and the error innervation (4).With a relatively good levator function and a low absence of innervation, a levator resection can be meaningful. In a pronounced synkinesis, this procedure is impossible, and a frontal suspension after the insertion of the muscle levator palppebrae from tarsus is recommended.

Case presentation

The case was a 10 month old baby girl, who presented at the Department of Strabology at the University "Ss.Cyril and Methodius" in Skopje, with ptosis of the right eyelid. The parents reported that ptosis was noticed within a few months after the birth. The mother of the baby had a natural childbirth without complication within the full gestation months. A physical examination revealed that head circumference was 40cm, height was 54cm and weight was 3500gr. The mother doesn't have a history of drug illness or orbital, facial or delivery trauma. She also denies the use of any drugs or alcohol. On physical examination it was noticed that rhythmic elevation of the right lid constantly occurred when the child sucked on a pacifier.The right ptosis was present, the palpebral apertures measuring 11x 3mm right and 11x4,5mm left in primary position. The right upper lid retracted rapidly some 1,5mm and fell back to its original position if the pacifier was pullout from hers mouth.

The upper lid ptosis is not covering the pupil. But the baby head posture usually is in upper position allowing good vision in all directions of the right eye and development of the stereo vision. No other abnormalities were present. In the primary position the eyes were in orthophoria. The Hirschberg test shows the symmetrical central corneal reflection in the pupil zone. The motility of the both eye bulbs in all direction were symmetrical and without restriction. The ciklоplegik skiascopy was done and there was a low hypermetropia +2D sph of both eyes. The examination of the fundus shows that there were no pathologic organic findings. The anterior segment was also normal. Laboratory investigations of the complete blood count, urea, serum creatine, liver function tests, blood sugar, serum electrolytes, thyroid function test and urine analysis were within normal limits. The neurological investigations were all without pathological signs.

This case of Markus Gunn Syndrome is still followed at the Department of Strabology at the Eye Clinic in Skopje. At this moment there is no need for surgery of the congenital ptosis because it is mild with moderate eyelid excursion.

From the heteroanamnesis, there were not similar cases in the both parents families. The uncle had a esotropia congenita as a child and he was successfully operated.

DISCUSSION

The etiopathogenesis of the Markus Gunn Syndrome is still not very known. The Markus Gunn Syndrome with jaw winking synkinesis 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 (5,6,7,8). It is believed that with time, the affected individual will recognize which movements are responsible for the synkinesis and will learn how to control or avoid them and thus minimize or mask the syndrome (9,10). In many children, the ptosis improves with time, but there is no scientific proof that this really dose take place.

Most of the patients with Marcus Gunn jaw winking synkinesis exhibited moderate eyelid excursion (10) as it was in our case. Since ptosis was mild in the most of the cases and had no association with strabismus, ambliopia or other conditions were establishing, no surgical procedures were necessary (7). The effective treatment of the severe ptosis is with surgical disabling the involved levator muscule and bilateral frontalis suspension and is some special cases disabling the involved levator muscule and unilateral frontalis suspension (10).

The Kirkham in his paper, describes unusual cases of the Marcus Gunn phenomenon with combinations with other malformations, as a congenital ptosis not after the movments of the same side lid or mandibula but with strabismus convergent position. The cause and rezone of this clinical presentation of the retraction Marcus Gunn Syndrome with ptosis and convergent starabismus has been described as misinervation of the external ocular muscles (11).

In previous reports, Markus Gunn Syndrome has been observed in 2 to 13% of patients with congenital ptosis, and although bilateral cases were reported, most were unilateral and occurred more frequently on the left side than the right (10). In the differential diagnosis of patients with ptosis, Markus Gunn Syndrom should be considered.

CONCLUSION

Markus Gunn Syndrome has to be recognized as a differential diagnostic separate conditions that has the different etiopathogenesis from the other congenital ptosis. Markus Gunn syndrome should be considered especially if the ptosis improves during feeding, chewing, sucking or any other mouth movement.

REFERENCES

1. Gunn RM: Congenital ptosis. Lancet 1883; 2:104
2. Falls HF, Kruse WC, Cotterman CW. Three cases of Marcus Gunn Phenomenon in two generations. Am J Ophthalmol. 1942; 32(116):53-59.
3. Sano K: Trigemino-oculomotor synkinesis. Neurol Med Chir. 1977; 1:29-51.
4. Ullerich K: Conservative behandlung der mandibulopalpebralen synkinesienach Markus Gunn. In modern problem der ekrankung der lider und des tranenapparates. Enke. 1978.
5. Torres MR, Calixto NJ, Oliviera LR, et al: Marcus Gunn phenomenon: differential diagnosis of palpebral ptosis in children. J Pediatr. 2004; 80:249-252.
6. Garkal RS: Markus Gunn phenomenon associated with synkineticoculo-palpebral movements. Br J Ophthalmol. 1961; 45:566-567.
7. Ghodasra DH, Nallasamy S, Binenbaum G. Congenital trigemino-abducens Synkinesis in a neonate. J APOS. 2009; 13:417-418.
8. Oesterle CS, Faulkner WJ, Clay R, Folk ER, Lucchese N, Putterman AM. Eye bobbling associated with jaw movement. Ophthalmology. 1982; 89:63-69.
9. Doucet TW, Crawford JS. The quantification, natural course and surgical results in 57 eyes with Marcus Gunn (jaw-winking) syndrome. Am J Ophthalmol. 1981; 92:702.
10. Demirci H, Fruch BR, Nelson CC. Marcus Gunn jaw-winking Synkinesis: clinical features and management. Ophthalmology. 2010; 117:1447-52.
11. Kirkham TH: Familial Markus Gunn phenomenon. Br J Ophthalmol. 1969; 53:282-283.