Marcus Gunn Jaw-winking syndrome

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KT is a 3-year-old girl and presents with a significant left congenital ptosis. Since birth, her parents observed that her ptosis dramatically subsides almost completely when she was chewing, sucking, or opening her mouth. Sometimes the ptosis becomes worse during downward gaze. She has normal developmental milestone without history of other ocular problems, and was otherwise well. There was no family history of similarly affected individuals. Examination showed that her pupils are isocoric (3mm/3mm) with normal light reflex. Her visual acuities and visual field are normal. She had a 3-4 mm left ptosis, with typical Marcus Gunn syndrome: the eyelid elevates to the normal position when her jaw is opening. All extra-ocular movements are normal and the neutral positions of eyeballs are central. Other neurological examinations are normal. Magnetic resonance imaging of brain including diffusion tensor color map is normal. Her family concerned about the cosmetic appearance of the ptosis and planned to arrange plastic surgery in the future.

Marcus Gunn first decribed this particular jaw-winking syndrome in 1883[1]. It accounts for approximately 5% of all cases of congential ptosis with various degrees. It is usually unilateral, but can present bilaterally in rare cases. Most patients with Marcus Gunn jaw-winking syndrome are identifed soon after birth by their own parents during feeding because of its typical phenomenon. When the baby or child is opening his or her mouth, sucking or chewing, the ptotic upper eyelid elevates to an equal or even higher level than the contralateral normal eyelid. Other movement with activation of ipsilateral lateral pterygoid muscle could also lead to

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Received June 24, 2009. Revised and Accepted July 10, 2009.
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eyelid elevation including jaw movement toward the contralateral side or jaw protrusion. This syndrome is sometimes associated with strabismus, other extra-ocular muscle palsy, or anisometropia, but not Marcus-Gunn pupil[2].

The pathophysiology of Marcus Gunn syndrome is now believed to be a form of abnormal synkinesis: an abnormal trigeminal motor branch innervating the lateral pterygoid muscle congenitally misdirected in the superior division of oculomotor nerve, which innervates the levator palpebrae superioris of the upper eyelid. The synkinetic speculation is supported by simultaneous contraction of the lateral pterygoid and levator muscle during voluntary activation of trigeminal nerve in electromyographic studies. Although some cases are reported in autosomal- dominant trait, and some cases are reported in pregnant women with exposure to misoprostol, most cases are sporadic and the true etiology is still unknown[2].

The correction of anisometropia, and strabismus is more important than the treatment of blepharoptosis itself. If the jaw-winking is insignificant in cosmetic, it can be ignored in the treatment of the ptosis. Because the persisting aberrant eyelid movement after levator surgery could sometimes result in normal resting position of eyelid but a more asymmetric eye-fissure during jaw-winking, the operation is suggested to be held and reserved for mild degree of jaw winking[3].

ACKNOWLEDGMENTS

We thank KT's parents for their permission to take the photography and video of KT, and their allowance to use KT's picture in scientific publication.

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