Inverse Marcus Gunn phenomenon

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A 4-month-old boy presents for routine pediatric consultation to perform children’s eye screening. He was born after an uncomplicated full term pregnancy and delivery. The neonatal period and subsequent development were normal. There is no relevant family history, other than the presence of a refractive error in his father. On physical examination was notice eyelid drooping of the left eye on
mouth opening (figure 1) that disappears on the basal position. The mother states that since his birth. The baby was alert and reactive. His weight, height and head circumference were at the 50th percentile. The pupils were reactive to light and in accommodation. Red reflexes were present bilaterally. Motility test and Hirschberg were normal. Ophthalmoscopy showed normal fundi in both eyes. The remainder of the physical examination was unremarkable including his neurologic status. These findings lead us to the diagnosis of Inverse Marcus Gunn phenomenon.

Synkinesis is the presence of a coordinated sequence of movements of muscles supplied by different nerves or by separate peripheral branches of the same nerve (1).

The inverse Marcus Gunn phenomenon is a rare congenital movement anomaly caused by cranial nerves synkinesis (2). This condition is characterized by eyelid drooping, generally the left side, during mouth opening or movements of mastication (1,3). Most cases are sporadic, although autosomal dominant mechanism had been reported (4).

The exact pathophysiology of this syndrome is not known but studies showed some nerves connection anomalies3. In 1978, Lubkin demonstrated by electromyography that the levator palpebrae muscle was inhibited or inactivated during jaw opening showing that the synkinetic movement was due to interactions at the level of the third and fifth cranial nerve complexes (4). Other synkinesis like this one have been reported, such as between fifth and sixth cranial (3).

There is another condition sometimes confused with the inverse Marcus Gunn synkinesis known as Marin-Amat syndrome. This entity also manifests as involuntary eyelid closure on jaw opening. Despite having the same physical features as inverse Marcus Gunn phenomenon, this condition is an acquired synkinesis resulting of aberrant regeneration of the facial nerve usually after a peripheral facial palsy (2,5).

Clinical findings in the examination are sufficient for the diagnosis, so complete history and physical examination is important (2). Although it does not require additional tests for confirmation, the electromyography can be useful to identify the affected nerves.

Treatment usually is not necessary for patients with a slight degree of ptosis (2,3). In this case we opted for a conservative treatment and pediatrics consultation following.

This entity is rarely described in children with only a few reported cases. This leads us to think that probably it may be underdiagnosed.

References