

MARCUS GUNN PHENOMENON

Abstract

Marcus Gunn phenomenon consists of a synkinesis between the masticatory musculature and the levator muscle of the contralateral eyelid. It is seen in patients with congenital ptosis. The phenomenon is normally unilateral, but bilateral cases are not unknown. Its exact etiology is not known. We report a case of bilateral Marcus Gunn phenomenon. The ptosis is secondary to a conflict between the oculomotor nerves and the posterior cerebral arteries. Our physiopathological hypothesis is that the motor neurons of V, possibly via “en passant” connections within the superior colliculus network, activate the nucleus of III – the homolateral portion of the central caudal nucleus in particular – causing reinforcement of its tonic action. This reinforcement leads to elevation of the contralateral eyelid, despite the paresis brought on by partial compression of the oculomotor nerve fibers.

Introduction

Marcus Gunn’s sign (or pupil) is well-known, particularly in cases of multiple sclerosis (MS), but the phenomenon of the same name is less well-known. This trigemino-oculomotor synkinesis is characterized by an upward jerk of the upper eyelid during exercise of the masticatory muscles. In most cases moderate and unilateral, this phenomenon is often partially compensated for by patients and thus occasions them only mild functional inconvenience. We report here a case of unusually late diagnosis whose effect was bilateral. Observation of this case has caused us to question the normal physiopathological explanation of the phenomenon.

Case report

A 16 year old girl was referred for ptosis of neonatal origin. The ptosis was bilateral, somewhat more marked on the left. It was not accompanied by ophthalmoplegia. The patient reported variation, not with fatigue but at mealtimes, causing a vision problem: she described it as a visual “jump” when she ate. On clinical examination, other than the ptosis, somewhat elongated facial features were noted, together with some atonality but without effect on facial motility. When the patient was asked to perform the action of diduction of the jaw, elevation of the contralateral eyelid was observed. On jaw protraction, the same elevation was seen to be bilateral. No elevation was observed during simple opening and closing of the mouth.

An encephalic MRI was basically normal, but close examination of the paths of the oculomotor nerves showed some conflict between these nerves and the left and right cerebral arteries. The two oculomotor nerves were bent at 120° by pressure from the two arteries crossing them at a right angle a few millimeters from their origin. The trigeminal nerves appeared normal.

A polygraph reading from the eyelid levator muscle and the external pterygoidians revealed, on one hand, persistence of a residual voluntary activity of the levators and on the other hand, that during lateral movement of the jaw, contraction of an external pterygoidian was accompanied by activation of the levator muscle of the contralateral upper eyelid.

Discussion

This phenomenon, first described by Marcus Gunn in 1883, is a rather rare clinical condition: 19 cases observed in 15 years among 146 congenital ptoses (Barthowski), 24 cases in 20 years (Khwarg) and 31 cases in 10 years (Bowier). It is characterized by involuntary activation of the eyelid levator on contraction of the contralateral masticatory musculature. It presents as involuntary activation of a muscle that is innervated by one branch of the oculomotor nerve as a result of voluntary activation of a different muscle that is innervated by the

root of the contralateral trigeminal nerve. It is therefore described as a crossed trigemino-oculomotor synkinesis. It is rare for the condition to be bilateral: from two of 71 cases (Pratt) to three of 24 (Khwarg).

The most usual culprit is the external pterygoidian, because the synkinesis occurs preferentially during diduction or protraction movements of the lower maxillary. Lateral displacement of the jaw causes elevation of the eyelid on the side opposite the jaw movement. This corresponds with our own observation in this case. However, this phenomenon can also be triggered by lowering of the jaw, by chewing or sucking, particularly in nursing infants (Koelsch), even by the act of smiling or protraction of the tongue (Torres).

A few unilateral post-traumatic cases have been described, as have familial associations (Kirkham, Pratt, Mrabet), but the phenomenon is almost always sporadic, congenital, presenting at birth or soon after (Koelsch). Diagnosis is sometimes late: in Bowier's series, the mean age at diagnosis was 11 years with extremes from a few months to 31 years. With the exception of one single case (Kodsi), the condition is always associated with congenital ptosis of varying severity. Marcus Gunn phenomenon is observed in 4% to 13% of congenital ptoses (Raverdy, Pratt, Barthowski). Diverse oculomotor synkinesis, however, is associated with up to 44% of congenital ptoses (Odehnl).

Other associated syndromes have also been described: for example, ambyopia (Awan), strabismus (Masany), nystagmus, fibrosis of extrinsic muscles (Yamada, Doco Fenzy), Duane's syndrome (Torres). In nearly a quarter of cases the superior rectus is also affected (Raverdy, Pratt).

Congenital ptoses are primarily caused by faulty action of the eyelid levator (striated muscle) and its annexes (suspensory ligament, internal aileron). These ptoses are normally considered as being myogenic in origin, associated with muscular degeneration (Frueh, Blin). More recent studies (Brotsky), however, tend to suggest that in some cases the problem is neurogenic and that the observed hypoplasia and fibrosis are due to a defect of innervation. The defect is assumed to be secondary to either degeneration of the fibers of the oculomotor nerve, or partial agenesis of the nucleus of III. In our observation, the ptosis is probably linked to compression of the upper contingent of the oculomotor nerve by the posterior cerebral arteries (Safran, Mrabet).

In fact, the innervation of the levator muscle of the eyelid originates in a thin branch issuing from the superior ramification of the oculomotor nerve. The fibers innervating the superior rectus and the levator initially follow a common path, diverging in the orbit. Those destined for the eyelid levator are situated in the upper part of the nerve bundle. These fibers derive from the caudal nucleus – a unique median nucleus that provides the bilateral innervation of the eyelid levators, acting within the nucleus of III. Within this nucleus, it is known (Warwick) that the fibers are crossed. For example, the upper right levator muscle is innervated by contralateral nerve fibers. The central caudal nucleus is seated in the superior colliculus in front of and above the aqueduct of Sylvius, and behind the Edinger Westphal nucleus, between the nuclei of the superior recti. As for supranuclear control of the levators, that is located in the first frontal circumvolution, above the area of control of conjugal eye movement (Serratrice, Auerbach).

The eyelid levator muscle is also served by proprioceptive innervation. This is provided by terminal branching (nasociliary nerve and frontal nerve) of the ophthalmic nerve, which is sometimes found anastomosed with branching of the superior maxillary nerve. The proprioceptive afference projects onto the mesencephalic nucleus of V (Alvarada). This in turn is located between the superior colliculus and the upper portion of the pons. It sits on the lateral edge of the mesencephalic periaqueductal gray area.

The masticatory muscles have various functions: the temporal, the masseter and the internal pterygoidian serve to elevate the lower jaw. The anterior belly of the digastric muscle and the mylohyoid serve to depress the jaw. Diduction is a function of the external pterygoidians. Bilateral contraction of these muscles

causes propulsion of the inferior maxillary. Proprioceptive innervation of these muscles arises from the V-3 and projects to the mesencephalic nucleus of the trigeminal. Motor innervation also arises from the V, whose motor root sits in the superior portion of the pons, ventro-lateral with respect to the floor of the 4th ventricle, inside and in front of the main pontal sensory root. The motor root of the mandibular nerve follows the path of the sensory root, circumvents Gasser's ganglion and exits through the *foramen ovale*. It then forms the anterior trunk of the mandibular sensorimotor nerve, which in turn divides into three branches: the temporobuccal nerves leading to the external pterygoidian muscles, the temporal and jugal mucosa, the deep temporal nerve leading to the temporal muscle, and the temporal-masseter nerve. The posterior trunk of the mandibular nerve innervates the internal pterygoidians and – via its inferior dental branch -- the mylohyoid muscle and the anterior belly of the digastric muscle.

In Marcus Gunn phenomenon, considering that the levator muscle is still functional, the observed ptosis must have a neural origin, as was the case in our observation. The synkinesis translates an aberrant re-innervation of the trigeminal system to the levator of the contralateral eyelid.

Our electrophysiological study proved that the synkinesis was crossed, because voluntary contraction of an external pterygoidian triggered involuntary activation of the contralateral levator. The phenomenon, therefore, cannot be due to ectopic re-innervation arising from the mandibular nerve. In light of the absence of activation of the levator during passive diduction of the jaw we can exclude any mechanism involving the proprioceptive pathways of the trigeminal, homolateral to the elevated eyelid. It seems important to us that the electrophysiological study revealed the existence of a voluntary activation of the levators of very weak amplitude, insufficient to cause movement.

As an explanation of Marcus Gunn phenomenon, certain authors point to agenesis of the caudal nucleus of the III, and re-innervation of the nerve trunk arising directly from the motor root of the V (Safran, Mrabet). In our observation, however, since congenital ptosis is secondary to partially damaged oculomotor nerves, this hypothesis cannot stand. Also, the crossed nature of the synkinesis does not favor a supra-nuclear origin for the phenomenon (connections between the supra-nuclear mastication centers and the eyelid levators) (Merle).

We can, therefore, suggest the hypothesis that Marcus Gunn phenomenon is associated with axonal reorganization of the motor roots of the V, via the neural networks of the cerebral trunk, the longitudinal posterior bandelette in particular (Mrabet, diagram 1). Reciprocal projections between the motor root of the V and the superior colliculus have been demonstrated (N Diaye). These projections allow the V to play a role in controlling orofacial function, as witnessed by the typical wide-eyed and open-mouthed appearance of people undergoing intense emotional experiences such as astonishment. Indeed, people commonly open their mouths to facilitate procedures requiring very open eyes, such as makeup sessions. This synkinesis between the motor root of the V and that of the III, well observed in certain species, particularly fish (Von Bartheld), has apparently not disappeared in humans.

We might therefore envisage that the V motor, perhaps via "en passant" connections within the superior colliculus network (Beyer Machule, Miller), activates the nucleus of the III – the homolateral part of the central caudal nucleus in particular – causing reinforcement of its tonic action. This reinforcement is what initiates elevation of the contralateral eyelid. It is the same process that explains other observed synkineses, either between the V and the III, with bobbing (Oesterle), between the V and the IV (Kathasi), or the V and the VI (Freedman).

Clearly, for Marcus Gunn phenomenon to appear in a congenital ptosis subject, the ptosis must be due to partial damage to the trunk of one or both oculomotor nerves, and the interneuronal circuits of the cerebral

trunk relaying nuclei of the V and III must be functional. To confirm this hypothesis, MRI of the oculomotor nerves of patients presenting the phenomenon, whether uni- or bi-lateral, would be necessary.

In many cases, particularly when the phenomenon is unilateral, symptoms are mild and patients learn to adapt by limiting the triggering motion. In the bilateral form, adaptation is more difficult. To the functional discomfort, such as that reported by our young patient, must be added the esthetic discomfort of having to eat meals in public. Surgical treatment may therefore be envisaged. It's a delicate subject, and the question should be brought up cautiously by the specialist surgical teams who are in a position to undertake it. The procedure would consist of de-insertion of the levator muscle followed by partial resection to reduce the ptosis, then suspension to the frontal muscle (Morax, Khwarg). Consideration must be given to the severity of the ptosis (Torres). In 84% of cases, clear reduction or even disappearance of the synkinesis is seen, and disappearance of ptosis in 68% (Barthowski).

In all probability, this very evident synkinesis bears witness to neuronal plasticity in organization of the cerebral trunk, and to mechanisms that are phylogenetically ancient. The phenomenon may have been recognized for more than a century, but it is very far from yielding all its secrets.