

Abnormal oculocardiac reflex in two patients with Marcus Gunn syndrome

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Abstract

Marcus Gunn phenomenon is seen in 4 to 6% of congenital ptosis patients. We report two cases of abnormal oculocardiac reflex during ptosis correction surgery. Marcus Gunn syndrome is an autosomal dominant condition with incomplete penetrance. It is believed to be a neural misdirection syndrome in which fibres of the motor division of the trigeminal nerve are congenitally misdirected into the superior pterygoid and the levator muscles. Anesthetic considerations include taking a detailed history about any previous anaesthetic exposure and any reaction to it as this syndrome has a high probability of being associated with malignant hyperthermia. It is also postulated that an atypical oculocardiac reflex might be initiated in these patients as seen in our patients, so precautions must be taken for its prevention and early detection.

Key words: marcus gunn syndrome, oculocardiac reflex, neural misdirection

Introduction

Marcus Gunn phenomenon or 'jaw – winking phenomenon' is seen in 4 to 6% of congenital ptosis patients. It is one of the most common congenital oculo-facial synkineses. These cases are diagnosed soon after birth when parents observe a ptotic lid which elevates spontaneously on sucking. A very rare variant, 'inverse' Marcus Gunn syndrome is characterized by the lid falling on opening the mouth because of inhibition of the levator muscle in association with lateral pterygoid contraction. We report two cases of abnormal oculocardiac reflex during ptosis correction surgery.

Case Report

The first patient was an 8 year old boy who presented to ophthalmology department for ptosis correction surgery. He

had unilateral congenital ptosis with no other systemic disease. Preanesthetic examination did not reveal any other abnormality. General anesthesia was induced using intravenous propofol and fentanyl 2 µg/kg. Trachea was intubated after neuromuscular blockade with intravenous rocuronium bromide and the surgery started. Anesthesia was maintained with oxygen, nitrous oxide and isoflurane. Surprisingly, as soon as the eyelid was handled, the patient developed multifocal premature ventricular contractions (PVC) and bradycardia (heart rate <45/min). On releasing the lid, the rhythm reverted to normal sinus rhythm but reappeared on resumption of surgery. The surgery was withheld and the patient was administered intravenous atropine (10 µg/kg), which restored his sinus rhythm. But still the PVCs (7-8 episodes) persisted, though the patient remained hemodynamically stable. The plane of anesthesia was deepened with increasing isoflurane and intravenous lidocaine (1.5 mg/kg) was administered, which ablated the arrhythmia. The rest of his intraoperative period was uneventful. The surgery lasted for 70 minutes. He was shifted to recovery room where the patient again had three episodes of unprovoked multifocal ventricular ectopics which resolved spontaneously without any medication.

The other child was a 12 year old boy who was also taken up for ptosis correction surgery. He had been operated 1 year earlier for the same, the perioperative period, however, was uneventful. Preanesthetic examination and investigations were all within the acceptable limits. General anesthesia was administered using standard techniques. As soon as the surgery was started and the eyelid was manipulated, he developed a

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variety of arrhythmias ranging from atrial ectopics, ventricular ectopics, bigeminy and bradycardia. The cardiac rhythm immediately reverted to normal sinus rhythm on releasing the lid. As soon as the lid was handled again, he redeveloped arrhythmias. The surgery was immediately stopped, inhalational agent (isoflurane) discontinued and intravenous lidocaine (1.5 mg/kg) administered. Cardiac rhythm was successfully restored normal. The rest of the surgery continued uneventfully and he was shifted to recovery room on completion of surgery.

Discussion

Marcus Gunn first described the syndrome in 1883.^[1] His patient had congenital ptosis of the left upper eyelid. Paradoxical lid retraction was evoked by movement of the jaw. Consequently the syndrome also became known as the “jaw-winking” syndrome. Other varieties of the Marcus Gunn syndrome have also been described. It is present in approximately 5% of neonates with congenital ptosis.^[2] It has been associated with amblyopia, anisometropia and strabismus.^[2]

Marcus Gunn syndrome is an autosomal dominant condition with incomplete penetrance.^[3] It is believed to be a neural misdirection syndrome in which fibers of the motor division of the trigeminal nerve are congenitally misdirected into the superior pterygoid and the levator muscles. This has been demonstrated on electromyographic studies. The wink phenomena may be elicited by opening the mouth, thrusting the jaw to the contralateral side, jaw protrusion, chewing, smiling or sucking.^[2,4]

There is a belief that the jaw-winking ptosis has a tendency to improve with age and this may be a reason why it is rarely seen in adults. The condition is almost always unilateral, usually involves the left side and is associated with ipsilateral superior rectus muscle underaction in approximately 80% of the cases.

Anesthetic considerations include taking a detailed history about any previous anaesthetic exposure and any reaction to it as this syndrome has a high probability of being associated with malignant hyperthermia.^[5] It is also postulated that an atypical oculocardiac reflex might be initiated in these patients

as seen in our patients, so precautions must be taken for its prevention and early detection.

A variety of stimuli arising in or near the eye especially following traction on the internal rectus or pressure on the eye ball may cause bradycardia, arrhythmias and cardiac arrest. In clinical practice this oculocardiac reflex (OCR) is most often encountered during squint surgery in children but it may also occur in eye muscle surgery, repair of detached retina, compression of Gasserian ganglion and enucleation of eye. OCR may be manifest by bradycardia, bigeminy, ectopic beats, nodal rhythm, AV block and cardiac arrest.

OCR has not been commonly reported with Marcus Gunn syndrome though there is always the probability of its occurring. There is only case report of its occurrence. Kwik reported an abnormal oculocardiac reflex in eyelid surgery of a young man^[6]. Arrhythmia occurred on eyelid manipulation and also occurred in recovery room, in the form of premature atrial contractions, wandering pacemaker and bradycardia. In our patients also, various types of arrhythmias occurred on manipulation of eyelid and were easily controlled. It should always be borne in mind and necessary preventive measures taken along with adequate cardiac monitoring to ensure a smooth perioperative course.

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