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Case Report

Hereditary Spastic Paraparesis-Use of Sugammadex Under General Anesthesia

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ABSTRACT:
Hereditary spastic paraparesis, is a rare genetically determined neurodegenerative condition. Clinical features are predominantly those of progressive lower limb weakness and spasticity. Although it has been noted that patients with progressive neurological disease may exhibit an exaggerated response to non-depolarizing neuromuscular antagonists, there is limited information in the literature. There is also limited information regarding the conduct of General anaesthesia and use of Sugammadex for reversal. Here we are presenting Anesthetic management of a patient with Hereditary spastic paraparesis under General Anesthesia posted for Open reduction and Internal fixation of Zygoma with successful Reversal form Inj Sugammadex.

Keywords- Hereditary Spastic Paraparesis, Sugammadex, General Anesthesia

INTRODUCTION:

Anesthetic management of Hereditary Spastic Parapresis (HSP). is challenging in view of Muscle spasticity with extra junctional proliferation of Acetylcholine receptors. The use of succinylcholine is contraindicated in view of risk of Hyperkalemia [1]. The variable response of Non depolarizing muscle relaxant has led to reduced dosage in such patients. Here we are reporting a case in which normal dose of Rocuronium used with successful reversal from newer drug Sugammadex. There are limited case reports with successful use of various Anesthetic agents [1-2].

CASE REPORT:

A 44 year old patient with history of Hereditary spastic paraparesis with history of fall in view of seizures presented with fracture zygoma posted for Plating of Zygoma under General Anesthesia (GA) .On Preanesthetic evaluation patient gave history of repeated fall in view of Hereditary spastic paraparesis. He was wheel chair bound with ongoing physiotherapy on Baclofen Tablets for Spasticity. There was family history of same disease and consanguineous marriage. The patient gave history of spastic paraparesis since the age of 15 years. His Bowel and Bladder habits were normal, except some Bladder precipitancy. The systemic examination was unremarkable. The airway assessment was normal.

After proper fasting status patient posted for Zygoma plating under General Anesthesia. The patient was monitored with standard ASA monitors along with Temperature and Neuromuscular monitor. 18 G IV line secured and 1000ml Ringer Lactate started. The patient was induced with Inj Fentanyl 2 mcg/kg and Propofol 2 mg/kg. The Muscle relaxant used was Inj rocuronium 1mg/kg. The Anesthesia was managed with Air/Oxygen mixture with Sevoflurane with Target MAC of 1. The whole procedure was done under Closed loop Anesthesia with Low Flow with Pressure controlled ventilator for Drager Zeus.The patient received Inj paracetamol 15mg/kg and inj Ketorolac intraoperatively as Analgesics. The Patient received 0.1 mg/kg Dexamethasone as Antiemetic. The surgery lasted for 1 hour. After TOF more than 80% , reversal done with Inj Sugammadex 2 mg/kg. The patient extubated, uneventful. The patient shifted to Post anesthesia Care unit for monitoring and eventually discharged to ward in stable condition.

DISCUSSION:

The Hereditary spastic paraparesis also known as Strumpell-Lorrain disease is one of the rarest genetic abnormality [4]. The disease mainly affects primarily the corticospinal tract with a distal to proximal retrograde axonal degeneration. Progression of the disease to the rest of the corticospinal tracts, to peripheral nerves, to the cerebellum or to the brain explains the other additional symptoms

There are various case reports of anesthesia management . The ideal anesthetic agent has not been established for general anesthesia for patients with HSP[1]. Many symptoms that are common in people with hereditary spastic paraplegia (HSP) are not caused directly by HSP but are instead caused indirectly by muscle spasticity, weakness, or hyperactive reflexes [1-4].

Here we are describing first case of Faciomaxillary surgery under GA .The challenges faced are muscle weakness which may pose difficulty in choosing Muscle relaxant. The hereditary spastic paraplegias (HSPs) are a group of con- ditions characterised by progressive weakness and spasticity of the lower limbs (1-4) . mainly characterized by variable degrees of stiffness and weakening of the muscles, with cognitive impairment, deafness, and ataxia in the more severe cases(5). At present, treatment of the HSPs is primarily directed symptomatically toward reducing muscle spasticity.The number cases reported under general anesthesia are only few in literature.. In the literature we found few cases of caesarean section and Spine surgery were found. Ours is first case with facial fracture and successful use of Sugammadex.The main concern ios delayed recovery from the effect of muscle relaxants. The use of Rocuronium and its specific reversal agent Sugammadex has reduced the incidence of disesase related complications.It has been shown that both sevoflurane and Desflurane can be used safely [1]. The regional anesthesia appears to be safe in indicated patients. As our case is done under General anesthesia with full dose of Rocuronium for case with proper perioperative Neuromuscular monitoring, Sugammadex completely reversed effect of muscle relaxants. Therefore Sugammadex appears to be safe in hereditary spastic parapresis as pathophysiology is genetic. The Sugammadex is successfully for reversing the muscle relaxant.

HSP is associated with challenges in anesthetic management as it is aneurodegenerative disease; therefore, it is important establish the pathophysiology of affected patients (5-7). First, perioperative Pulmonary complications such as include pulmonary infections. Patients with neurological diseases commonly exhibit poor respiratory muscle tone, with associated reduction of respiratory reserve (1). The administration of Depolarizing muscle relaxants, such as suxamethonium, can cause hyperkalemia (3-5). In few patients there may be hypersensitive to non-depolarizing muscle relaxants. therefore, normal or slightly increased doses may be sufficient for intubation. In this case, the patient exhibited no respiratory dysfunction. Postoperatively, the patient's respiratory muscles recovered completely from the muscle relaxant; this was reflected in clinical signs, and neuromuscular monitoring. .

CONCLUSION:

The Sugammadex can be used safely in normal dose in patient with Hereditary Spastic Paraparesis. The availability Neuromuscular monitoring in perioperative period will guide us in the proper management of Patients with this Neurogenerative condition. HSP comprised a heterogeneous group of Genetic diseases, such that there was no certainty all anesthetic drugs would have the same action among all variants of HSP. Careful perioperative monitoring and assessment of the effect of anesthetic drugs are needed. We hereby conclude that Sugammadex can be used successfully in patient with Hereditary Spastic Paraparesis.

Conflicts of Interest: None

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