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CASE REPORT

PERI-OPERATIVE MANAGEMENT OF A CHILD WITH JUVENILE MYASTHENIA GRAVIS UNDERGOING THORACOSCOPIC THYMECTOMY - A CASE REPORT

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Abstract

Juvenile myasthenia gravis (JMG) is a rare autoimmune disease of children. Thymic hyperplasia is more common in JMG and patients with refractory symptoms benefit from thymectomy. This reports a successful thoracoscopic thymectomy without lung isolation in a 13-year old with JMG. This technique allowed faster recovery with less post-operative complications. The anaesthetic challenges associated with neuromuscular blocking agents were safely overcome by non muscle relaxant technique by the use of potent opioid remifentanyl combined with anaesthetic agents.

Key Words: Juvenile myasthenia gravis, remifentanyl, thoracoscopic thymectomy

Case History

A 13-year-old girl was referred from the neurology unit with JMG with refractory muscle weakness affecting daily activities including schooling despite high doses of pyridostigmine and immunosuppressants. The condition was confirmed by electromyography and positive Acetyl Choline Receptor (ACR) antibody levels and contrast enhanced CT chest showed thymic hyperplasia.

Her lung functions showed a moderate restrictive pattern with Forced Vital Capacity (FVC) 59% of predicted but she did not have other lung diseases or other autoimmune diseases. Other pre operative investigations were normal. Her routine medications were continued as usual but anxiolytics were avoided.

Basic monitoring was established. Pre induction Train of four (TOF) count in neuromuscular monitor was four. A volume of 250 ml of normal saline was infused before induction. Patient was induced with a slow bolus of remifentanyl 1 ug/kg and propofol 2mg/kg without a significant blood pressure drop. After about 60 sec good relaxation of jaw was noted. Smooth intubation was achieved. Anaesthesia was maintained with isoflurane and N_2O in 50% O_2 and remifentanyl infusion at 0.1mic/kg/min with slight dose adjustment according to response. There was no bucking and no curare clefts in the capnograph.

She underwent thoracoscopic thymectomy in supine position, with access to mediastinum from left side. Surgery was performed under general anaesthesia and intubation was done with a single lumen endotracheal tube. Lung collapse to obtain space for dissection was achieved by creating a capnothorax of 8mmHg. Airway pressure was monitored closely along with other respiratory and cardio-vascular parameters. Bleeding was less than 50ml and patient was stable throughout the procedure. Bilateral intercostal tubes were inserted

Ten minutes before the end of surgery morphine 6 mg was given to cover the postoperative pain. At the end of surgery TOF count remained four. Remifentanyl infusion was stopped. She was successfully extubated and sent to ICU for further care. By post-op day 4 her pyridostigmine dose was reduced and was discharged home on day 5.

Discussion

JMG is defined as myasthenia gravis in children less than 16 years of age. It is the commonest type of myasthenia in children, other types being neonatal and congenital myasthenia.[1] In JMG, it is common to find a hyperplastic thymus gland compared to adults.[1]

Thymectomy is indicated in patients with MG who have evidence of thymoma as well as in patients with non thymoma related refractory MG[2]. In recent studies it has been shown to increase the probability of clinical improvement in 50%, being asymptomatic and achieving medication free remission in further 35% of patients[2]. Nevertheless the place of thymectomy in pre pubertal JMG, ocular MG and in patients over 60 years is questionable and there is no consensus up to date[3]. Thymectomy undertaken early in the course of the illness has shown to be more effective than late[2].

Thymectomy can be performed by open as well as thoracoscopic methods. Trans sternal – transcervical approach which needs median sternotomy and neck dissection, despite being highly invasive, is the standard procedure as it enables maximum exposure to thymic tissue. However thoracoscopic thymectomy is an equally effective method which has significantly low morbidity and mortality when compared to conventional method even though it lacks the accessibility to the neck. Most worried side effects of surgery are phrenic nerve and recurrent laryngeal nerve injuries, myasthenic crisis and hospital acquired chest infections.

Myasthenia gravis being a disease of the neuromuscular junction, brings special anaesthetic challenges. It impairs the functions of neuromuscular system specially the respiratory functions and the medications used to treat MG interact with anaesthetic drugs. JMG will add challenges associated with paediatric age group and its rare occurrence gives minimum experience. Patients with JMG are sensitive to non depolarizing muscle relaxants and there is potential interaction of anticholinesterases administered as therapeutic agents, hence many anaesthetist now prefer non muscle relaxant anaesthetic techniques. Even though these patients are sensitive for muscle relaxation properties of anaesthetic agents and opioids, high doses are required to achieve excellent intubating conditions without use of muscle relaxants. Hence, achieving good relaxation while minimizing the side effects of high doses of anesthestic agents is a challenge to the anaesthetist. We used remifentanyl and propofol at induction without muscle relaxants. Remifentanyl being a potent drug and having synergistic properties with propofol was able to provide excellent intubating conditions at a dose of 1 ug/kg/min combined with usual induction

dose of propofol. Remifentanyl is metabolized by tissue and plasma esterases making it ultra short acting which is very important to achieve quick recovery in patients with muscle disorders. Sudden hypotension associated with above combination of drugs was minimized by a fluid bolus prior to induction. To avoid further requirement of muscle relaxants we continued the remifentanyl infusion at a rate of 0.1mic/kg/min. Non muscle relaxant technique is popular in paediatric population. Trans-sternal and thoracoscopic thymectomies have been successfully performed in JMG with anaesthetic agents combined with remifentanyl or thoracic epidural analgesia or both without any muscle relaxants.[1,4,5]

Since lung isolation with a cumbersome double lumen tube was not used, conventional endotracheal tube was easily tolerated with isofurane and remifentanyl infusion. All the other anesthetic challenges associated with one lung ventilation such as desaturation, shunting, atelectasis were minimized as surgical access was achieved by only partial lung collapse with created capnothorax.

Conclusion

Minimally invasive thoracoscopic thymectomy without lung isolation combined with short acting anaesthetic agents without muscle relaxants results in excellent recovery and long term treatment in JMG.

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