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A CASE OF MYASTHENIA GRAVIS WITH OBSTRUCTED INGUINAL HERIA.EMERGENCY GROIN EXPLORATION MANAGED SUCCESSFULLY WITH COMBINED SPINAL EPIDURAL ANESTHESIA.

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Abstract : Myasthenia gravis is an autoimmune disorder affecting post-synaptic Ach receptor presenting with muscular weakness involving skeletal muscles, muscles of eyes, bulbar muscles etc. Myasthenic patients pose unique anesthetic challenges. They are sensitivite to non-depolarizing muscle relaxants,there may be a need for prolonged post-operative mechanical ventilation and possibility of being a chronic respiratory cripple. Here I present a case of myasthenia gravis patient coming for emergency groin exploration for obstructed inguinal hernia.

Keyword :myasthenia gravis, regional anaesthesia, emergency, preoperative preparation

Case History : Kannan 38year old male, a k/c/o:myasthenia gravis for 8 years, c/o weakness of both upper and lower limbs towards the end of the day. H/O easy fatigability present for 8 years, presented with h/o right inguinal swelling for 6years. Now admitted with pain over right groin and the swelling not reducing in size for 2 days. History of fever for 2 days. Nausea , vomiting and obstipation present for 1 day. Fever was of moderate grade, not associated with chills or rigor or any rashes. Three episodes of vomiting, vomitus containing undigested food, not blood tinged or bilious. Right groin pain was present since two days and generalized abdominal pain for one day. Pain was dull, aching in nature, not radiating to back. History of mild abdominal distension present. No history of diarrhoea, hematemesis or melena. No history of dysuria or haematuria. No history of trauma to abdomen. No history of difficulty in swallowing, breathing ,drooping of eyelids, drooling of saliva , any visual disturbances or hoarseness of voice. No history of cough or expectoration now. History of last oral intake was 24 hrs. back.

Past history: History of myasthenia gravis for the past 8 years and patient was on regular treatment with tab. pyridostigmine 250 mg qid and tab.prednisolone 20mg b.d. History of admission to icu with ?myasthenic crisis mechanically ventilated for three weeks, H/O: tracheostomy done for prolonged ventilation and successfully weaned from ventilators 8 months back. No documentation of discharge summary was available with the patient. No histories of any other medical illnesses like Diabetes, Hypertension, bronchial asthma, epilepsy, pulmonary tuberculosis. No history of previous surgery or drug allergy. No history suggestive of any other autoimmune illness.

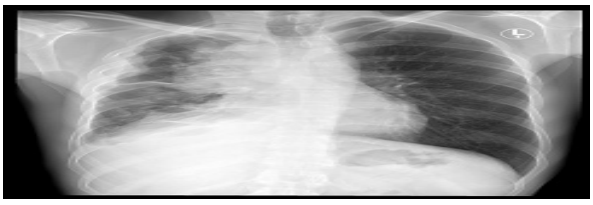
Family history: No history of any similar disease in the fa mily.
Personal history: patient takes mixed diet, No history of smoking or alcohol intake.

General Physical Examination

Thin built, height 158 cm, weight 60 kg. Patient conscious, oriented, febrile, no pallor, no clubbing, no jaundice, no cyanosis or pedal oedema. A scar was present over the midline in the neck (tracheostomy). PR= 98/min, BP=110/70 mm Hg, RR=20/min and Temp=100 F.

Systemic Examination:

CVS – S1S2 +/ no murmurs,RS – Normal vesicular breath sounds +. Occasional rhonchi+ over both axillae,SpO2 at room air = 98%.breath holding time-20 seconds. Cough test was negative. P/A –distended, generalized tenderness+, tenderness more over right groin. no guarding or rigidity. Spleen, Liver not palpable. Preoperative examination: Airway: modified mallampati Class II, Neck movements, Spine, Dentition normal. Lab evaluation: Hb% =11.5gm%,TLC=10,800cells/cu.mm, Platelet= 1.5L/cu.mm, RBS= 140 mg/dl, Urea = 36mg/d, Creatinine = 0.8 mg/dl. Chest x-ray PA view showed thymic enlargement. Electrocardiography was Normal.



Patient was catheterised and urine output was 100 ml/hr. inj.Ranitidine 150 mg i.v and inj.metoclopramide 10 mg i.v was given. Surgery planned: Right groin exploration. Anesthetic plan: Combined spinal epidural anaesthesia.

Pre-operative preparation:

Patient communicated about possibility of prolonged post-operative ventilation. Icu bed with ventilator was kept ready. Serum electrolytes level was checked and ensured that it was normal. We ensured that patient has taken routine tab. pyridostigmine via ryle's tube on the day of surgery. Approriate sized endotracheal tubes(also smaller sized #6.5 , 7,7.5, 8) was kept ready, in view of possible tracheal stenosis.

Premedication:
inj.midazolam 1 mg i.v. inj. Hydrocortisone 100 mg i.v,inj. ondensetron 6 mg i.v was given. Epidural block was done at T 10-T11 intervertebral space with 16 G tuohy needle and 18G epidural catheter placed in epidural space, tip at approximately at T9. Epidural test dose was given. Sub-arachnoid block was given at L3-L4 intervertebral space and 3 ml of 0.5%bupivacaine was injected. The level of sensory blockade was T6. Intra operatively, apart from routine pulseoximetry, noninvasive BP, ECG, temperature and urine output monitoring, respiratory effort was monitored closely. Intra operative hypotension was treated with fluids and Inj. ephedrine 6mg i.v. Epidural catheter activated after 1 hr of start of the procedure and totally 9 ml of 0.5% bupivacaine was given in 3 ml aliquots. Intra operative period was uneventful. By avoiding a general anaesthesia, we have avoided the use of muscle relaxant and prolonged post-operative ventilation. Surgery lasted for 2 hours 30 minutes. Intra-operatively 2500ml of crystalloids was infused and the urine output was 200ml. oxygen was supplemented intra-operatively at 5L/min. The operative finding was omentum found as content of obstructed inguinal hernia.

Post-operative care:

Oxygen supplementation:
Patient was supplemented with 5L/min oxygen by face mask for 12 hours. No significant desaturation was observed.

monitoring:
Patient was monitored for respiratory rate and o2 saturation. all monitoring done intraoperatively was continued in the postoperative period. serum electrolytes and renal function tests were done and ensured that it was within normal limits.

Analgesia:
Epidural analgesia with 0.125% bupivacaine with 2 mic/ml fentanyl solution as continuous infusion at 6 ml/hr. Patient was pain-free at this dose.

Anticholine esterase therapy:
Patient's routine dose of anti-choline esterase was administered via ryle's tube after 12 hours postoperatively. **Peri-operative steroids**: inj. Hydrocortisone 25 mg IV was infused every 8 hours for 24 hours. Usual preoperative dose was then continued via ryle's tube.

Discussion
MG is an autoimmune disorder caused by circulating antibodies to nicotinic acetylcholine receptors at the neuromuscular junction . The antibodies reduce the numbers of receptors available for muscular stimulation by acetylcholine, apparently by blockade and increased degradation of the receptor. There is no correlation between the antibody titre and the severity of the disease. Up to 25% of patients have a concurrent thymoma, and about 10% have evidence for other autoimmune diseases. More recently, antibodies against the MuS receptor, which is involved in the formation of the neuromuscular junction, have been identified in MG patients. The clinical course of myasthenia gravis is marked by periods of exacerbation and remission. Muscle strength may be normal in well-rested patients, but weakness occurs promptly with exercise, infection, electrolyte abnormalities, pregnancy, emotional stress, and surgery. Antibiotics, especially the aminoglycosides, can aggravate the muscle weakness. Weakness of pharyngeal and laryngeal muscles results in dysphagia, dysarthria, and difficulty handling saliva. Patients with myasthenia gravis are at high risk of pulmonary aspiration of gastric contents. Myocarditis can result in atrial fibrillation, heart block, or cardiomyopathy. Patients with myasthenia gravis often require ventilatory support after surgery. Therefore, it is important to advise these patients during the preoperative interview that they may be intubated and ventilated when they awaken.

Criteria that correlate with the need for mechanical ventilation during the postoperative period following transsternal thymectomy include disease duration of longer than 6 years, the presence of chronic obstructive pulmonary disease unrelated to myasthenia gravis, a daily dose of pyridostigmine higher than 750 mg, and a vital capacity less than 2.9 L. These criteria are less predictive of the need for ventilatory support following transcervical thymectomy, indicating that this less invasive surgical approach produces less respiratory depressionBy avoiding general anesthesia, the risk of aspiration, the need for prolonged ventilation and polypharmacy in a case of myasthenia gravis would be avoided. The Myasthenia Gravis Foundation of America classifies the clinical presentation according to a modified scale initially presented by Osserman and Genkins : class I (ocular muscles only); class II (symptoms plus mild generalize weakness); class III (eye plus moderate weakness); class IV (eye plus severe weakness); and class V (intubation, ventilation). This case belongs to class II according to this scale. The specific diagnosis involves blood tests for antibodies; electromyographic recordings; cholinesterase inhibitor test (edrophonium test); and imaging (to identify thymoma). Cholinesterase inhibitors (neostigmine, pyridostigmine) and corticosteroids andimmunosuppressive drugs (cyclosporine, azathioprine) are used for the treatment . In some patients, plasmapheresis is indicated to decrease circulating antibodies (four to eight treatments over 2 weeks). In addition, thymectomy is performed in most patients leading to improvement in clinical symptoms in most patients, and in some patients to a complete remission, which sometimes require several months to determine. In general, MG is not a progressive disease, and the symptoms may fluctuate or even spontaneously disappear within several years. With appropriate therapy, the life expectancy is normal.

Anesthetic challenges in patients with myasthenia gravis:
Respiratory and bulbar functions should be carefully evaluated during the preoperative evaluation. Although the respiratory drive and the CO2 response are usually intact, patients may have a profoundly diminished vital capacity. Efforts should be made to determine the absence of a larger thymoma, which may cause tracheal compression or even airway collapse during induction of general anaesthesia. Cardiac arrhythmias and myocarditis have been described in MG patients, suggesting that preoperative ECG recordings should be part of the preoperative work-up. While assessing the airway, the degree of bulbar involvement requires special attention. espesially in cases like ours, where there is history of prolonged intubation, tracheal stenosis is a possibility. so in case of elective surgeries it is mandatory to take CT neck to look for tracheal diameter. in case of tracheal stenosis, anesthetist should be prepared with asmaller sizes of endotracheal tubes. Medical management before surgery aims to optimize the patient's muscular function. The decision to continue or hold immediate preoperative anticholinergic medication is based on an individual basis. Patients with severe MG should receive preoperative anticholinergic medication despite the increased risk of potentiation of vagal responses and decreasing metabolism of local anesthetics and succinylcholine in the intraoperative period. Preoperative plasmapheresis has been shown to be very effective preoperatively in severe MG for improving pulmonary function. The reduction in plasma esterases by plasmapheresis, however, prolongs the duration of action of drugs like succinylcholine, esmolol, mivacurium, and remifentanil. Sedative drugs may be dosed very carefully or even avoided completely in MG patients because of the risk for respiratory compromise.

Premedication: Myasthenic patients may have little respiratory reserve and hence depressant drugs for preoperative premedication should be used with caution, and avoided in patients with bulbar symptoms, but an anticholinergic may be useful. Hydrocortisone 'cover' should be given to those on long-term corticosteroid therapy. anti emetics and anti aspiration prophylaxis should be

given. Patients can safely undergo regional anesthesia, and it seems to be the preferential regimen in MG patients whenever possible. The doses of local anesthetics (ester types and amides) should be reduced in patients receiving cholinesterase inhibitors to avoid prolonged blocks and the potential for a myasthenic crisis. If general anesthesia is planned, induction and maintenance may involve intravenous agents, such as propofol, thiopental, and etomidate, and volatile anesthetics. Volatile anesthetics exert muscular relaxation by impairing the neuromuscular transmission, with isoflurane being twice as potent as halothane in MG patients, and independent of the severity of the disease .

Neuromuscular blockers may be used cautiously in MG patients, but their differential effects need to be considered. Compared with patients without MG, succinylcholine has decreased efficacy at low doses and a higher incidence of phase-two block at high doses. At a dose of 1 to 1.5 mg/kg, succinylcholine can be expected to achieve clinical efficacy and duration as expected in a normal patient (1.5–2.0 mg/kg is safe for rapid sequence induction in MG patients) . In contrast, MG patients show high sensitivity to nondepolarizing muscular blocking agents. The reduced number of acetylcholine receptors may require only 10% of the normal dose to elicit a reasonable neuromuscular block. Even then the duration of the block may be prolonged, especially if long-acting drugs, such as pancuronium or rocuronium and medium-acting drugs, such as vecuronium (ED95 56%) or atracurium, are preferred, although producing a longer than normal block. The effects of reversal drugs are unpredictable, especially in patients on chronic anticholinesterase treatment, and the excessive administration may precipitate a cholinergic crisis (generalized muscle weakness, bradycardia, increased secretion, and gut motility). The variability in sensitivity to muscular blocking agents does not correlate with the clinical severity of MG, but some report predictability by perioperative monitoring of the train-of-four, which may guide the dosing regimen. In addition, drug interactions need to be considered, because substances that are known to exacerbate the clinical symptoms of muscular weakness (aminoglycosides, vancomycin, quinidine, ester-type local anesthetics, furosemide, calcium antagonists, b-blockers) may also amplify the clinical effects of neuromuscular blocking agents .

The two most important concerns during the postoperative period are mechanical ventilation and sufficient pain management. Postoperative pain control may be a challenge because of the sensitivity of MG patients to any drug with respiratory depressant effects, such as opioids and benzodiazepines. Regional anesthesia may be beneficial for the management of postoperative pain. Neuraxial techniques using epidural opioids were shown to be safe and effective, reduced the overall requirements for systemic narcotics, provided excellent pain relief, and resulted in improved postoperative respiratory function in patients with MG.

It is recommended to resume the anticholinesterase therapy as soon as possible after surgery. The anaesthetic management of the myasthenic patient must be individualized to the severity of the disease and the type of surgery. The use of regional or local anaesthesia seems warranted whenever possible. Whenever local or regional anaesthesia is used, the dose of the local anaesthetic may be reduced in patients to decrease the possible effects of anaesthetics on neuromuscular transmission. This may be particularly important when ester local anaesthetics are administered to patients receiving anticholinesterase therapy (inhibit plasma cholinesterase). General anaesthesia can be performed safely, provided the patient is optimally prepared and neuromuscular transmission is adequately monitored during and after surgery.

Anaesthetic techniques: Two techniques have been recommended for general anaesthesia in the myasthenic patient, although none is superior. Because of the unpredictable response to suxamethonium and the marked sensitivity to non-depolarizing muscle relaxants, some anaesthetists avoid muscle relaxants and depend on deep inhalational anaesthesia, for tracheal intubation and maintenance of anaesthesia.

These agents allow neuromuscular transmission to recover, with rapid elimination of these agents at the end of surgery. In theory, desflurane and sevoflurane may offer some advantages, due to their low blood solubility. Sevoflurane is probably superior to desflurane, due to its lower incidence of excitatory airway reflexes during inhalational induction. However, others utilize a balanced technique which includes the use of muscle relaxants, without the need for deep inhalational anaesthesia, titrating small doses (10– 25% of the ED95) of intermediateacting relaxants monitoring with a peripheral nerve stimulator for both intubation and surgical relaxation, if required. The decision as to whether to reverse residual neuromuscular blockade at the end of surgery is controversial. Some argue that the presence of anticholinesterases and antimuscarinics will confuse efforts to differentiate weakness due to inadequate neuromuscular transmission from cholinergic crisis in the recovery room. Some prefer spontaneous recovery and extubation when the patient has demonstrated adequate parameters for extubation (head-lift, tongue protrusion).

Similarly, the presence of fade (T4/T1 < 0.9) in the preanaesthetic period predicts decreased atracurium requirements in patients with MG. This technique, along with preoperative pulmonary function testing, may be useful in determining preoperative baseline function. Total intravenous anesthesia (TIVA) for the management of myasthenics has been reported. Haemodynamic instability in older patients makes this approach more difficult, whereas younger patients usually tolerate it. The use of remifentanil as part of TIVA may alleviate some of the hemodynamic instability.

When possible, many clinicians prefer to utilize regional or local anesthetic techniques. Regional techniques may reduce or eliminate the need for muscle relaxants in abdominal surgery. Epidural techniques offer the advantage of postoperative pain control with minimal or no opioid use. Ventilatory function must be monitored carefully after surgery. There are few tests of neuromuscular function which correlate with adequate ventilation. It has been shown recently in normal patients that many of the recommended tests such as maintained response to tetanic stimulation of a peripheral nerve can return to normal, while the pharyngeal and neck muscles necessary to protect the airway can still be partially paralysed. The different response of peripheral versus bulbar muscles may be more evident in myasthenic patients, particularly those suffering from bulbar and/or respiratory muscle weakness. It is essential that sustained respiratory muscle strength be confirmed before extubation of the trachea and resumption of spontaneous ventilation. Myasthenic patients may be at increased risk of developing postoperative respiratory failure – following trans-sternal thymectomy, up to 50% of patients require prolonged postoperative ventilation.

Four risk factors have been identified: (Anesthesiology 980; 53: 26-30)

Duration of myasthenia gravis for longer than six years (12 points). (Duration of MG proved to have the greatest value in predicting the need for ventilatory support). A history of chronic respiratory disease other than respiratory dysfunction directly due to MG (10 points).

A dose of pyridostigmine greater than 750 mg per day, 48 hr before operation (8 points). A preoperative vital capacity < 2.9 L (4 points) These risk factors were weighted according to their significance as predictors; a total score of more than 10 points is identified as those patients likely to need postoperative pulmonary ventilation for more than three hours. This patient having history of myasthenia for 8 years and taking 250 mg of pyridostigmine, the score is 20 points. This predicts that there is need for post-operative ventilation.

Conclusion: Myasthenia gravis is an autoimmune disease of neuromuscular junction. Ideally careful neurological consultation for optimizing preoperative drug therapy should be done. Pre-operative pulmonary function tests should be done to assess the need for post-operative ventilation. During emergency surgery, bedside pulmonary function tests are rough guides. Wherever possible regional anaesthesia is preferred to avoid using neuromuscular blocking agents and getting into controversy of whether to reverse or not. Perioperative steroid supplementation, assessing for presence of other autoimmune disorders, avoiding aminoglycoside antibiotics, being prepared to mechanically ventilate the patient, should the necessity occurs, forms the mainstay of anesthetic management in myasthenic patients.

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