DIFFICULT TO WEAN OFF VENTILATORY SUPPORT IN MYASTHENIC PATIENTS UNDERGOING THYMECTOMY

Zia Ali, Sarfraz Khan Janjua

Combined Military Hospital, Rawalpindi

INTRODUCTION

Myasthenia gravis is a chronic autonomic disorder characterized by varying degrees of skeletal muscle weakness. The name myasthenia gravis, which is Latin and Greek in origin, literally means, "grave muscle weakness."[1]. Respiratory insufficiency is the main complication in inadequately controlled (myasthenic crisis) or over medicated patient (cholinergic crisis) along with ptosis, excessive secretions and generalized muscle weakness. Myasthenic patients usually respond to medical management and ventilatory support if required is gradually weaned off however in some cases weaning off the ventilator may be very difficult. Increased age (> 50 years), baseline severity of myasthenic weakness and complications appear to be the principle factors associated with prolonged crisis leading to difficulties in weaning off the ventilatory support [2]. We are reporting a well controlled patient of myasthenia gravis under going thymectomy who required prolonged ventilatory support postoperatively and was difficult to wean off.

CASE REPORT

Our patient, a 70 years old lady, presented with 02 months history of easy fatigability, dysphagia and diplopia. Her symptoms aggravated in intensity as the days passed. On clinical examination she had bilateral ptosis, diplopia and upper limb raising test (muscle fatigue test for 1 minute) was positive. Peak expiratory flow rate was 150 liter/minute and she had motor power of grade III in all the limbs with intact sensory system. Routine lab investigations i.e, full blood count, ESR, blood sugar, urea, electrolytes, liver function tests and thyroid function tests were with in normal limits.

Her X-ray chest showed widening of the madiastinum and CT scan chest -revealed a well defined triangular soft tissue in anterior madiastinum (Fig). She was on tab pyridostigmine 60 mg 6 hourly and tab prednisolone 15 mg 8 hourly.

Thymectomy was performed through sternal split incision under general anaesthesia with minimal use of muscle relaxants (atracurium besylate). Analgesia was provided through thoracic epidural catheter with 0.25-0. 125% bupivacaine and inj tramadol 100mg/20ml. Post operative recovery was satisfactory and patient was extubated and shifted to ICU. Postoperatively she was given tablet azathioprine 50 mg OD, tablet prednisolone 15 mg 8 hourly and tablet pyridostigmine 60 mg 6 hourly. On second post operative day the patient became restless and tachypnoeic and was placed on ventilator. On 7th post operative day her tracheostomy was performed. She was assessed daily to wean off and was given repeated trials but she used to exhaust and mechanical ventilation had to be resumed. Frequent tracheobronchial toilets, chest physiotherapy and psychological motivation were continued. The dose of pyridostigmine was reduced due to excessive salivation and bradycardia. Her plasmapheresis was done in two phases but she did not show much improvement. After that intravenous immunoglobulins 400 mg/kg body weight daily were infused for three consecutive days along with intensive nursing care, nutritional support and chest physiotherapy. She was placed on adaptive support ventilation, gradually encouraging her own ventilatory efforts. On 23rd post operative day she was placed on spontaneous ventilatory mode with

Correspondence: Lt Col Sarfraz Khan Janjua, Department of Anaesthesia, Combined Military Hospital, Rawalpindi.
Email: sarfrazjanjua@hotmail.com

Received July 14, 2007; Accepted Nov 16, 2007

continuous pressure support which was gradually decreased. Ultimately she was weaned off successfully on 43 post operative day. Tablet pyridostigmine, prednisolone and azathioprine were continued and she was referred to the neuro physician for follow up.

DISCUSSION

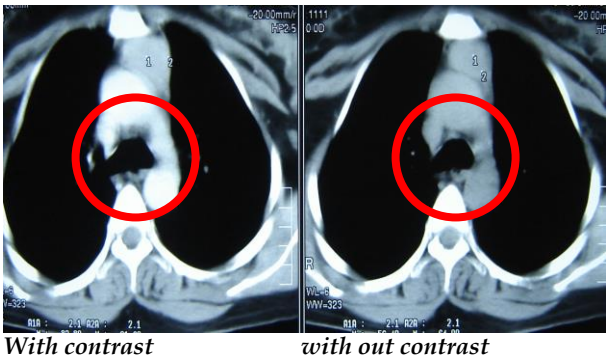
Myasthenia gravis is an autoimmune neuromuscular disease leading to fluctuating muscle weakness and fatigability. Weakness is typically caused by circulating antibodies that block acetylcholine receptors at the post-synaptic neuromuscular junction and inhibiting the stimulative effect of the neurotransmitter acetylcholine.

Myasthenia gravis is treated with cholinesterase inhibitors, corticosteroids, immunosuppressants like azathioprine and cyclophosphamide which have steroid sparing effect as well, plasmapheresis, transfusion of immunoglobulins and in selected cases thymectomy [3].

Myasthenic patients for thymectomy may require post operative ventilatory support which is predicted by the evaluation of muscle groups affected especially the respiratory muscles, drug therapy, co existing illness, disease duration > 6 years, pyridostigmine dose > 750 mg /day, concomitent pulmonary disease, peak inspiratory pressure < 25cm H2O, maximum expiratory pressure < 40cm H2O and vital capacity < 30ml/kg [4].

Our patient had no comorbid disease, the disease duration was less than 6 years and pyridostigmine dose was less then 750 mg /day. However she had thymoma confirmed histopathologically and developed myasthenic crises postoperatively. Transfusion of immunoglobulins was carried out after plasmapheresis. Plasmapheresis after immunoglobulins therapy is not recommended as it can wash away the immunoglobulins.

The patients who have previous history of crisis, oropharyngeal weakness, or thymoma are more prone to respiratory



With contrast *with out contrast*

Figure: CT Scan Chest shows a well defined triangular soft tissue in anterior madiastinum.

failure. Possible triggers include infection, aspiration, physical and emotional stress, changes in medications, myopathies secondary to electrolyte imbalance and neuropathies [5]. All these factors lead to difficultly in weaning off from ventilator. Infections are associated with 30% to 40% of crisis; the most common are upper respiratory tract viral infections, bronchitis and bacterial pneumonia. Aspiration pneumonitis is associated with 10% of crisis. Anaemia can also increase weakness, and transfusions are recommended when haematocrit values are under 30%. In patients with chronic carbon dioxide retention (suggested by elevated serum bicarbonate levels), PCO2 should be kept above 45 mmHg to avoid alkalosis and bicarbonate wasting, which makes weaning more difficult.

Duration of intubation is an important predictor of functional outcome after crisis. Thomas, et al have described that 77% of patients intubated for more than 2 weeks had functional dependence at discharge compared with 36% of patients intubated for less than 2 weeks [6].

Respiratory failure is a life threatening complication of myasthenia gravis that necessitates care in an intensive care unit. Two possible mechanisms can result in life threatening respiratory compromise in myasthenic patients. First is the respiratory muscles weakness which can lead to impaired lung expansion, hypoventilation and weak cough. Secondly, oropharyngeal weakness can result in inability to clear air ways and

aspiration of secretions can occur. Fibreoptic bronchoscopy is useful in such circumstances and airway patency can be restored under vision by redirected suctioning [6].

These patients must be followed up closely as thymectomy is neither a definite treatment nor always complete, even redo thymectomy may be required in some cases and the patient may have to be maintained on medical therapy.

REFERENCES

1. Nicolle MW. Myasthenia gravis. Neurologist. 2002; 8(1): 2-21.
2. Thomas CE, Mayer SA, Gungor Y, Swarup R, Webster EA, Chang I, et al. Myaesthenic crises: clinical features mortality, complications and risk factors for prolonged intubation. Neurology. 1997 May; 48(5): 1253-60.
3. Berroushot J, Baumann I, Kalischewski P, Sterker M, Schneider D. Therapy of myasthenia crisis. Crit Care Med. 1997; 25(7): 1228-35.
4. Naguib M, el Dawlatly AA, Ashour M, Bamgboye EA. Multivariate Determinants of the need for post operative ventilation in myasthenia gravis. Can J Anaesth. 1996; 43(10):1006-13.
5. Isabel P, Nieto, MD, Juan A, Pedro P, Robledo MD, Manuel C. Multivariate determinants of the need for postoperative ventilation in myasthenia gravis. Ann Thorac Surg. 1999; 67: 1568-71.
6. Thomas C, Mayer S, Gungor Y, Swarup R, Webster EA, Chang I, et al. Myasthenic crisis: clinical features, mortality, complication and risk factor for prolonged intubation. Neurology. 1997; 48(5): 1253-60.