Nursing care plan for patients undergoing thymectomy for Myasthenia Gravis

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ABSTRACT

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One half of cortical thymoma patients develop myasthenia gravis (MG), an autoimmune disease affecting the voluntary muscles, while 15% of MG patients have thymomas. Thymectomy has been a mainstay in the treatment of myasthenia gravis and the management of such surgical patients is extremely demanding both at the physician's and at the nurse's level. In this paper we review some of the nursing interventions for patients with MG undergoing surgical removal of the thymus gland.

INTRODUCTION

Myasthenia gravis (MG) is a chronic autoimmune disorder caused by a loss of acetylcholine receptors (AchR) in the postsynaptic neurons of the neuromuscular junction (NM)[1]. The very name of the disease, which is both Greek and Latin in origin, means "grave muscle weakness". Patients with MG typically present with considerable weakness and abnormal fatigue of the skeletal (voluntary) muscles, which increase during periods of activity or repetitive use and improve after periods of rest. Although virtually any voluntary muscle may be affected by the disease, ocular muscles, those controlling facial expression and swallowing are most frequently involved.

Women are affected more often than men with a ratio 3:2[2]. Also, women younger than the age of 40 are at increased risk for developing MG, whereas people at the 50-70 range are at an equally increased risk irrespective of their gender[2]. Additional risk factors for developing MG include familial history, thyroid diseases, auto-

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immune disorders and demyelinating CNS diseases[3].

The pathophysiology of MG involves a defect in the transmission of nerve impulses to muscles due to compromise of the neuromuscular junction. Circulating autoantibodies block, interfere or alter Ach receptors at the postsynaptic NM[1]. Myasthenia gravis is most often associated with dysfunction of the nicotinic AchR, although less commonly the offending cause might be traced back to autoantibodies attacking muscle-specific kinase protein[4].

There appears to be a link between the thymus gland and acquired MG, although it is not entirely clear whether its role in the pathogenesis of the disease is primary or secondary. The thymus gland lies in the upper chest area just beneath the breastbone. It plays a leading role to the development and maturation of immune system during early life, by promoting immunological self-tolerance which is the capacity of the body to recognize self vs. foreign antigens[5]. A great proportion of patients with MG have a background of thymic hyperplasia and a 10-15% also has thymoma[6,7]. The latter have more severe disease and higher levels of AChR autoantibodies.

The cornerstone of MG symptomatic management are anticholinesterase agents, such as neostigmine and pyridostigmine[8,9]. The latter improve neuromuscular transmission and increase muscle strength, by inhibiting the activity of acetylcholinesterase, an enzyme that catalyzes the degradation of Ach. Hence, more Ach is available at the neuromuscular junction to convey the signal. Immunosuppressive drugs such as prednisone, cyclosporine and azathioprine may also be used as part of the treatment[10]. These drugs act by suppressing the production of anti-antibodies against Ach receptors.

Finally, thymectomy, the surgical removal of the thymus gland, reduces symptoms in more than 70 percent of patients without thymoma and may also cure some individuals. The purpose of this paper is to describe the basic elements of a nursing care plan for a patient undergoing thymectomy due to myasthenia gravis.

SYMPTOM RECOGNITION

A nurse needs to be familiar with the typical clinical manifestations of the disease. These include ocular symptoms such as ptosis and diplopia, occurring in more than 50% of patients[11]. Bulbar symptoms such as dysphagia, fatigable chewing and dysarthria may be seen in up to 15% of patients[12]. They may be noted by the need of patients to use their fingers to push the jaw so that their mouth is kept closed, particularly through meals with meat or other concentrated solid foods. Fluids may be required to ease swallowing. Choking, nasal regurgitation and in most severe cases aspiration pneumonia may be seen. Nasal speech occurs due to palatal weakness and, expectedly, may worsen with prolonged speaking. Facial weakness manifests

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itself as a transverse smile, known as myasthenic snarl and an otherwise expressionless face. In less than 5% patients present with proximal limb weakness[3]. In generalized MG the respiratory muscles are affected which results in breathing difficulty while supine. It may result in respiratory failure and then the patient is said to be at a myasthenic crisis.

MANAGEMENT

General considerations

Meals should be taken during the periods of maximal strength, e.g. earlier part of the day or ½ hour after the administration of AchE medication. The patient should be positioned upright with his or her head slightly forward and rest before drinking or eating. Foods should be soft, not sticky and not requiring excessive chewing. The patient shall be offered small bites, be instructed to chew slowly, swallow and take a rest in between the bites. The meals should be small and frequent. Large meals are to be given at the morning and smaller at the evening. Talking during eating should be discouraged to avoid fatigue and risk for aspiration. Sufficient fluid intake should be ensured along with nutritional intake. The nurse needs to be aware of common complications, e.g., urinary incontinence, bowel incontinence, diarrhea (as a manifestation of AchE overdose), psychological breakdown, etc. Stressful activities, either physical or emotional, should be limited since they could aggravate the symptoms.

Pharmaceutical management

Acetylcholinesterase inhibitors, such as neostigmine and pyridostigmine, decelerate the degradation of Ach and ease the patient's symptoms[8,9]. Nurses need to be familiar with the potential adverse effects of these agents, such as abdominal cramps, diarrhea and fasciculations. It is very important to be able to differentiate MG crisis and cholinergic crisis. Doses need to be titrated to the individual patient, aiming at a balance between side-effects due to overdosing and muscle weakness due to underdosing. A nurse must also participate in the education of the patients and their families, on how to administer the correct dose, on time and be familiar with side effects.

Patients with MG may additionally receive immunosuppressive agents as part of their treatment. Since these drugs are not MG-specific and affect the general immune function, naturally they put patients at an increased risk of infections and delay wound healing at the context of a thymectomy. Therefore, nursing personnel must engage appropriate infection control practices including good hand hygiene and sterile techniques for invasive procedures. They also need to convey the same message to the patients and their families and advise them to avoid exposure to people with a known infection.

There is a vast amount of drugs that when used for other conditions, unrelated to MG, may ag-

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gravate muscle weakness and trigger a crisis[13]. Therefore, nurses treating such patients should be aware of these drugs and use them with extreme caution, fully monitoring the patients for potential side effects. Some common drugs that fall into this category include macrolide and aminoglycoside antibiotics (e.g. azithromycin, vancomycin, etc.), quinolone antibiotics (ciprofloxacin, levofloxacin, norfloxacin), beta-blockers such as propranolol, calcium channel blockers, magnesium salts (laxatives, antacids), local anesthetics, analgesics, anxiolytics, sedatives and hypnotics. Surely, this list is not inclusive of all drugs that could potentially interact with the disease. The nurse should review the medication profiles and contact the treating physician prior to the administration of any drug whatsoever.

Surgical management

Patients undergoing thymectomy may present with certain postoperative complications, such as respiratory failure due to myasthenic crisis, infection, etc. For patients with preoperative respiratory compromise, treatment with plasmapheresis or intravenous immunoglobulin may be warranted prior to the surgery or during the immediate postoperative period to lower the probability of a myasthenic crisis[14].

At the postoperative period patients should be monitored closely, particularly their respiratory status, including rate, depth and work of breathing, oxygen saturation and ventilation tolerance. Also pulmonary function testing, specifically forced vital capacity (FVC) and negative inspiratory force (NIF) are recommended by some authors[15,16]. Any bronchial secretions should be thoroughly suctioned to secure a clear airway. Due to the effect of drugs administered by the anesthesiologist for anesthesia, analgesia and sedation, patients are at a considerable risk for becoming symptomatic postoperatively[15]. Nurses need to balance the administration of medications that may exacerbate MG, including opioids. The aim is to achieve a comfort level for the patient that will allow him to breathe as effortlessly as reasonably achievable.

Patients undergoing thymectomy, depending on the approach used, may need one or more chest tubes to monitor and control thoracic drainage. Hourly chest tube drainage of greater than 100 ml is considered excessive[17]. It is the nurse's responsibility to monitor the tube's drainage output, check the patency, look for any evidence of air leak and signs of infections.

There are certain factors that may put the patient undergoing thymectomy at an increased risk for postoperative complications, such as[18] preoperative vital capacity < 1.2 L, bulbar symptoms, a history of preoperative myasthenic crisis, high AchR antibody serum level > 100 nmol/L and an intraoperative blood loss exceeding 1000 mL. Thus, nurses should be at an elevated level of alertness, anticipating a postoperative MG crisis in these patients and acting pro-

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actively. Besides respiratory failure, other MG-crisis symptoms may include tachycardia, tachypnea, hypertension, pale and cool skin and generalized increased weakness. The in-hospital mortality of patients experiencing an MG crisis postoperatively is increased[2] and all efforts should be made to prevent it.

The average daily dose of an AchE inhibitor may be decreased following a thymectomy. Clinical improvement though may take up to several months to reach maximal point[19,20]. Therefore, patients need to be educated appropriately by a multidisciplinary team which includes nurses. Awareness of the potential interactions of drugs unrelated to MG should be conveyed to the patient and family.

CONCLUSIONS

In order to optimally treat patients with myasthenia gravis undergoing thymectomy there is a steep learning curve. It is very important to be able to recognize early the symptoms and signs of common MG complications and particularly myasthenic crisis. Prompt identification combined with aggressive nurse interventions are the keys to ensure the best possible outcomes.

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Author Disclosures:

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