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Review Article

Approach to Emergencies in Neuromuscular Disorders

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Abstract

The entire gamut of neuromuscular diseases presenting in an emergency can be varied with some dramatic presentations requiring immediate stabilization and management. Respiratory involvement is the leading indication for admission of patients with neuromuscular disorders to the Intensive Care Unit. Here, in this article we detailed mentioned how to clinically diagnose neuromuscular emergencies and criteria for immediate intervention in ICU. We also discussed management of neuromuscular diseases requiring emergency intervention in gross as well as with special reference to individual disease. So, clinicians always keep in mind that neuromuscular diseases is one of the important differential diagnosis in patients referred to emergency department or ICU. A cautious history and thorough clinical evaluation often help us to plan for proper management.

Keywords: Diagnosis; ICU; Neuromuscular Diseases

Introduction

The entire gamut of neuromuscular diseases presenting in an emergency can be varied with some dramatic presentations requiring immediate stabilization and management. Respiratory involvement is the most common reason for these patients to land up in emergency room or Intensive Care Unit of a hospital. Another equally important reason is the involvement of bulbar muscles that finally results in aspiration pneumonia. Rarely these patients may have cardiac arrhythmias due to autonomic dysfunction or cardiomyopathy. Depending upon the type of neuromuscular disorder, involvement can occur at the spinal cord level in the anterior horn cell, near the spinal cord in the sensory ganglia, in the sensory or motor nerves or at the neuromuscular junction level. Immediate diagnosis of the conditions and institution of appropriate therapies can improve survival and mitigate the suffering of the patients.

Approach to determine the cause of weakness

As a general rule, the following four things are required to reach to a conclusive diagnosis, the first being as important as the rest of the three. These are (1) Knowledge of possible disorder/Spectrum of conditions presenting with Neuromuscular Emergency, (2) History, (3) Physical Examination and (4) Laboratory investigations.

Spectrum of conditions presenting with neuromuscular emergency

Significant respiratory or bulbar involvement may come acutely in conditions such as Guillaine Barre Syndrome or acute myasthenic crisis where presentation is dramatic with a typical history and few differentials. On the other hand, conditions like anterior horn cell disease, botulism, snakebite, persistent tick bite etc., may not come with a clear history and a detailed targeted history and examination is required to come to the diagnosis.

The main causes of generalized weakness encountered in an intensive care setting can be easily remembered using the mnemonic 'MUSCLES' as given below in Table 1.

M	Medications: Steroids, Neuromuscular Blockers (Pancuronium, Vecuronium), Zidovudine, Amiodarone
U	Undiagnosed neuromuscular disorder: Myasthenia, LEMS, inflammatory myopathies,
	Mitochondrial myopathy, acid maltase deficiency.
S	Spinal cord disease (ischemia, compression, trauma, vasculitis, demyelination)
C	Critical illness myopathy, polyneuropathy
L	Loss of muscle mass (cachectic myopathy, rhabdomyolysis)
E	Electrolyte disorders (hypokalemia, hypophosphatemia, hypermagnesemia)
S	Systemic illness (porphyria, AIDS, vasculitis, paraneoplastic, toxic)

Table 1: Mnemonic for differential diagnosis of generalized weakness in the intensive care.

The flowchart below represents an algorithm that should help clinicians adopt a systematic approach when dealing a patient with severe weakness in the critical care unit.

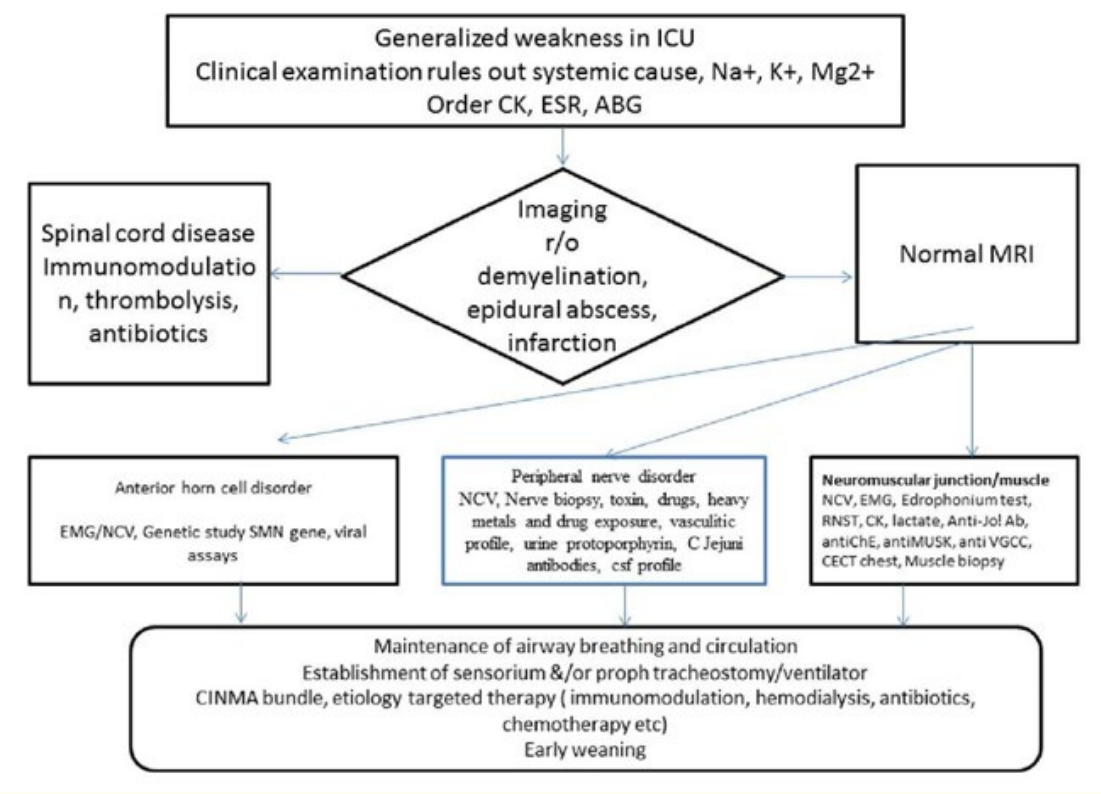

Figure 1

The primary non-neurological causes of generalized weakness comprise of metabolic derangements, poisonings and snake envenomation, diabetic ketoacidosis, congestive heart failure, respiratory conditions like exacerbation of chronic obstructive pulmonary disease/asthma, acute respiratory distress syndrome or pneumonia. Electrolyte imbalance like hypokalemia, hypermagnesemia, and hypocalcemia can also cause generalized fatigue and weakness. Preserved or hyperactive reflexes in the absence of any sensory involvement point out to the involvement of the anterior horn cell or spinal cord lesion whereas absent reflexes with varying levels of sensory involvement point out to a demyelinating neuropathy. Imaging is done to rule out lesions in the spinal cord and/or brainstem and to provide supportive evidence demyelination or focal lesion if suspected.

Clinical history

The most important cause of acute deterioration in a chronic neuromuscular disease is the acute focal or systemic infection particularly the chest infection. Therefore, the history of pre-existing disease such as myasthenia gravis, amyotrophic lateral sclerosis or muscular dystrophy needs to be elucidated along with the history of acute onset deterioration, terminal stage of disease or chest infection. History of preexisting systemic disorders like malignancy, connective tissue disease is also important. Dietary history of canned food points towards botulinum toxicity.

History of tick bite will rule out tick paralysis and the history of recent vaccination will rule out vaccine induced paralysis. History of drug intake is important as it may per say cause muscle or neuromuscular weakness and also precipitate disease like myasthenia. Various drugs causing muscle weakness are to be kept in mind while evaluating any acute onset muscle weakness. Diuretics are known to precipitate hypokalemia. Certain drugs have direct myotoxic effect; these include statins, colchicine, penicillamine, zidovudine, cyclosporine, cocaine, chloroquine, L-tryptophan and corticosteroids. Other drugs such as amiodarone, cytarabine and streptokinase may cause demyelinating neuropathy. Magnesium containing antacids may cause hypermagnesemia in patients with chronic renal insufficiency.

The onset of disease whether acute, sub-acute or chronic helps in narrowing down the differentials. Acute onset flaccid weakness is a presentation of Gullian-Barre Syndrome,

Hypokalemic/hyperkalemic periodic paralysis, botulinum toxicity and tick paralysis. Sub-acute onset weakness is seen in inflammatory myopathies, diphtheria, porphyria, myasthenia gravis, LEMS. Most of the other neuromuscular diseases have a chronic course of illness and they would come to emergency during acute onset deterioration or crisis. Tetanus is usually clinically suspected from the history of a trivial wound followed by hyperpyrexia and trismus. Critical illness neuromyopathy usually manifests together and has varying grades of motor weakness with sensory involvement mainly seen as a difficulty in weaning from mechanical ventilation.

Physical examination

Clinical evaluation should focus on examining the patient's sensorium, cranial nerves, motor power, reflexes, sensory examination and plantar response. Other systems should be evaluated rapidly to exclude close differentials. Thorough examination of hair and axilla is required, especially in pediatric patients for ticks as they present with ascending paralysis. Acute onset flaccid weakness with pure motor or mild sensory symptoms but no involvement of bladder points towards GBS, tick paralysis, myasthenia, botulism, diphtheria, porphyria or hypokalemic periodic palsy. Involvement of bulbar muscles points towards myasthenia gravis or acute exacerbation of polymyositis. Bilateral facial nerve involvement is seen in GBS. Cranial nerve involvement, impaired pupillary response, loss of sphincter tone and impaired tendon reflexes are seen in botulism. Decreased reflexes or areflexia is are also seen in GBS, diphtheria and LEMS. Bulbar weakness/pseudobulbar weakness suggested by dysarthria, dysphagia, cough during eating and nasal regurgitation. Autonomic dysfunction is seen in GBS, porphyria, diphtheria and LEMS. Respiratory system examination must be done and signs of respiratory distress should be assessed in all patients presenting with acute neuromuscular weakness. Signs of respiratory distress includes single breath count less than 15, tachypnea, inability to complete sentences, inability to lie down/leaning forward, central cyanosis, restlessness, drowsiness unexplained, use of accessory muscles like sternocleidomastoid, indrawing of intercostal muscles and nasal flaring. Respiratory weakness occurs in neuromuscular diseases because of involvement or respiratory muscles, phrenic nerve, bulbar weakness, drugs precipitating acute crisis like in myasthenia, chest infection in chronic diseases like muscular

dystrophy or inflammatory myopathy leading to acute emergencies. Cardiac involvement occurs due to cardiomyopathy, LV hypertrophy and fibrosis which contribute to conduction block, ventricular arrhythmias and sudden cardiac death (SCD).

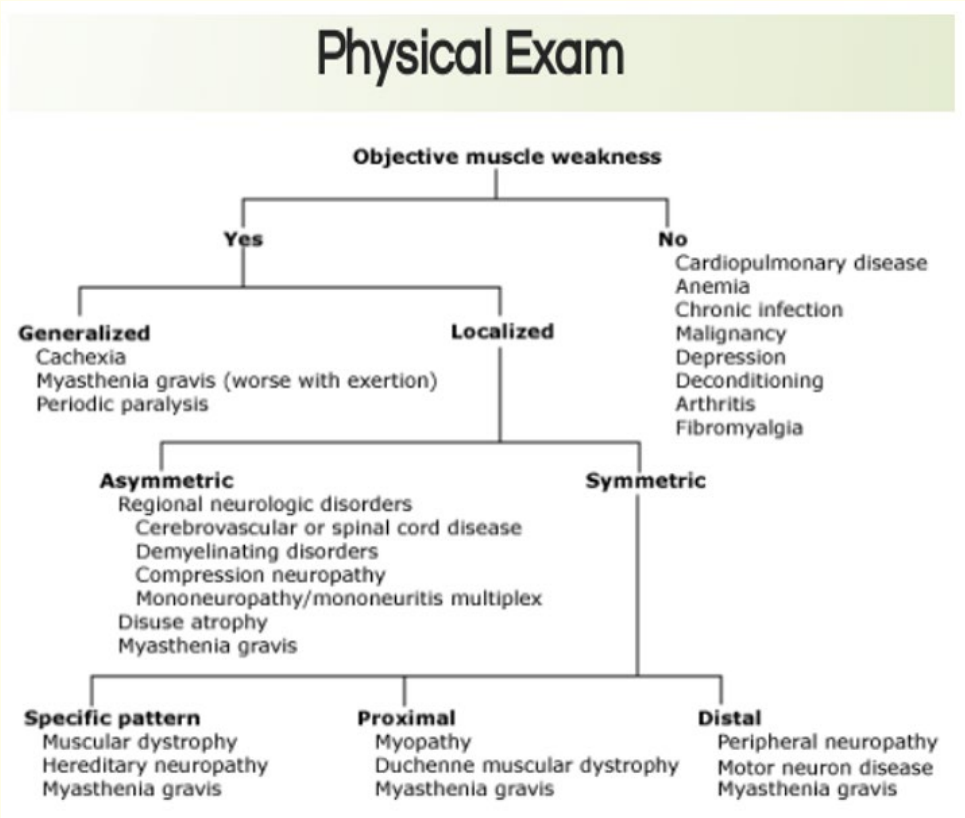

Figure 2

Laboratory investigations

Immediate laboratory tests should be done to rule out metabolic abnormalities. Blood gas analysis should be performed to rule out acidosis, which can cause air hunger and respiratory fatigue. Blood cultures, erythrocyte sedimentation rate (ESR) and creatine phosphokinase (CPK) should also be assessed. Blood chemistry is important specifically serum potassium levels. Hypokalemia/ hyperkalemia and even normokalemia may present with acute onset flaccid weakness. Certain rare conditions like porphyria require urine evaluation for protoporphyrins. Acetylcholinesterase antibodies are positive in approximately 85% of generalized myasthenia gravis and 50% of ocular myasthenia, with high specificity in specificity (100%) in ocular myasthenia gravis with IgG1 and IgG3 subclass proteins were higher among myasthenia patients compared with healthy controls. However MUSK antibodies are observed only in 50% of those with generalized myasthenia gravis and only in few percentage in ocular myasthenia. Other antibodies like Anti striational antibodies are important because they are positive in myasthenic patients with thymoma and are associated with the late-onset MG subgroup and constitute severe form of MG. Similarly Anti-titin antibodies are seen to be positive in older-onset myasthenia gravis patients of more than 60 years age. They are detected in 49–95% of T-MG, with anti-RyR found in 70–80% of cases and anti-Kv1.4 in 40–70% of cases. In GBS various antibodies like anti GQ1b, anti GT1a, anti GD1b and anti GD3 are seen in Miller- Fischer variant whereas high titers of anti-GM1, anti-GD1b, anti GD1a and anti GalNAc-GD1a are seen in AMAN variant with preceding campylobacter jejuni infection.

Specific neurological tests like nerve conduction studies and electromyography are valuable in the lesion to nerve and muscle respectively. However, in patients with neuromuscular junction illness, *RNST* (repetitive nerve stimulation test) can be especially useful. Finally, muscle biopsy is extremely helpful in the diagnosis of inflammatory muscle disease and/or myopathies, which can present acutely with severe weakness.

Conclusion

It is wise to keep in mind that the differential of disorders affecting the nerve or muscle presenting to the emergency is extremely varied. A cautious history and clinical evaluation will often help us to narrow our differentials to an anatomical level. Further investigations can be directed accordingly to pinpoint the diagnosis so that appropriate therapeutic modalities can be applied during the salvage period of the patient. Irrespective of etiology, the management of a patient on mechanical ventilation should be long enough to tide over the crisis yet prompt to recognize and start weaning strategies. The strategic development of the critical illness neuromyopathy should be avoided by strictly following the CINMA bundle. Finally, it is important to remember that every disease in the emergency unit is a challenge that can be surmounted by aggression with sound clinical knowledge and practice.

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