Select Topics in Neurocritical Care

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KEYWORDS

• Neurocritical care • Myasthenia gravis • Guillain-Barre syndrome • Meningitis • Encephalitis • Rhombencephalitis

KEY POINTS

- The most important determinant of the need for intensive care in neuromuscular disease is pending respiratory failure.
- Clinical presentation can differentiate meningitis from encephalitis.
- The most common cause of immune-mediated encephalitis in adults is antibody mediated encephalitis.
- Guillian Barre syndrome is a commonly missed diagnosis in the ED which may progress to significant morbidity.

INTRODUCTION

Neurocritical care aims to improve outcomes in patients with life-threatening neurologic illness. The scope of neurocritical care extends beyond the more commonly encountered and important field of cerebrovascular disease, as described previously.[1,2] This article focuses on neuromuscular, neuroinfectious, and neuroimmunologic conditions that are significant causes of morbidity and mortality in the acutely neurologically ill patient. As understanding continues to increase regarding the physiology of these conditions and the best treatment, rapid identification, triage, and treatment of these patients in the emergency department (ED) is paramount.

NEUROMUSCULAR DISEASE

Unlike those in the central nervous system, lesions in the peripheral nervous system can be difficult to localize and may delay diagnosis, with detrimental effects. The 2 main categories of neuromuscular disease that frequently require intensive care are peripheral demyelinating disease (Guillain-Barre syndrome [GBS]) and neuromuscular junction disease (myasthenia gravis). The clinical course can be highly variable, ranging from strictly outpatient treatment to prolonged ventilator dependence in an intensive care unit (ICU) setting, therefore appropriate early triage is important.

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Peripheral Demyelinating Disease

With an estimated incidence of 1 to 4 per 100,000, GBS and its variants are rare but represent the most common cause of acute paralysis.[3] Guillain-Barre syndrome typically presents with ascending weakness, paresthesias, and areflexia, but at least 4 subtypes have been described with variable presentations. Miller-Fisher syndrome accounts for roughly 5% of GBS cases and presents with ataxia, ophthalmoplegia, and areflexia. Acute motor axonal neuropathy and acute motor sensory axonal neuropathy are differentiated by severe damage to peripheral nerve axons in addition to demyelination, and present as acute paralysis and loss of reflexes with or without sensory loss. Finally, acute panautonomic neuropathy is a rare variant that can be rapidly fatal, and presents with encephalopathy and autonomic instability.[4]

Pathophysiology

Guillain-Barre syndrome is an autoimmune disease with antibodies directed against targets on peripheral nerves. These antibodies usually form 4 to 7 days after an antecedent infection. Most common infections include *Campylobacter jejuni* and cytomegalovirus.[5] Although an association between GBS and influenza has been reported, specifically after the swine flu in the late 1970s, more recent studies have shown no specific correlation.[6] The typical course of GBS progresses through 4 phases: (1) interval between the inciting illness and onset of neuromuscular symptoms, (2) progressive weakness lasting less than a month, (3) plateau, and (4) recovery. Quadriplegia may occur in a rapidly progressive form of GBS, in which onset of respiratory failure can occur within 48 hours.[5]

Clinical presentation and diagnosis

Generalized weakness is a common complaint in the emergency department, and therefore a high index of suspicion is needed to correctly diagnose GBS. A retrospective case series of 20 patients with GBS over 5 years in a large ED found that only 25% were accurately diagnosed during their first visit and on average 2 visits were needed for diagnosis.[4] In patients presenting with ascending weakness or parasthesia after an illness, a neurologic examination is needed to specifically focus on ocular movement abnormalities and absent reflexes. A lumbar puncture should also be performed to assess for albuminocytologic dissociation, in which an elevated protein level is seen (>100 mg/dL) without an elevation in cell count.[3] Electromyography can eventually be helpful to determine the extent and type of damage but it is typically normal in the acute setting and not performed urgently in the ED.

Neuromuscular Junction Disorders

Disorders of the neuromuscular junction refer to interruptions in the transmission of acetylcholine from the nerve terminal to the muscle cell. In developed countries, the most common neuromuscular junction disorder is myasthenia gravis with an incidence of 1 to 3 per 1 million people. As with GBS, the variability of clinical presentation and disease course is significant and a high index of suspicion is needed for diagnosis. A higher incidence of myasthenia gravis is seen in women, with the peak onset occurring in childbearing years. Up to 20% of patients with myasthenia gravis will have a myasthenic crisis, characterized by the need for mechanical ventilation, within 2 years of diagnosis.[7]

Pathophysiology

Myasthenia gravis is an autoimmune disorder with antibodies most commonly directed toward the acetylcholine receptor on the muscle cell membrane. The binding, blocking, and modulating antiacetylcholine receptor antibodies are frequently tested with the binding antibody present in 90% of cases.[7] Medications of many classes