HOSPITAL PHYSICIAN®

NEUROLOGY BOARD REVIEW MANUAL

STATEMENT OF EDITORIAL PURPOSE

The *Hospital Physician Neurology Board Review Manual* is a peer-reviewed study guide for residents and practicing physicians preparing for board examinations in neurology. Each manual reviews a topic essential to the current practice of neurology.

NOTE FROM THE PUBLISHER:
This publication has been developed without involvement of or review by the American Board of Psychiatry and Neurology.

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Multiple Sclerosis

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Multiple Sclerosis

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INTRODUCTION

Multiple sclerosis (MS) is a T-cell mediated autoimmune disease triggered by unknown exogenous agents (eg, viruses, bacteria) in individuals with a specific genetic background. The disease can produce various combinations of symptoms and neurologic signs that reflect the location of a lesion or lesions in different areas of the brain, spinal cord, and optic nerves. Historically, MS was considered to be a disease of the brain white matter, but recent data suggest that primary involvement of gray matter is also quite prevalent in the disease.[1]

MS is the most common neurologic condition affecting young adults in their most productive years. Onset typically is in the third or fourth decade of life but in 5% of patients can be as early as 8 or as late as 70 years of age. As with most autoimmune illnesses, females are affected more often than males. Between 8000 and 10,000 new cases of MS are diagnosed each year in the United States, with an incidence ranging from 1 in 1500 people in the Southwest to 1 in 750 in the Pacific Northwest. The overall prevalence of MS in the United States is considered to be 400,000 to 500,000 patients; worldwide, MS prevalence is estimated to be close to 2.5 million people.[2] Prevalence varies between areas of high, medium, and low risk, with MS being more prevalent in populations located further north from the equator in the Northern Hemisphere and further south from the equator in the Southern Hemisphere. Reasons for this distribution are unknown, although there are data supporting variable geographic distribution of genetically susceptible populations and possible environmental factors.[3]

MS is the most common of several demyelinating disorders. The others include acute disseminated encephalomyelitis (ADEM), neuromyelitis optica (NMO, or Devic's disease), Marburg variant of MS, Schilder's disease, and Balo's concentric sclerosis. ADEM is a rapidly progressive, acute, often postinfectious or postimmunization inflammatory demyelinating condition. ADEM often responds to high-dose steroid treatment and tends to be monophasic, with good resolution of the neurologic deficit.

NMO/Devic's disease is a recurrent or monophasic inflammatory demyelinating illness affecting the optic nerves (bilateral optic neuritis) and spinal cord (myelitis).[4,5] Resolution of the neurologic deficit tends to be incomplete, clinical response to MS treatment (immunomodulatory medications) ineffective, and progression of disability rapid. Immunosuppressive agents and plasma exchange are sometimes effective in treatment of Devic's disease. A recently discovered serum antibody, NMO-IgG, is 70% sensitive but nearly 100% specific for Devic's disease diagnosis.[6]

Marburg variant of MS is an acute, aggressive, and rapidly progressive form of MS, which is poorly responsive to treatment. Schilder's disease is a rare, progressive demyelinating disorder that begins in childhood and is characterized by dementia, aphasia, seizures, personality changes, poor attention, tremors, balance instability, incontinence, and muscle weakness. Schilder's disease responds poorly to treatment and is generally characterized by a progressive, disabling course.[7]

Balo's concentric sclerosis is a rare variant of MS pathologically characterized by alternating rings of demyelination and spared myelin. The clinical course of Balo's concentric sclerosis was considered similar to that of Marburg variant of MS, and most reported cases have involved young adults and resulted in death within weeks to months. Recently, an increasing number of cases have been described as having prolonged survival or spontaneous remission. The most commonly reported clinical manifestations are headache, aphasia, cognitive or behavioral dysfunction, and/or seizures. Cerebrospinal fluid (CSF) studies often reveal a mononuclear inflammatory reaction and occasionally oligoclonal bands.[8]

This review presents 6 cases encompassing a range of clinical presentations of MS, MS subtypes, differential diagnosis, treatment options, and prognosis. We will also consider an algorithm for clinical decision making in cases of patients who present with MS.