or radiating back pain localized to the area of ischemia is frequent. Areflexia due to spinal shock is often present initially; with time, hyperreflexia and spasticity appear. Less common is infarction in the territory of the *posterior spinal arteries*, resulting in loss of posterior column function either on one side or bilaterally.

Causes of spinal cord infarction include aortic atherosclerosis, dissecting aortic aneurysm, vertebral artery occlusion or dissection in the neck, aortic surgery, or profound hypotension from any cause. A "surfer's myelopathy" in the cervical region is probably vascular in origin. Cardiogenic emboli, vasculitis (Chap. 385), and collagen vascular disease (particularly SLE [Chap. 378], Sjögren's syndrome [Chap. 383], and the antiphospholipid antibody syndrome [Chap. 379]) are other etiologies. Occasional cases develop from *embolism of nucleus pulposus* material into spinal vessels, usually from local spine trauma. In a substantial number of cases, no cause can be found, and thromboembolism in arterial feeders is suspected. MRI may fail to demonstrate infarctions of the cord, especially in the first day, but often the imaging becomes abnormal at the affected level.

In cord infarction due to presumed thromboembolism, acute anticoagulation is not indicated, with the possible exception of the unusual transient ischemic attack or incomplete infarction with a stuttering or progressive course. The antiphospholipid antibody syndrome is treated with anticoagulation (Chap. 379). Lumbar drainage of spinal fluid has reportedly been successful in some cases of cord infarction and has been used prophylactically during aortic surgery, but it has not been studied systematically.

Inflammatory and Immune Myelopathies (Myelitis) This broad category includes the demyelinating conditions MS, NMO, and postinfectious myelitis, as well as sarcoidosis and systemic autoimmune disease. In approximately one-quarter of cases of myelitis, no underlying cause can be identified. Some will later manifest additional symptoms of an immune-mediated disease. *Recurrent episodes of myelitis* are usually due to one of the immune-mediated diseases or to infection with herpes simplex virus (HSV) type 2 (below).

MULTIPLE SCLEROSIS MS may present with acute myelitis, particularly in individuals of Asian or African ancestry. In Caucasians, MS attacks rarely cause a transverse myelopathy (i.e., attacks of bilateral sensory disturbances, unilateral or bilateral weakness, and bladder or bowel symptoms), but it is among the most common causes of a partial cord syndrome. MRI findings in MS-associated myelitis typically consist of mild swelling of the cord and diffuse or multifocal "shoddy" areas of abnormal signal on T2-weighted sequences. Contrast enhancement, indicating disruption in the blood-brain barrier associated with inflammation, is present in many acute cases. A brain MRI is most helpful in gauging the likelihood that a case of myelitis represents an initial attack of MS. A normal scan indicates that the risk of evolution to MS is low, ~10–15% over 5 years; in contrast, the finding of multiple periventricular T2-bright lesions indicates a much higher risk, >50% over 5 years and >90% by 14 years. The CSF may be normal, but more often there is a mild mononuclear cell pleocytosis, with normal or mildly elevated CSF protein levels; the presence of oligoclonal bands is variable, but when they are found, a diagnosis of MS is more likely.

There are no adequate trials of therapy for MS-associated transverse myelitis. Intravenous methylprednisolone (500 mg qd for 3 days) followed by oral prednisone (1 mg/kg per day for several weeks, then gradual taper) has been used as initial treatment. A course of plasma exchange may be indicated for severe cases if glucocorticoids are ineffective. **MS is discussed in Chap. 458.**

NEUROMYELITIS OPTICA NMO is an immune-mediated demyelinating disorder consisting of a severe myelopathy that is typically longitudinally extensive, meaning that the lesion spans three or more vertebral segments. NMO is associated with optic neuritis that is often bilateral and may precede or follow myelitis by weeks or months, and also by brainstem and, in some cases, hypothalamic involvement. Recurrent myelitis without optic nerve involvemement can also occur in NMO; affected individuals are usually female and often of Asian ancestry. CSF studies reveal a variable mononuclear pleocytosis of up to several hundred cells per microliter; unlike MS, oligoclonal bands are generally absent. Diagnostic serum autoantibodies against the water channel protein aquaporin-4 are present in 60–70% of patients with NMO. NMO has also been associated with SLE and antiphospholipid antibodies (see below) as well as with other systemic autoimmune diseases; rare cases are paraneoplastic in origin. Treatment is with glucocorticoids and, for refractory cases, plasma exchange (as for MS, above). Preliminary studies suggest that treatment with azathioprine, mycophenylate, or anti-CD20 (anti–B cell) monoclonal antibody may protect against subsequent relapses; treatment for 5 years or longer is generally recommended. **NMO is discussed in Chap. 458.**

SYSTEMIC IMMUNE-MEDIATED DISORDERS Myelitis occurs in a small number of patients with SLE, many cases of which are associated with antibodies to antiphospholipids and/or to aquaporin-4. Patients with aquaporin-4 antibodies are likely to have longitudinally extensive myelitis by MRI, are considered to have an NMO-spectrum disorder, and are at high risk of developing future episodes of myelitis and/or optic neuritis. The CSF in SLE myelitis is usually normal or shows a mild lymphocytic pleocytosis; oligoclonal bands are a variable finding. Although there are no systematic trials of therapy for SLE myelitis, based on limited data, high-dose glucocorticoids followed by cyclophosphamide have been recommended. Acute severe episodes of transverse myelitis that do not initially respond to glucocorticoids are often treated with a course of plasma exchange. Sjögren's syndrome (Chap. 383) can also be associated with NMO spectrum disorder and also with cases of acute transverse or chronic progressive myelopathy. Other immune-mediated myelitides include antiphospholipid antibody syndrome (Chap. 379), mixed connective tissue disease (Chap. 382), Behçet's syndrome (Chap. 387), and vasculitis related to polyarteritis nodosa, perinuclear antineutrophilic cytoplasmic (p-ANCA) antibodies, or primary central nervous system vasculitis (Chap. 385).

Another important consideration in this group is sarcoid myelopathy that may present as a slowly progressive or relapsing disorder. MRI reveals an edematous swelling of the spinal cord that may mimic tumor; there is almost always gadolinium enhancement of active lesions and in some cases nodular enhancement of the adjacent surface of the cord; lesions may be single or multiple, and on axial images, enhancement of the central cord is usually present. The typical CSF profile consists of a mild lymphocytic pleocytosis and mildly elevated protein level; in a minority of cases, reduced glucose and oligoclonal bands are found. The diagnosis is particularly difficult when systemic manifestations of sarcoid are minor or absent (nearly 50% of cases) or when other typical neurologic manifestations of the disease—such as cranial neuropathy, hypothalamic involvement, or meningeal enhancement visualized by MRI—are lacking. A slit-lamp examination of the eye to search for uveitis, chest x-ray and CT to assess pulmonary involvement and mediastinal lymphadenopathy, serum or CSF angiotensin-converting enzyme (ACE; CSF values elevated in only a minority of cases), serum calcium, and a gallium scan may assist in the diagnosis. The usefulness of spinal fluid ACE is uncertain. Initial treatment is with oral glucocorticoids; immunosuppressant drugs, including the tumor necrosis factor α inhibitor infliximab, have been used for resistant cases. **Sarcoidosis is discussed in Chap. 390.**

POSTINFECTIOUS MYELITIS Many cases of myelitis, termed *postinfectious* or *postvaccinal*, follow an infection or vaccination. Numerous organisms have been implicated, including Epstein-Barr virus (EBV), cytomegalovirus (CMV), mycoplasma, influenza, measles, varicella, rubeola, and mumps. As in the related disorder acute disseminated encephalomyelitis (Chap. 458), postinfectious myelitis often begins as the patient appears to be recovering from an acute febrile infection, or in the subsequent days or weeks, but an infectious agent cannot be isolated from the nervous system or CSF. The presumption is that the myelitis represents an autoimmune disorder triggered by infection and is not due to direct infection of the spinal cord. No randomized controlled trials of therapy exist; treatment is usually with glucocorticoids or, in fulminant cases, plasma exchange.

ACUTE INFECTIOUS MYELITIS Many viruses have been associated with an acute myelitis that is infectious in nature rather than postinfectious. Nonetheless, the two processes are often difficult to distinguish. Herpes