Acute Disseminated Encephalomyelitis

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CLINICAL HISTORY

A 49-year-old woman presented to the emergency department with a change in mental status. The patient had recently had the flu, but otherwise had been feeling well. The rest of her history was unremarkable. CT and MR imaging were performed with the imaging findings described below. A lumbar puncture showed an increased number of lymphoid cells, and cultures were negative for bacteria, mycobacterium, or fungi.

FINDINGS

A CT scan of the brain without intravenous contrast (not shown) did not show any abnormalities. Axial T2-FLAIR image (Figure 1) demonstrates multiple small regions of hyperintense signal within the subcortical white matter. Axial T2-FLAIR image (Figure 2) through the basal ganglia also showed regions of hyperintense signal. Sagittal T2-FLAIR image (Figure 3) demonstrated hyperintense signal of the undersurface of the corpus callosum. A few lesions showed mild enhancement following contrast administration (not shown).

DIAGNOSIS

Acute disseminated encephalomyelitis.
DISCUSSION

Acute disseminated encephalomyelitis (ADEM) is a monophasic inflammatory demyelinating disease that typically occurs after vaccination or following a viral infection. The most common agents responsible for the prodromal phase include Epstein-Barr virus, cytomegalovirus, herpes simplex virus, and mycoplasma. However, a particular agent is identified in only a minority of ADEM cases. The disease may also be idiopathic. This condition is caused by an autoimmune response that forms antibodies that attack and destroy myelin.

Neurologic symptoms include decreased level of consciousness that varies from lethargy to coma. Additional symptoms include hemiparesis, paraparesis, cranial nerve palsies, and seizures. Most of the time, symptoms resolve spontaneously. However, permanent sequelae may be seen in up to 25% of cases. A small percentage of the time the disease can be fatal. The degree of recovery is not related to the severity of the illness, as patients who developed quadriplegia, blindness, or coma have been able to recover fully. Relapses after ADEM are uncommon.

Demyelinating lesions associated with ADEM usually form 10 to 14 days after the viral infection or vaccination. Lesions may involve both the brain and spinal cord. Lesions predominately involve the white matter at the grey-white matter junction; however the grey matter may also be involved. Lesion size varies from punctate to large tumefactive masslike lesions. CT is often negative at initial presentation, although low-attenuation lesions may be seen. Contrast-enhanced CT may show ring-enhancing lesions. MRI is the modality of choice in evaluating ADEM. Findings include bilaterally asymmetric T2-FLAIR hyperintense regions and show variable enhancement that may be central or peripheral. Cranial nerve enhancement may be seen. Leptomeningeal enhancement appears in rare instances. Restricted diffusion on diffusion-weighted imaging (DWI) is variable. No new lesions should be seen on MRI after six months, though there may be incomplete resolution of hyperintense lesions. Imaging manifestations overlap with those of multiple sclerosis. Diagnosis, therefore, is based on history and cerebrospinal fluid analysis. Analysis of the CSF usually shows a lymphocytic predominance and an increase in myelin basic protein.

A severe form of ADEM is acute hemorrhagic leukoencephalitis (Hurst's disease) characterized by...
diffuse multifocal perivascular demyelination and hemorrhage confined to the cerebral white matter with sparing of the subcortical U-fibers. Histologically, there is necrotizing angitis with petechial hemorrhages. Death usually occurs within a week from severe cerebral edema.

The overall goal of treatment is to stop the central nervous system inflammatory response as quickly as possible. High-dose corticosteroids are the treatment of choice in inflammatory demyelinating CNS diseases. Other options include the use of anti-inflammatory and immunosuppressive treatments such as IV immunoglobulin (IVIG) and plasmapheresis.

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BIBLIOGRAPHY


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