ICU
MANAGEMENT & PRACTICE

Mayo Clinic Clarifies Diagnosis and Treatment for Rare, Serious Blood Vessel Disease of Brain and Spinal Cord

This disease is often suspected based on a combination of symptoms, along with the initial results of brain imaging with CT or MRI. The Mayo study clarifies the usefulness of the evaluation techniques and suggests that a combined diagnostic approach is needed. Primary central nervous system vasculitis is difficult to diagnose because it is not a single disease, but a variable syndrome that appears to consist of several distinct subsets of different diseases that can mimic other less-serious conditions, thus eluding treatment. Frequent symptoms are headache, altered cognition, or stroke, and visual symptoms. Recognizing the symptoms and understanding the role of evaluation techniques, may lead to earlier identification and treatment of patients.

This Mayo Clinic analysis of 101 patients seen with primary central nervous system vasculitis over 21 years represents one of the largest reported groups of patients with this disorder. The Mayo Clinic research report on improved understanding of PCNSV appears in the current edition of Annals of Neurology (http://www3.interscience.wiley.com/cgi-bin/jissue/78504407).

Significance of the Mayo Clinic Research

Primary central nervous system vasculitis has two traits that make it difficult, yet important, to study:

-- It is rare and poorly understood. Few medical centers see enough patients with confirmed diagnoses to collect enough data to analyze.

-- It carries a high potential for serious complications, even death. A blood vessel that malfunctions may disrupt blood supply and oxygen to the brain or spinal cord, leading to a stroke.

While angiography has been the preferred diagnostic technique because it is less invasive and carries fewer risks than brain biopsy, no studies had determined its accuracy or established angiographic criteria for the diagnosis. "Progress in understanding PCNSV has been slow because its occurrence is infrequent and its identification is difficult," explains Robert D. Brown Jr., M.D., chair of the Mayo Clinic Department of Neurology, and the lead investigator. "Our study begins to answer some essential questions that can clarify diagnosis and hopefully lead to earlier, and successful, management."

Symptoms and Treatment

Patients with PCNVS in the Mayo Clinic study had the following symptoms: headache was the most common, followed by changes in ability to think or remember (cognitive impairment); weakness on one side of the body; and difficulty speaking (aphasia). Of the 101 patients, 97 were prescribed initial treatment consisting of oral or intravenous doses of the steroid drug prednisone; some received oral or intravenous doses of the chemotherapy drug cyclophosphamide. Most showed favorable responses. "This encouraging finding emphasizes the need for early diagnosis and initiation of therapy that may help avoid serious and irreversible complications," says Dr. Brown.

Key Findings

Some the study's main findings:

-- Angiography imaging of blood vessels appears to complement brain biopsy in determining diagnosis.

-- PCNSV is not a single disease, but a variable syndrome that appears to consist of several distinct subsets of different diseases.

-- Though treatment is available and most patients responded to therapy, relapse was common, occurring in 25 percent of patients studied.

-- Death and disability rates among patients were highest in those who had a defined brain dysfunction, such as cognitive impairment, stroke and disease of the large blood vessels on angiography.

-- The annual incidence rate of primary central nervous system vasculitis was 2.4 cases per 1 million people.

Collaboration and Support

Other members of the Mayo Clinic research team include Carlo Salvarani, M.D.; Kenneth Calamia, M.D.; Teresa Christianson; Stephen Weigand; Dylan Miller, M.D.; Caterina Giannini, M.D.; James Meschia, M.D.; John Huston III, M.D.; and Gene Hunder, M.D. Their work was supported with grants from Mayo Foundation for Medical Education and Research.

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