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Normal Pressure Hydrocephalus: New Findings and Old Questions

Normal pressure hydrocephalus (NPH), a form of communicating hydrocephalus, is characterized by normal mean CSF pressure. MR imaging is essential to establish the presence of ventricular dilatation and to exclude other conditions that also may be responsible for the clinical triad of gait disorder, mental slowing, and urinary incontinence. Clinical features that support the diagnosis of NPH include a recent or remote history of subarachnoid bleeding or meningitis and mental changes of a ''subcortical dementia,'' typified by slowness of responses (bradyphrenia), and dullness and apathy, different from early Alzheimer's disease. The gait is typically slow and unsteady with magnetic responses (as if the feet are stuck to the floor) and gait apraxia. The ''CSF tap test,'' with which some CSF is removed by means of lumbar puncture, may be of value despite false-negative and false-positive tests, but only if the test is well standardized (such as comparing the time required to walk a standard distance before and after large volume CSF drainage). Lumbar puncture is essential to establish that the CSF pressure is normal and to exclude a chronic meningitis that can be responsible for large ventricles and the clinical features of NPH. An elevated protein ($\leq$3000 mg/dL) may be the sole indicator of a spinal neoplasm responsible for the syndrome of NPH.

The major diagnostic challenge is to differentiate NPH from cerebral atrophy and deep white matter ischemic, both far more common causes of the clinical triad than NPH. Although NPH and cerebral ischemic changes often concur, the clinician's major question for the neuroradiologist is whether the ventricular enlargement observed is consonant with the degree of enlargement of the cortical sulci (ie, is it hydrocephalus ex vacuo due to cerebral atrophy or is it communicating hydrocephalus?). Unfortunately, many radiologists fail to address this question in their clinical reports. Other MR imaging features of NPH that have been proposed include the ''aqueductal flow void sign'' and increased CSF stroke volume (1), but these remain controversial (2). Isotope cisternography has also been proposed for the evaluation of NPH. Despite the occurrence of ventricular reflux of isotope in NPH, its presence does not reliably predict which patients will have favorable outcomes after shunt surgery; similarly, its absence does not preclude a favorable response to shunt surgery.

MR imaging in cases of hydrocephalus also reveals thinning of the periventricular white matter associated with hyperintensities of the subependymal and deep white matter. Such findings, suggesting small vessel ischemia, are commonly associated with hypertension, diabetes, and aging, as well as with NPH. There has been much speculation regarding their pathogenesis. Unfortunately, there has been insufficient correlation of the imaging data and the neuropathologic findings. Although the degree of subcortical ischemia is inversely correlated with outcome from shunt surgery, this should not preclude the diagnosis of NPH.

New insight into the significance of the white matter changes in hydrocephalus is provided by Tullberg et al (page 1665) who describe two different changes in the white matter in NPH, ''smooth periventricular hyperintensities'' and ''irregular deep white matter hyperintensities.'' The authors observed that only the irregular deep white matter hyperintensities were improved after successful shunt surgery. They also conclude that the decision to perform shunt surgery should not be negatively influenced by the presence of diffuse white matter hyperintensities or lacunar infarctions, which concurs with findings of previous investigators (3). Moreover, they did not find any MR imaging features that were prospectively associated with the diagnosis of NPH. Thus, they support the opinion that the decision to perform shunt surgery should be based largely on clinical findings and MR imaging studies that indicate that brain atrophy is not responsible for the ventriculomegaly.

Unfortunately, many old questions regarding the cause and diagnosis of NPH remain unanswered (4). What are the rates of CSF formation and absorption in healthy volunteers and in patients with NPH? Is the transmural pressure gradient from the ventricles to the subarachnoid space responsible for the enlargement of the lateral ventricles? How does NPH bring about the clinical triad? How much CSF is formed within the cerebral hemispheres compared with the secretion of CSF by the choroid plexus in healthy volunteers versus in that of patients with hydrocephalus? What mechanism underlies the restoration of white matter thickness after successful shunt surgery? How reliable is CSF drainage in predicting the response to shunt surgery? NPH is an uncommon problem that is subject to over-diagnosis as well as under-diagnosis. We anticipate that diagnosis and patient care will be-

come more coherent when the pathophysiology of NPH is better understood.

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