Increased Intracranial Volume in Normal Pressure Hydrocephalus: Evidence for Benign External Hydrocephalus as the Etiology

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Introduction

Normal pressure hydrocephalus (NPH) was initially described as an "idiopathic" disease, ie, the cause was not known. It is diagnosed on the basis of a clinical triad of gait disturbance, dementia, and incontinence in elderly patients presenting with the radiographic picture of communicating hydrocephalus. Phase contrast CSF velocity imaging has been shown to be useful in selecting which patients will respond favorably to ventricular shunting for NPH. Specifically, the volume of CSF pulsating back and forth through the aqueduct over the cardiac cycle ("aqueductal CSF stroke volume" [SV]) is increased.

A few years ago, it was suggested that NPH actually begins in infancy as "benign external hydrocephalus", a condition in which the CSF resorptive capacity is decreased due to delayed maturation of the arachnoidal granulations. Since these children are less than a year old, their sutures can still expand, thus CSF accumulates in the subarachnoid space over the convexities as well as within the enlarged ventricles, causing the children to present with increasing head circumference. If NPH patients truly did have benign external hydrocephalus as infants, they should have greater intracranial volumes than age and sex-matched controls thereafter. Evaluation of that hypothesis was the purpose of this study

Materials and Methods

All patients with CSF velocity imaging studies for clinically suspected NPH over the prior 2 years were retrospectively reviewed. Only those with SVs 50% higher than normal were included in this study as NPH patients. Age and sex matched controls were taken from the same time period. The T2 weighted images of NPH patients and controls were loaded on to a Vital Images workstation and the intracranial volumes were measured. 18 male patients were compared to 26 male controls. 25 female patients were compared to 56 female controls. The mean SV for the NPH males was 149 uL; for NPH females it was 127 uL (compared to a normal literature value of 42 uL).

Results

The mean intracranial volume for NPH males (1690 ml) was significantly larger than the intracranial volume (1584 ml) for the controls ($p < .04$). The mean intracranial volume for NPH females (1495 ml) was significantly larger than the intracranial volume (1407 ml) for controls ($p < .004$).

Discussion

This study shows that NPH patients have intracranial volumes 90-100 ml or 5-6% larger than age and sex-matched controls. The larger volume confirms that the initial insult occurred before the sutures fuse at 1 year of age, as would be the case with benign external hydrocephalus. This would imply that the CSF resorption has always been decreased, not just when it is discovered in old age.

The patients somehow remain asymptomatic until their later years when deep white matter ischemia leads to a second insult, eg, decreased parenchymal CSF resorption or softening of the brain. The "second hit" leads to progressive ventricular enlargement and the symptoms of NPH.

Radiologists probably see the CT and MRI studies of "pre-NPH" patients routinely and dismiss the mild (5-6%) ventricular enlargement as "ventricles at the upper limits of normal". These patients should be observed for early signs of a gait disturbance in their elderly years as NPH is a treatable cause of dementia when ventricular shunting is performed early in the course of the disease.