Cerebral bleed after shunt for normal pressure hydrocephalus with cerebral amyloid angiopathy

Coincidence or consequence?

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The triad of gait impairment, cognitive decline, and urinary incontinence in the setting of normal CSF pressure has long been described, yet our ability to predict who will experience a prolonged, meaningful response to ventriculoperitoneal (VP) shunting remains limited.¹⁻⁴ Unfortunately, the procedure is associated with major risk, and improvements following shunting are not guaranteed and are often short-lived. Assessing the harm to benefit ratio can be problematic.^{2,4} It is imperative not only to identify those likely to respond to shunting but also to identify those with a higher risk of adverse events or those unlikely to respond. Here we report a case of right frontal lobe hemorrhage following VP shunt placement in a patient with preoperative MRI evidence of cerebral amyloid angiopathy (CAA).

Case

A 76-year-old right-handed retired laborer with a history of type 2 diabetes, hypertension, chronic urinary hesitancy without progression, and chronic back pain was evaluated for progressive gait decline and cognitive decline over the previous 2 years.

The Mini-Mental State Examination score was 21/30, with impairment in orientation, working memory, and recall; strength was normal, with decreased vibratory sensation below the knees; and deep tendon reflexes were brisk throughout with extensor plantar responses bilaterally. He had difficulty standing from a seated position without assistance and a wide-based, unsteady gait with decreased stride length without a magnetic gait quality. Vitamin B₁₂ and folic acid were unremarkable. EMG/nerve conduction studies revealed evidence of bilateral lumbar radiculopathy. MRI of the cervical spine revealed mild central canal stenosis

Practical Implications

The use of susceptibility-weighted MRI sequences for detection of microhemorrhages should be considered in the diagnostic evaluation of possible normal pressure hydrocephalus.

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Figure Axial FLAIR MRI prior to the insertion of the ventriculoperitoneal shunt (A), T2* gradient echo MRI demonstrating multiple small hypodensities consistent with microhemorrhages (B), and axial CT scan (C)

(A) Serial caudal-rostral axial fluid-attenuated inversion recovery (FLAIR) and coronal (T2) images demonstrating a disproportionate increase in the temporal horns and posterior lateral ventricles, mild periventricular white matter hyperintensities, and a “tight high” sign (circle) of decreased intrahemispheric sulcus near the vertex. (B) Serial caudal-rostral axial T2* gradient echo MRI demonstrating multiple small hypodensities starting in the left cerebellar hemisphere and continuing up through the occipital, temporal, and inferior right frontal cortices with a right hemisphere predominance. (C) Axial CT scan demonstrating right frontal lobe hemorrhage post ventriculoperitoneal shunt.

at C4–5. MRI of the brain (figure, A) demonstrated ventriculomegaly on fluid-attenuated inversion recovery sequences with posterior predominance, modest global atrophy, a decrease in the intrahemispheric sulcus at the vertex (“tight high” sign⁵), and scattered subcortical white matter changes. The MRI was also consistent with imaging criteria for probable CAA,⁶ with multiple microhemorrhages in the occipital, temporal, and frontal lobes (figure, B). The relevance of the microhemorrhages was not mentioned in the clinical record.

A lumbar drain did not improve gait in a standardized 3-day assessment, but the patient’s family reported gait improvement following discharge. Two months later, a VP shunt was placed. Twenty-four hours following the procedure, a CT scan demonstrated a right frontal lobe hemorrhage (figure, C). Three months later, when referred to our service, he had considerable apathy and abulia; seizures began 9 months later. His gait remained unchanged.

DISCUSSION

Normal pressure hydrocephalus (NPH) is a rare disorder. Community-based studies indicate an incidence of 1.19/100,000/year.² Although some patients have a significant and

meaningful improvement after VP shunt insertion,^{1,3} hospital-based estimates of treatment-responsive NPH can be as low as 0.22/100,000/year.⁴ Improvement following shunt is more likely with short symptom duration, lesser degrees of cognitive impairment (with a pattern of subcortical-frontal impairment more typical), a syndrome dominated by gait impairment without other features of Parkinson disease, and a clear improvement in gait following high-volume CSF tap or external lumbar drain.^{3,7,8} Improvement after VP shunt insertion is less likely in the absence of the cardinal features of NPH: peripheral causes of urinary symptoms should not be used to complete the triad.

Identifying a reliable marker of NPH is difficult because of the varied pathologies that can contribute to the symptoms, and the presence of non-NPH pathology predicts poorer response after VP shunt insertion.^{2,7,8} In a recent report evaluating right frontal lobe biopsies during shunt insertion for NPH, CAA was identified in 6 of 18 biopsies. The patients with Alzheimer pathology had less favorable responses to shunting,⁸ but cerebral hemorrhage was not reported in those patients with CAA. Improvements in MRI technology have enabled reliable detection of cerebral microhemorrhages, an essential feature of CAA. Since CAA is a risk factor for spontaneous lobar hemorrhage, it is reasonable to assume that it will increase the likelihood of hemorrhage after insertion of a VP shunt.

Complications in shunt surgery occur in between 25% and 30% of patients,²⁻⁴ and death or severe residual morbidity occurs in 7%–10%.^{2,4} Yet some patients clearly do obtain sustained benefit from shunt insertion,³ so identifying appropriate candidates remains crucial. While relatively rare, lobar hemorrhage is a particularly debilitating complication of shunt surgery, so it is imperative to identify those at greatest risk. Given the increased risk of spontaneous lobar hemorrhage in CAA, this case raises the possibility that it may also increase the risk of hemorrhage during VP shunting. Clinicians and researchers should systematically review susceptibility-weighted MRI sequences for microhemorrhages prior to shunt insertion and consider the findings in risk stratification for the procedure.

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