Reversal of dementia in normotensive hydrocephalus after removal of a cauda equina tumor

Case report

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An elderly man presented with signs of normotensive hydrocephalus. Elevated protein content in the spinal fluid led to the diagnosis of an “asymptomatic” cauda equina neurilemoma and its removal. Within 6 weeks the patient’s mental status had dramatically improved. Chronic transudation of plasma proteins including fibrinogen into the subarachnoid space had probably impeded spinal fluid reabsorption. It is suggested that the leakage of fibrinogen into the cerebrospinal fluid may be the cause of hydrocephalus in other clinical settings in which there is an elevation of the spinal fluid protein.

Key Words • spinal tumor • fibrinogen • hydrocephalus • dementia • CSF protein

MESSERT and Wannamaker reviewed the syndrome of occult hydrocephalus and confirmed its clinical manifestations of dementia, apraxia, lack of emotional spontaneity, decreased voice volume, and incontinence. Pneumoencephalography is a diagnostic aid; the results are consistent with normotensive hydrocephalus if the lateral ventricles are enlarged, air fails to pass over the convexities, and the callosal angle is less than 120° on the anteroposterior view. In positive cases isotope cisternography demonstrates ventricular influx and occasional flow into the Sylvian cisterns, but no passage of isotope over the convexities. Delayed passage of isotope over the convexities is considered a “mixed” pattern.

If these criteria are met and there is clinical and electroencephalographic (EEG) evidence of progressive decompensation, then it is likely that the symptoms can be reversed. We report a case that fulfilled the above requirements, but had an unusual pathogenesis.

Case Report

This 75-year-old man presented with complaints of confusion and unsteadiness that had begun insidiously 6 months previously. He had a history of rheumatic heart disease and chronic bronchitis.

Examination. The patient was disoriented to time, place, and situation. The cranial nerves were normal. There was no papilledema. He could move all his limbs, and the...
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FIG. 1. Correlation between mental status, CSF protein levels, and procedures recorded at intervals during the patient's hospitalization.

tendon reflexes were normal; however, muscle tone was increased, and Babinski, snout, and grasp reflexes were present bilaterally. His gait was apraxic. Sensory perception and synergy were normal.

Laboratory investigations on admission demonstrated a peripheral leukocytosis, normal hematocrit, electrolytes, and arterial blood gases, and tests for syphilis were negative.

A skull series and brain scan were normal; the EEG was diffusely slow. Cervical spine films demonstrated mild spondylosis. Lumbar puncture on admission showed a pressure of 40 mm H2O, with xanthochromic fluid containing 510 mg/dl protein. There were no cells, and all microbiological studies were negative.

During his hospital stay, the patient’s mental status fluctuated; the severity of his dementia was directly proportional to the cerebrospinal fluid (CSF) protein level, and indirectly proportional to the frequency of lumbar puncture (Fig. 1). The authors suspected that a spinal tumor was responsible for the elevated CSF protein.

A myelogram on the 24th hospital day demonstrated a 3-cm subarachnoid mass over L-5. Pneumoencephalography on the 31st day showed enlarged ventricles and absence of air over the convexities. On the same day CSF fibrinogen was 15 mg/dl (normal 0). A cystometrogram 6 days later confirmed that the patient’s bladder was spastic.

Operation. The next day the patient underwent a lumbar laminectomy and a cauda equina neurilemoma was removed. Histological findings were characteristic: areas of vascular proliferation, thrombosis, and recanalization were seen. Electron microscopy revealed a normal basement membrane. Immunofluorescent microscopic findings were non-contributory.

Postoperative Course. During the convalescent period, two radioisotope cisternograms demonstrated a “mixed” pattern, with ventricular influx and very slow progression over the convexities.9,18 One month postoperatively the patient was fully oriented, demonstrated basic insight, and was walking well. The CSF protein and fibrinogen levels were normal. He continues to do well.
Discussion
The patient presented with a syndrome identical to that of occult hydrocephalus. It was noted that the severity of his confusion was directly proportional to the CSF protein level (Fig. 1). The elevated CSF protein was not explained by the single diagnosis of normotensive hydrocephalus, and prompted the search for a spinal cord tumor.

Many authors have discussed the origin of the elevated CSF protein associated with spinal cord tumors. Suggested mechanisms include change in permeability of extradural blood vessels as a result of direct pressure, breakdown of tumor material, hemorrhage from the tumor, transudation through tumor vessels, and active secretion of proteins by the tumor.

Based on the results of fibrinogen assays, electrophoresis, immunofluorescent microscopy, electron microscopy, and tissue culture, we believe that the CSF protein was a plasma transudate which passed through the fragile walls of blood vessels within the neurilemoma. The history and acellular CSF analysis ruled out a hemorrhage in the recent past.

What is the mechanism by which elevated CSF protein causes increased intracranial pressure? A minor component is osmotic pressure, but the major factor is the change in the reabsorptive surface; the arachnoid granulations become clogged with proteinaceous material, and the valves fail to open.

Elevated CSF protein levels and intracranial pressure have been observed not to be directly proportional in the same patient. We therefore analyzed the CSF for a substance which might be present in approximate proportion to the total CSF protein. This analysis was started late in the management of the patient's case, but was fruitful. There was a correlation between the mental status and CSF fibrinogen levels, which was highly suggestive.

Compensated hydrocephalus with a normal intracranial pressure is an actively destructive process due to the force exerted upon the expanded walls of the ventricular system. We think this force was related to the patient's mental status, to the CSF protein levels, and to the CSF fibrinogen levels.

Since normally no fibrinogen is present, its presence in this case suggests that chronic plasma leakage produced a fibrin coating of the subarachnoid pathways which resulted in hydrocephalus. We caution then that the syndrome of occult hydrocephalus can be produced by reversible spinal cord pathology and probably by other causes of elevated CSF protein. It is advisable to perform fibrinogen assays, since fibrinogen may be the factor common to many forms of communicating hydrocephalus even in the absence of gross hemorrhage.

In a review of the literature we found 25 cases of spinal tumors associated with hydrocephalus, dementia, elevated CSF protein, and papilledema. We believe that in those cases the CSF dynamics and the specific protein contents were not fully evaluated. To our knowledge, no other cases presenting with dementia, apraxic gait, and incontinence have been found to have increased CSF protein and fibrinogen content, normal CSF pressure, and an "asymptomatic" spinal tumor.

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