Tentorial block of cerebrospinal fluid associated with a lumbar neurofibroma

Case report

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A case is reported in which raised intracranial pressure with papilledema due to hydrocephalus resulting from a subarachnoid block at the tentorial hiatus was associated with a lumbar intradural neurofibroma. The possible significance of elevated protein content in cerebrospinal fluid samples taken above and below the tumor is discussed.

Key Words: spinal cord tumor 
- increased intracranial pressure 
- hydrocephalus 
- papilledema 
- tentorial hiatus 
- cerebrospinal fluid protein

The relatively rare occurrence of papilledema with a spinal cord tumor is well documented, although the pathogenesis of the association remains in doubt. The following case is reported because of an unusual finding that suggests a possible mechanism for the production of the papilledema.

Case Report

A 66-year-old man was admitted to the Wessex Neurological Centre on November 16, 1970. He was unable to give an adequate history because of recent intellect impairment, but according to his wife he had been apathetic and depressed with poor memory for 4 months.

Examination. The abnormal physical findings were: intellect impairment, bilateral papilledema, paraparesis more marked on the right, and a left extensor plantar response. A provisional clinical diagnosis was made of a midline intracranial space-occupying lesion, either supra- or infratentorial.

Skull radiographs showed demineralization of the dorsum sellae suggesting raised intracranial pressure; the calcified pineal gland was in the midline. The electroencephalogram showed a left frontotemporal slow-wave focus, and a brain scan (technetium 99m pertechnetate) indicated an ill-defined area of increased uptake in the left frontotemporal region on the lateral view which could not be identified on the anterior view. A left carotid angiogram revealed marked enlargement of the lateral ventricles but no other abnormality. Air ventriculography confirmed enlargement of the supratentorial ventricles; the aqueduct could not be visualized but the fourth ventricle lay in its normal position and air passed into the
cisterna magna. No air passed over the surface of the brain. A lumbar pneumoencephalogram showed that gas could be manipulated into the basal cisterns, but none passed through the tentorial hiatus despite repeated attempts to fill the supratentorial subarachnoid space (Fig. 1). The cerebrospinal fluid (CSF) obtained from the ventricles was clear and colorless with no abnormal cells; the protein level was 50 mg/100 ml. A diagnosis of communicating hydrocephalus due to a tentorial block of unknown etiology was made.

On November 24, 1970, a ventriculoatrial shunt was inserted employing a Spitz-Holter valve opening at medium pressure. Within 7 days the patient became normally oriented and his intellectual state improved. On the 9th postoperative day psychometric tests revealed a verbal scale I.Q. of 103 and a performance scale I.Q. of 85; in the Williams test of delayed recall the patient scored 34 and in the name-learning test 39/60.

At this time the patient complained of severe low-back pain and a difficulty in passing urine, which progressed to retention, requiring catheterization. Physical examination now showed, in addition to the paraparesis, bilateral sensory loss over dermatomes L1-2. Radiographs of the lumbosacral spine were normal. At lumbar myelography on December 7, 1970, fluoroscopy of the contrast column with the patient tilted head-down revealed a poor flow and a complete obstruction at the level of the third lumbar vertebra (Fig. 2). The protein content of the cerebrospinal fluid (CSF) was 2800 mg/100 ml, this specimen having been obtained from below the level of the lesion.

**Operation.** On December 9 a lumbar laminectomy of L1-4 inclusive was performed. A solid intradural tumor measuring 4 x 2 cm with no clear point of origin was found at the level of the body of L-3 and was totally removed; histological examination showed it to be a benign neurofibroma. Cerebrospinal fluid taken from the subarachnoid space immediately above the level of the tumor showed a protein content of 500 mg/100 ml.

**Postoperative Course.** The patient's intellectual improvement continued and on March 31, 1971, nearly 4 months after the laminectomy, micturition was normal, there was only slight weakness of flexion at the left hip, and slight sensory change over the L1-2 dermatome on the left side. Psychometry showed his verbal I.Q. to be 128 and a performance I.Q. of 123; in the Williams test of delayed recall the patient scored 9 and in the name-learning test 59/60. When last seen in November, 1972, the patient was perfectly well.

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Fig. 1. Lumbar pneumoencephalogram following a ventriculogram, anteroposterior view (left), lateral view (right). All parts of the ventricular system occupy normal positions but all four ventricles are enlarged. The extraventricular gas is confined to cisterns in the infratentorial compartment, none passing beyond the tentorial hiatus to outline the convolutions over the cerebral hemisphere.
Increased intracranial pressure with lumbar neurofibroma

**Discussion**

The mechanism that occasionally produces increased intracranial pressure and papilledema in association with spinal cord tumors remains a source of considerable speculation. Factors considered by various authors include the site and type of tumor, the increased protein content of the cerebrospinal fluid, cerebral edema, and the possibility of congenital semipermeability of the meninges. While tumors of the upper cervical canal and craniovertebral junction may obstruct the flow of CSF in the region of the foramen magnum and thereby cause raised intracranial pressure and papilledema, this mechanism cannot explain the occurrence of these features in tumors situated at lower levels in the spinal canal. The type of tumor is also unlikely to be the main factor since no consistent cell type has been present in the cases described. Although spinal ependymoma are traditionally associated with papilledema, no more than 60% of these cases are ependymomas, the other cases including spinal neurofibroma, meningioma, and glioma.

The hypothesis that has received most attention is that the increased protein content of the CSF is itself responsible for the raised intracranial pressure. In our case the protein content was high not only in the CSF below the tumor (2800 mg/100 ml) but also in that immediately above the tumor (500 mg/100 ml); this was also reflected in the high level found within the ventricular system (50 mg/100 ml). This increased protein content is thought to be due either to a meningeal reaction to the presence of the tumor or to specific secretions from the tumor itself. Schaltenbrand suggested that the raised protein content increases the viscosity of the CSF, thereby decreasing the rate of its absorption. Although elevated levels of ventricular CSF protein have been noted in previous reports, there is none in which the protein content was measured immediately above the level of the spinal lesion. Indeed, in some cases a normal protein content has been recorded in the CSF taken from below the spinal obstruction.

In addition to the production of increased protein, the secretion of an abnormal protein of sufficient molecular size and in sufficient quantity to obstruct the pores of the arachnoid villi is an attractive theory which has received some support. However, Welch and Friedman demonstrated that the pores of the normal arachnoid villi will permit the passage of particles up to 7 to 8 μ in diameter, considerably larger than the normal fibrinogen molecule.

Various authors have postulated that the increased protein content provokes an arachnoiditis which leads to internal hydrocephalus, raised intracranial pressure, and papilledema. Arseni and Maretis described two patients with spinal cord tumors in whom there was a dilated ventricular system and an apparent obstruction of the outlets of the fourth ventricle. Both patients underwent posterior fossa exploration and were said to have shown arachnoiditis; in

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Fig. 2. Positive contrast oil myelogram showing complete obstruction to flow at the level of the body of L-3. Arrow indicates site of lumbar injection of contrast medium.
one case histological confirmation was obtained. However, in neither case was the extent of involvement in the basal cisterns demonstrated either by lumbar pneumoencephalography or radioisotope cisternography. In our case, combined ventriculography and lumbar pneumoencephalography revealed the precise site of CSF obstruction, namely, the tentorial hiatus, and it is possible that the high protein content above the spinal lesion was directly responsible for the obstructive process in the subarachnoid space. Previous evidence suggests that this process is reversible because excision of the spinal tumor results not only in a fall in protein levels to normal, but also a lessening of the papilledema.

Although the cause of the papilledema associated with spinal cord tumor is still far from explained, investigations should include an examination of the ventricular system and intracranial subarachnoid spaces. Further knowledge of the patterns of CSF proteins at different sites in such cases may help to elucidate the problem in the future.

References


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