Cauda Equina Neurinoma Associated with Normal Pressure Hydrocephalus

—Case Report—

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Abstract

A 45-year-old male was hospitalized because of gait disturbance, urinary incontinence, and memory disturbance. A computed tomographic (CT) scan revealed dilatation of the cerebral ventricles, and a metrizamide CT scan showed reflux into all ventricles. Cerebrospinal fluid obtained at the time of metrizamide CT was highly xanthochromic. Myelograms demonstrated a tumor between the inferior portion of the fifth lumbar vertebra and that of the second sacral vertebra. After removal of a spinal tumor involving nerve fibers at the cauda equina, his symptoms disappeared and the spinal fluid protein normalized. Three months after surgery, a CT scan revealed reduction of cerebral ventricular size. There are 10 reported cases of spinal tumors resulting in normal pressure hydrocephalus. All these and present cases involved protein in the spinal fluid, which might have been the cause of the hydrocephalus.

Key words: spinal tumor, normal pressure hydrocephalus, cerebrospinal fluid protein

Introduction

Since normal pressure hydrocephalus (NPH) was first reported by Hakim and Adams⁷⁾ in 1965, many such cases have been encountered. The condition is most often secondary to subarachnoid hemorrhage, trauma, or meningitis; it is rarely associated with spinal tumors.

We treated a patient with a cauda equina neurinoma accompanied by NPH whose symptoms completely resolved after tumor removal. We present the details of this case and discuss the mechanism of hydrocephalus secondary to spinal tumors.

Case Report

A 45-year-old male was hospitalized with chief complaints of gait disturbance, urinary incontinence, and memory impairment. He had first noted gait

disturbance in August of 1986, while ascending and descending stairways, and attributed this difficulty to aging. At the same time, however, he became forgetful, to the extent that it interfered with his work. In September of 1986, he developed dull headaches and nocturnal urinary incontinence, and on September 22, 1986, was hospitalized for evaluation.

On admission, he exhibited no motor or sensory disturbance. Neither nuchal stiffness nor spinal tenderness on percussion was observed. He had no low back pain on coughing. His tandem gait was normal, and Romberg's sign was absent. The Achilles tendon reflex was normal. He was alert and attentive. Neurological examinations relating to lumbosacral lesions revealed slight anal sensory disturbance but normal urinary bladder function. Blood chemistries and biochemical data were normal. No abnormalities were found on plain x-rays of the skull and spine and cerebral angiograms. A computed tomographic (CT) scan of the head revealed a cyst in the cavum septi pellucidi and dilatation of both lateral and the third ventricles (Fig. 1).

Fig. 1 CT scan showing markedly dilated ventricles and a cyst in the cavum septi pellucidi.

Fig. 2 CT scans taken 6 hours after intrathecal metrizamide injection, showing full reflux into all ventricular systems.

A lumbar puncture was performed between the fourth and fifth lumbar vertebrae, and 8 ml of 170 mg I/ml metrizamide was infused. The metrizamide first appeared in the ambient and quadrigeminal cisterns 1 hour after infusion. The cerebral sulci were visualized at the same time. Metrizamide flowed into the ventricles after 3 hours and was diffusely distributed throughout the ventricles after 6 hours (Fig. 2). Communicating hydrocephalus was diagnosed from these findings. Analysis of the cerebrospinal fluid (CSF) obtained during lumbar puncture revealed normal pressure (150 mmH₂O) and marked xanthochromia. Although the cell count was 1/3 and the sugar level 53 mg/dl, the protein concentration was markedly elevated (640 mg/dl). Both the Pandy and Nonne-Apelt reactions were strongly positive, and fibrin deposits were observed. The protein fractions were as follows: albumin 61.6% (394 mg/dl), α_1 -globulin 3.8% (24 mg/dl), α_2 -globulin 10.8% (69 mg/dl), β -globulin 9.5% (61 mg/dl), and γ -globulin 14.4% (92 mg/dl). These values were nearly the same as those of serum proteins. The CSF immunoglobulin fractions were IgG 80 mg/dl, IgA 8 mg/dl,

and IgM 3 mg/dl.

Myelograms revealed a clearly demarcated, circular filling defect from the lower margin of the fifth lumbar vertebra to that of the second sacral vertebra, as well as swelling of nerve roots. However, the spinal dural canal was not completely obstructed (Fig. 3). CT myelograms also showed a low-density area with a smooth margin in the spinal canal at the first sacral vertebral level (Fig. 4).

An ependymoma or neurinoma arising from the cauda equina was suspected, and the tumor was removed on November 14, 1987. With him in the prone

Fig. 3 Metrizamide myelograms, revealing a filling defect at the lower L5-S2 levels.

Fig. 4 CT myelogram at the S1 level, showing a radiolucent, oval mass.

Fig. 5 CT scan taken 3 months after surgery, showing the decreased ventricular size and appearance of the cortical sulci.

position, an 8-cm skin incision was made from the spinous process of the fifth lumbar vertebra along the sacral processes. The arches of the first and second sacral vertebrae were resected, and a longitudinal incision was made in the dura. A dark red, parenchymatous, soft elastic, encapsulated mass was found under the arachnoid. The tumor was slightly on the right side; the normal cauda equina fibers were pressed to the left. The tumor appeared to originate in a right cauda equina nerve fiber. The tumor, which measured $2 \times 2 \times 2.5$ cm and was elliptical, and the cauda equina nerve fiber were removed en bloc. Histological examination showed the tumor to consist of long, fusiform cells; it was diagnosed as an Antoni-B type neurinoma with myxoid degeneration.

Postoperatively, the urinary incontinence and gait disturbance improved. A lumbar puncture performed 1 month after surgery showed the CSF to be aqueous and transparent, with a pressure of 125 mmH₂O and a protein level of 25 mg/dl. A CT scan of the head obtained 3 months after surgery revealed diminished ventricles and the appearance of cerebral sulci (Fig. 5). Within 5 months postoperatively, his memory disturbance disappeared.

Discussion

The development of hydrocephalus and papilledema secondary to spinal tumors was first reported by Kyrielleis in 1931,¹⁰ and 66 cases have since been documented.^{2,8,19} However, there have been very few reported cases of spinal tumors in which mental symptoms were among the chief complaints. The phenomenon was first reported by Neil-Dwyer¹⁵ in 1973, only nine additional cases have been reported to date.^{2,3,10,12,13,17,18} These cases, as well as ours, are summarized in Table 1. All but one pa-

tients were middle-aged or elderly, and there was no sex difference. In addition to mental symptoms, gait disturbance occurred in six and urinary incontinence in four. It is noteworthy that this symptom complex resembles that of NPH. In all 10 cases in which lumbar puncture was performed, the protein level in the lumbar CSF exceeded 500 mg/dl and was much higher than that in the cerebral ventricles and cisterns. The histological diagnoses were neurinoma in five cases, neurofibroma in three, ependymoma in two, and oligodendroglioma in one. Ependymoma was associated with initial symptoms of intracranial hypertension or choked disk, whereas neurinoma and neurofibroma frequently produced gradually progressive hydrocephalus and, eventually, mental symptoms. This difference is attributed in part to the difference in anatomy of tumor vessels. Many ependymomas are diagnosed early, since spinal symptoms usually appear suddenly, as a result of subarachnoid hemorrhage from the tumor. Since many reported patients did not complain of spinal symptoms at the time mental symptoms became severe, the spinal symptoms were detected only after hydrocephalus had been diagnosed, ventricular shunting had been performed, and the mental symptoms had been alleviated. Backache and spinal root symptoms are often absent in cases of spinal tumors arising from the cauda equina, and this should be kept in mind. Recent advances in diagnostic techniques, especially imaging technology, have improved the detection rate of spinal tumors. In five cases, including ours, only tumor removal improved or eliminated mental symptoms, urinary incontinence, and gait disturbance. In these cases, postoperative CT scans revealed diminished ventricles and spinal fluid protein levels were normalized.

There are many hypotheses concerning the mechanism by which spinal tumors cause hydrocephalus.^{1,4-6,11,18} In our case, the cerebrospinal canal was not completely blocked by the tumor, nor was subarachnoid hemorrhage observed, as occurs in Fincher's syndrome, which is characterized by bloody spinal fluid due to hemorrhage from a spinal tumor and choked disc. Evidence from animal experiments indicates that the cauda equina is an important site for absorption of CSF,^{4,20,21} and this is supported by the fact that hydrocephalus occurs after radical surgery for spinal meningocele.^{4,20} In our patient, the spinal subarachnoid space was enlarged, suggesting that the spinal tumor disturbed the normal spinal fluid circulation and thereby caused the hydrocephalus.

However, this explanation alone is not sufficient. Norstrom *et al.*¹⁶ found intracranial hypertension in

Table 1 Summary of reported cases of lower spinal tumors associated with mental symptoms

Author (Year)	Age, Sex	Chief symptoms	CSF	Histological diagnosis, tumor location	CT, PEG, and/or RI cisternography	Treatment→outcome
Neil-Dwyer (1973) ¹⁵	66, M	depression, dementia, paraparesis	lumbar protein: 2800 mg/dl, ventricular protein: 50 mg/dl	neurofibroma, L3	PEG: communicating hydrocephalus	ventricular shunt→back pain, TR→complete recovery
Rau (1974) ¹⁷	65, F	dementia, memory disturbance	lumbar protein: 880 mg/dl	ependymoma, L5	—	TR only→normal mentation
Bamford and Labadie (1976) ²¹	75, M	confusion, gait disturbance, paraparesis	xanthochromic, lumbar protein: 510 mg/dl	neurofibroma, L5	PEG: communicating hydrocephalus	repeated lumbar puncture→mental status improvement, TR→normal mentation, cisternal protein: 21 mg/dl
Koyama <i>et al.</i> (1977) ¹⁰	58, F	confusion, disorientation, dementia	xanthochromic, lumbar protein: 2700 mg/dl	neurinoma, L1	PEG: communicating hydrocephalus	ventricular shunt→no change, TR→normal mentation
Ridsdale and Moseley (1978) ¹⁸	49, F	headache, depression, gait disturbance, memory disturbance	xanthochromic, lumbar protein: 3200 mg/dl, ventricular protein: 203 mg/dl	neurofibroma, Th12-L1	PEG: communicating hydrocephalus	ventricular shunt→improved alertness, back pain, TR→complete recovery
	62, F	confusion, dementia, vomiting, gait disturbance	xanthochromic, lumbar protein: 2000 mg/dl, ventricular protein: 99 mg/dl	neurinoma, L2-3	PEG: communicating hydrocephalus	ventricular shunt→leg pain, TR→complete recovery
	21, M	headache, confusion	lumbar protein: 945 mg/dl, ventricular protein: 48 mg/dl	oligodendroglioma, L5-S1	CT: ventriculomegaly, periventricular lucency	steroid and irradiation→death
Messer and Brinker (1980) ¹³	73, F	confusion, gait disturbance, back pain, urinary incontinence	xanthochromic, lumbar protein: 1200 mg/dl	neurinoma, L1	CT: ventriculomegaly, periventricular lucency	TR only→normal mentation, ambulatory with cane, ventricular size decrease
Lusins and Kotsilimbass (1983) ¹²	59, M	gait disturbance, urinary incontinence, dementia	xanthochromic, lumbar protein: 1020 mg/dl	ependymoma, lumbar area	RI cisternography: ventricular reflux, NPH	TR only→disappearance of symptoms
Feldmann <i>et al.</i> (1986) ³	68, F	confusion, back pain, memory disturbance, urinary incontinence	cisternal protein: 106 mg/dl	neurinoma, L2-4	CT: ventriculomegaly	TR only→complete recovery, ventricular size decrease
Present case	45, M	memory disturbance, gait disturbance, dull headache, urinary incontinence	xanthochromic, lumbar protein: 640 mg/dl	neurinoma, L5-S2	CT: ventriculomegaly, metrizamide CT: ventricular reflux	TR only→complete recovery, ventricular size decrease, lumbar protein: 25 mg/dl

PEG: pneumoencephalography, RI: radioisotope, TR: tumor removal.

only three of 101 patients with cauda equina tumors. Further, hydrocephalus does not accompany epidural tumors or spinal herniation, in which the CSF circulation in the subarachnoid space is completely blocked.

The most widely accepted hypothesis is that the increase in the spinal CSF protein or its fractions delays absorption of spinal fluid.^{4,5} For example, patients with Landry-Guillain-Barré syndrome have hydrocephalus and choked disc and exhibit symptoms very similar to those of patients with hydrocephalus due to spinal tumors.^{9,14} This implicates the increased CSF proteins in the onset of hydrocephalus. Among the cases described in Table 1, the spinal fluid protein level was higher than 500 mg/dl in all patients with mental symptoms attributed to

hydrocephalus. Moreover, in many cases tumor removal was followed by normalization of the protein level, disappearance of hydrocephalus, and improvement of mental status.

Various mechanisms have also been proposed to explain the increase in CSF protein. According to one theory,¹ the tumor itself releases a substance that causes the protein content to rise. Another hypothesis holds that the protein increase is mediated by immunological factors, since elevated IgG levels have been found in the spinal fluid obtained from patients with spinal cord tumors accompanied by hydrocephalus.⁵ In our case, however, IgG in the spinal fluid increased to 80 mg/dl but accounted for only 12.5% of the total CSF protein. We found that the spinal fluid protein fractions paralleled those of

serum. If the increase in CSF protein were immunologically mediated, one would expect higher γ -globulin and IgG values, as occurs in tuberculous sclerosis and meningitis.

Deposition of fibrin and the presence of IgM in CSF are thought to reflect dysfunction of the blood-brain barrier. We believe, as was pointed out by Bamford and Labadie,²⁾ that in the cases under discussion the elevation in spinal CSF protein was due simply to leakage of serum protein from newly generated tumor vessels having no blood-brain barrier function. Since the meninx is not irreversibly damaged by an increase in CSF protein alone, it can be assumed that after our patient's tumor had been removed, the protein that had seeped from its vessels was gradually absorbed, resulting in resolution of the hydrocephalus. However, the mechanisms involved in the onset of hydrocephalus remain hypothetical, and further study is required.

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