

Lumbar Schwannoma Associated with Hydrocephalus

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We describe a rare case of 52-year-old woman with lumbar schwannoma associated with hydrocephalus. In our case, the signs and symptoms of intracranial hypertension were not resolved even after the complete removal of the lumbar schwannoma. We also reviewed the literature on the association of hydrocephalus with spinal cord tumor.

KEY WORDS : Hydrocephalus · Schwannoma.

Introduction

The spinal cord tumors associated with hydrocephalus are unusual. The association of hydrocephalus with benign extramedullary spinal cord tumor is particularly rare. Complete removal of the intraspinal lesion, in the absence of leptomeningeal metastatic disease, is therapeutic for eradicating the signs and symptoms of intracranial hypertension in the majority of cases^{7,14,17}. Increased protein content in cerebrospinal fluid (CSF) plays an important role in causing intracranial hypertension even though it does not explain all the cases. In our case, the protein level of CSF was also high on initial spinal tapping and normalized after the complete removal of the lumbar schwannoma. But it was not therapeutic for eradicating the signs and symptoms of intracranial hypertension. So we placed a ventriculoperitoneal shunt using a programmable valve.

Case Report

On September 14, 2001, a 52-year-old right-handed woman was admitted to the neurology department of our hospital with the chief complaint of an aggravated headache and gait disturbance. On admission, there was no mental change, memory disturbance or urinary symptoms. The patient had received antihypertensive drugs for 6 years

and antidiabetic drugs for 1 year. The findings of general examination were unremarkable and the neurologic examination demonstrated severe bilateral papilledema and constricted visual fields on automatic perimetry. Visual acuity was 20/25 in the right eye and 20/33 in the left. The brain computed tomography (CT) showed the generalized ventricular enlargement (Fig. 1A).

On September 17, 2001, a lumbar puncture was performed on the L3-4 level for radioisotope cisternography. The opening pressure of the spinal tapping was 21 cmH₂O and the

CSF was a clear deep orange color. About 5 ml CSF was drained for analysis and 1 ml radioisotope was inserted to intrathecal space. The CSF protein was elevated (500 mg/dl) and the cytology of CSF was negative. The obtained radioisotope cisternography showed no radioisotope signal above the upper lumbar. After the spinal tapping, the patient complained of back pain, severe radiating pain in both legs, an aggravated headache and drowsiness.

On September 18, 2001, the patient was transferred to our department. An external ventricular drainage (EVD) was placed immediately and the puncture pressure was 20 cmH₂O and the CSF protein was elevated (298 mg/dl). After the EVD, the aggravated headache and then drowsiness improved, but the spinal symptoms did not improve. The high concentration of protein in the CSF and the findings of the radioisotope cisternography suggested impaired CSF communication between lumbar spinal and more proximal compartments. We performed a lumbar magnetic resonance imaging (MRI) for evaluating the cause of the intraspinal CSF blockage.

A lumbar spinal MRI scan (Fig. 2) documented an extramedullary mass lesion, located from the upper level of the L1 body to the upper level of the L3, 8.5 cm in length. The mass contained cystic components in the upper and lower poles of

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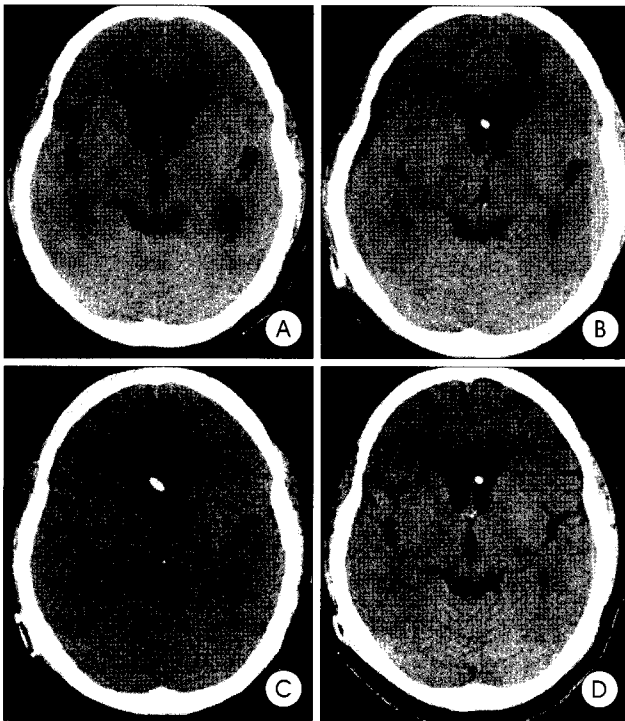


Fig. 1. A : Initial brain computed tomography(CT) scan demonstrating the enlarged frontal horn of lateral ventricle and effacement of sulci. B : Brain CT, obtained one week after the ventriculoperitoneal shunt surgery, demonstrating subdural fluid collection on the right frontotemporal area. The size of frontal horn is decreased but the sulcus markings are still not apparent. C : Brain CT, obtained one month after the readjustment of valve pressure to 17cmH₂O, demonstrating no more subdural fluid collection on the right frontotemporal area and the apparent sulcus marking. D : The latest brain CT, demonstrating no definite interval change compared with previous CT.

the mass. The solid component of the mass showed heterogeneous high signal intensity on T2-weighted image and isointensity on T1-weighted image and well enhancement with gadolinium on T1-weighted image. There was no pathologic alteration of marrow signal intensity.

On September 20, 2001, the patient underwent a total laminectomy from T12-L3 for tumor removal. After dural opening, a capsulated solid wellvascularized mass adherent to the nerve roots of the cauda equina was found (Fig. 3A). The tumor was completely removed by microsurgical technique without damaging nerve roots (Fig. 3B). Histologically, it was schwannoma (Fig. 3C, D). The spinal symptoms were improved after the removal of the tumor. The EVD pressure was monitored postoperative day (POD) 8. The protein content of CSF started to decrease gradually from POD 1 (83mg/dl) to POD 8 (23mg/dl). The EVD pressure was persistently high (mean: 18.4-27.3 cmH₂O) and amounts of CSF drainage (mean : 291ml/day) at the level of EVD height 25~30cm above the external auditory canal did not decrease during follow-up days. On POD 8,



Fig. 2. Preoperative lumbar magnetic resonance(MR) image demonstrating an extramedullary mass lesion, located from the upper level of the L1 body to the upper level of the L3, 8.5cm in length. The mass containing cystic components in the upper and lower poles of the mass. The solid component of the mass showing a heterogeneous signal intensity on T2-weighted image(A) and isointensity on the T1-weighted image(B) and well enhancement with gadolinium on T1-weighted image(C). There is no pathologic alteration of marrow signal intensity.

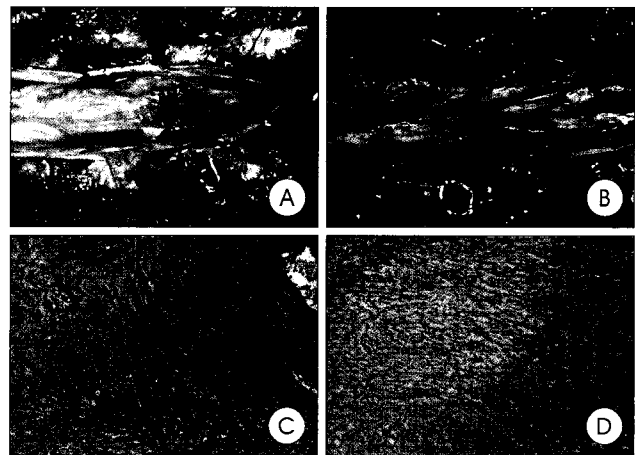


Fig. 3. Operative findings demonstrating a capsulated solid wellvascularized mass adherent to the nerve roots of the cauda equina(A), and complete removal of tumor by microsurgical technique without damaging nerve roots (B). Histopathologic findings revealing typical schwannoma (C&D). C : The micrograph shows high cellular area of elongated spindle cells with long cytoplasmic processes arranged in interwoven fascicles (Antoni A area) (H&E stain, ×100). D : The micrograph shows loose hypocellular portion with myxoid and microcystic changes (Antoni B area). Nuclei of tumor cells are small round and regular (H&E stain, ×100).

the EVD was clamped for 4 hours, and then the patient complained of a headache and EVD pressure was 47cmH₂O at reopening the EVD clamp. So we thought it was dangerous to remove the EVD without any other CSF diversion system. The change of CSF component and EVD pressure and

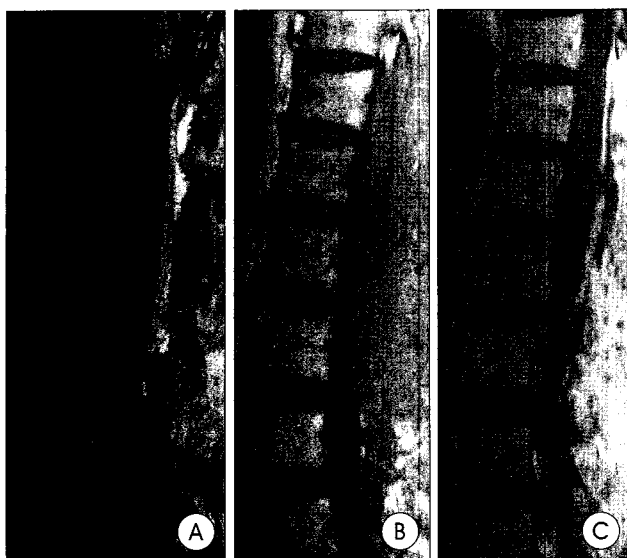


Fig. 4. Follow-up MR image (T2-weighted image(A), T1-weighted image(B), gadolinium enhanced T1-weighted image(C)) of the lumbar spine, obtained one year after lumbar surgery, shows no recurrence of the lumbar tumor.

Table 1. Changes of cerebrospinal fluid components, external ventricular drainage pressure and drainage amount

No. (day)	WBC (μl)	RBC (HPF)	Protein (mg/dl)	Glucose (mg/dl)	Pressure (cmH ₂ O)	Drainage(cc)/EVD height(cm)
1*	0-1	0-1	500	44	21	5
2**	20	many	298	113	20-27	444/25
3	18	15-25	111	138	23-36	102/25
4***	15	15-20	83	89	20-28	229/25
5	5	many	71	99	20-27	325/25
6	3-5	many	46	86	21-26	346/25
7	14	many	38	94	21-28	275/25
8	13	10-15	38	108	14-28	234/25
9	7	9-12	27	91	15-27	303/30
10					15-30	222/30
11	2	10-15	28	98	16-26(47 †)	235/30
12	3	20-25	23	70		

1* : spinal tapping was done, 2** : EVD was done, 4*** : tumor removal was done, 47 † : EVD pressure when reopening the EVD after 4 hours clamp. HPF : high power field($\times 400$)

drainage amount is listed on table 1. A ventriculoperitoneal shunt was placed using a programmable valve on September 29, 2001. The valve pressure was adjusted to 15cmH₂O. One week after shunt surgery, a brain CT showed subdural fluid collection on right frontotemporal area (Fig. 1B). Overdrainage of the CSF was suspected and the valve pressure was readjusted to 17cmH₂O. A one month follow-up brain CT showed the subdural fluid collection on right frontotemporal area disappeared (Fig. 1C). One year after surgery, a follow-up MRI of the lumbar spine showed no recurrence of the lumbar tumor (Fig. 4). And the neurologic examination

demonstrated disappearance of bilateral papilledema. Visual acuity improved to 20/20 in the right eye and 20/20 in the left. But the constricted visual field did not improve. On latest follow-up, on July 5, 2004, the brain CT (Fig. 1D) showed no interval change and the patient has enjoyed good health and resumed her premorbid work after the initial operation.

Discussion

Hydrocephalus associated with spinal tumor is well known but not common phenomenon. Histologically, the most common neoplasm is ependymoma followed by malignant tumors, neurinoma, and meningioma^{3,7,17}. The incidence of intraspinal tumors revealed by increased intracranial pressure is about 1%⁹.

The pathogenesis of a benign spinal lesion association with hydrocephalus or intracranial hypertension remains unclear and various explanations are proposed for this phenomenon.

A mechanical explanation is based on the observation that the spinal subarachnoid space serves as an elastic reservoir to buffer the physiological variations of CSF pressure determined by rapid modifications of arterial pressure, venous pressure, and body position⁹. The anatomical and functional isolation of the spinal subarachnoid space from the intracranial compartment caused by the presence of a spinal obstruction could prevent the compensation of CSF pressure variations and cause papilledema and ventricular dilation.

Other authors^{3,8,17} have proposed that in some patients the spinal pathways of CSF resorption around the spinal nerve sheaths could be more abundant than in the normal population, and subsequently their functional exclusion due to a subarachnoid blockage induced by a spinal tumor could explain the impairment to CSF resorption and the development of intracranial hypertension.

A widely accepted hypothesis is that elevation of protein in CSF occlude the arachnoid granulation or retard CSF flow by increasing viscosity, resulting in hydrocephalus^{1,4,6,15}. Many authors have discussed the origin of the elevated CSF protein associated with spinal cord tumors. Suggested mechanisms include changes in permeability of extradural blood vessels as a result of direct pressure, breakdown of tumor material¹⁶, hemorrhage from the tumor¹¹, transudation through tumor vessels⁹, and active secretion of proteins by tumor⁵. In our case, the protein content in CSF was elevated on initial spinal tapping. Immediately after complete removal of lumbar mass, the protein content in CSF obtained from the EVD started to decrease and finally normalized on POD 8. Subsequent normalization of the CSF protein content after removal of

spinal tumor may explain that spinal tumor may play an important role in elevating the protein content in the CSF. However, this hypothesis does not explain in hydrocephalus in patients with normal or slightly elevated CSF protein concentration proximal to a spinal subarachnoid obstruction^{1,6,7,13,17}. The other explanation for this phenomenon include arachnoiditis¹⁰, leakage of fibrinogen into the CSF^{1,2,12}. Findings in our case suggest that reduced spinal subarachnoid buffer space, impaired spinal pathways of CSF resorption and elevated proteins in the CSF may be significant causative factors of hydrocephalus.

Complete surgical excision of the intraspinal lesion, in the absence of leptomeningeal metastatic disease, is therapeutic for eradicating the signs and symptoms of intracranial hypertension in the majority of cases^{7,14,17}. We also expected that complete removal of the lumbar schwannoma would improve the intracranial hypertension and shunt surgery could not be necessary. And then we planned to observe the changes of CSF components and pressure through the EVD system. We already put a EVD with a long subcutaneous tunneling (15cm) and it made possible to observe CSF changes for a long period than usual without CSF infection. The protein content of CSF started to decrease gradually from POD 1 but the EVD pressure was persistently high and amounts of CSF drainage did not decrease during follow-up days. On POD 8, the EVD was clamped for 4 hours, and then the patient showed signs of intracranial hypertension. So we decided to put a ventriculoperitoneal shunt with programmable valve and then readjusted the valve pressure 1 week later due to the overdrainage of CSF.

Conclusion

In the case of unexplained hydrocephalus, a spinal tumor can be highly suspected if the ventricular dilatation exists in association with increased CSF protein. In this case, a MRI would be advisable. The occurrence of hydrocephalus in association with a benign spinal lesion may not be attributed to single etiologic or pathogenetic factor. Although the CSF diversion system after removal of the lumbar tumor can be

avoided, sometimes it may be necessary, depending on the clinical conditions.

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