



Case Report

Cerebrospinal fluid shunt for normal pressure hydrocephalus patient exacerbates cord symptoms due to spinal tumor

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ABSTRACT

Background: Normal-pressure hydrocephalus (NPH) and spinal intradural extramedullary benign tumors rarely exist together. Here, a 72-year-old female who presented with NPH symptoms (i.e., gait disturbance and dementia) newly developed symptoms of spinal cord compression attributed to a previously undiagnosed schwannoma.

Case Description: A 72-year-old female was diagnosed with NPH without disproportionately enlarged subarachnoid space hydrocephalus. The lumbar puncture revealed an elevated cerebrospinal fluid (CSF) protein level of 0.141 g/dl, but with normal pressure. The patient's NPH symptoms improved after lumbar-peritoneal shunt placement. However, a year later, she subacutely developed a progressive Brown-Sequard syndrome. On the cervical magnetic resonance (MR), an intradural extramedullary lesion was found at the C5-C6 level which at surgery, proved to be a schwannoma. A review of this patient and three others with NPH and intradural extramedullary benign tumors revealed that 4.3 months following CSF shunting for NPH, they developed rapidly progressive cord deficits, attributed to their benign spinal tumors.

Conclusion: Before the placement of shunts for NPH, patients should undergo holospinal MR imaging studies to rule out attendant spinal intradural extramedullary tumors.

Keywords: Cerebrospinal fluid protein, Disproportionately enlarged subarachnoid space hydrocephalus, Normal-pressure hydrocephalus, Shunt surgery, Spinal schwannoma

INTRODUCTION

The concurrence of normal-pressure hydrocephalus (NPH) and spinal intradural extramedullary tumors has rarely been reported.^[7] Here, we evaluated a 72-year-old female who following shunting for NPH developed myelopathy attributed to a previously undiagnosed C5-C6 schwannoma.

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CASE REPORT

A 72-year-old female presented with increased dementia (Mini-Mental State Examination [MMSE] score = 11) and gait disturbance. The brain computed tomography (CT) and magnetic resonance (MR) studies confirmed enlarged ventricles without a brain tumor; she was diagnosed with non disproportionately enlarged subarachnoid space hydrocephalus (DESH: a form of NPH^[3]) [Figures 1a and b]. The spinal tap revealed clear and colorless cerebrospinal fluid (CSF) with a normal opening pressure (i.e., 160 mmH₂O). Although the CSF showed no cells, the protein level was increased (141 mg/dL). Following a spinal tap, the patient's symptoms improved (i.e., MMSE score = 29, improved gait disturbance). Therefore, she underwent lumbar-peritoneal shunt placement (Codman-Hakim programmable valve set at 160 mmH₂O). Improvement in cognitive function (MMSE score = 29) and gait disturbance were observed postoperatively.

One-year follow-up

A year later, she presented with "abnormal sensations in the left hand and both lower extremities." Within 1 month, findings extended to the right hand and she developed a progressive myelopathy (i.e., weakness in the left upper and both lower extremities 2/5 motor level) with diffuse hyperreflexia and abnormal sensory findings. The brain CT confirmed the ventricular size remained unchanged [Figure 1c]. However, the cervical MR imaging (MRI) with contrast documented a benign intradural extramedullary lesion (i.e., 1.4 × 9.0 × 2.3 cm) at the C5-C6 level [Figure 2]. The entire cervical tumor mass was dissected away from the C6 nerve root; the lesion was positive for S-100 protein, confirming that it was a schwannoma [Figure 3]. Postoperatively, the patient's spinal symptoms improved, and there was no exacerbation of her hydrocephalus.

Of interest, her shunt pressures were maintained at 200 mmH₂O both preoperatively and postoperatively and did not require further adjustment potentially indicating that shunt placement had been unnecessary.

DISCUSSION

Notably, in this patient with a diagnosis of NPH, the CSF cell protein elevation reflected the presence of an accompanying spinal tumor (i.e., range 0.03–4.073 g/dl).^[5,7] Wajima *et al.* reviewed similar cases with NPH and spinal intradural extramedullary spinal tumors; in all cases, the intracranial pressure was normal, but enlarged ventricles were observed.^[7] Notably, most of their tumors occur in the lumbar spine, with two cases being found in the cervical region.^[5-7]

Cause of NPH with spinal tumors

There are several considerations regarding the cause of NPH with spinal tumors.^[2,4,7] An increase in tumor-related CSF protein likely contributes to an aseptic arachnoiditis and adhesions, resulting in poor CSF flow resulting in reflux and hydrocephalus.^[7] Vestibular schwannomas may also impact CSF flow through similar mechanisms.^[1] In Wajima *et al.*, the 13 patients with both NPH and intradural extramedullary lesions underwent tumor removal before shunting avoid shunt obstruction due to high-protein levels; nine of 13 patients improved with tumor removal alone without the need for shunt placement.^[7]

Avoiding NPH shunt placement before spinal tumor removal to avoid precipitous neurological deterioration

Wajima *et al.*, 13 patients with both NPH and intradural extramedullary spinal lesions underwent tumor removal before shunting with nine subsequently improving and not

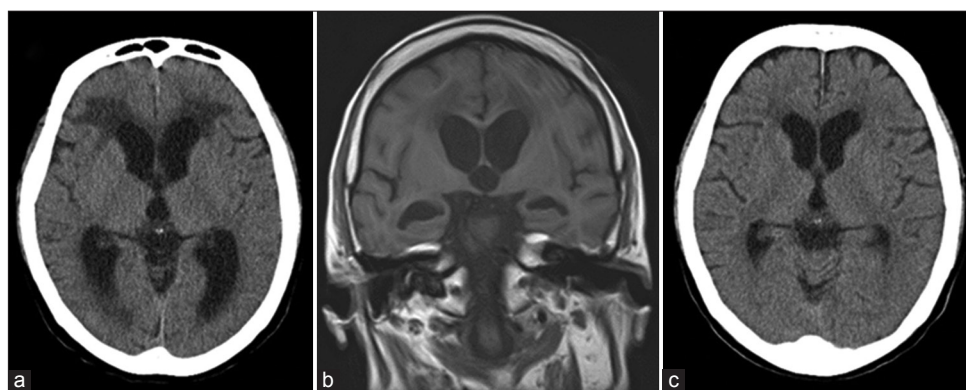


Figure 1: Head computed tomography (CT) scan (a) and head magnetic resonance imaging (MRI; b) at onset showing enlargement of the ventricles. No typical disproportionately enlarged subarachnoid space hydrocephalus findings can be seen. There is no evidence of exacerbation of the hydrocephalus, such as enlargement of the ventricle, or over drainage on the head CT scan (c) when symptoms worsened.

Table 1: Reported cases of NPH shunting performed before intradural extramedullary spinal tumor removal.

Case no.	Author (year)	Age/Sex	CSF protein (g/dl)	Xanthochromia	Drainage or shunt	Duration of diagnosis	Level of tumor	Pathology
1	Kudo (1987) ^[6]	55/F	0.98	—	VPS	3 months	C2-4	Schwannoma
2	Koshu (1993) ^[5]	60/F	0.03	—	VPS	P.O. paraplegia	C4-5	Schwannoma
3	Present case	74/F	0.141	—	LPS	1 year	C5-6	Schwannoma
4	Kudo (1987) ^[6]	49/F	3	—	VPS	2 months	T12-L1	Schwannoma

F: Female, CSF: Cerebrospinal fluid, VPS: Ventriculoperitoneal shunt, LPS: Lumbar-peritoneal shunt, P.O: Postoperative, C: Cervical, T: Thoracic, L: Lumbar

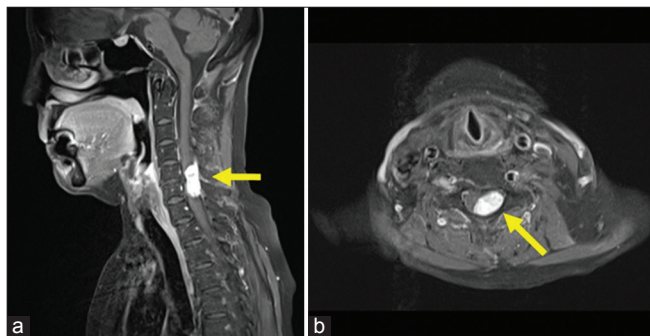


Figure 2: A cervical contrast-enhanced magnetic resonance imaging examination (a and b) showing a 1.4 × 9.0 × 2.3 cm neoplastic lesion (arrow) in the dura mater at the C5-C6 level. This is diagnosed as an intradural extramedullary tumor.

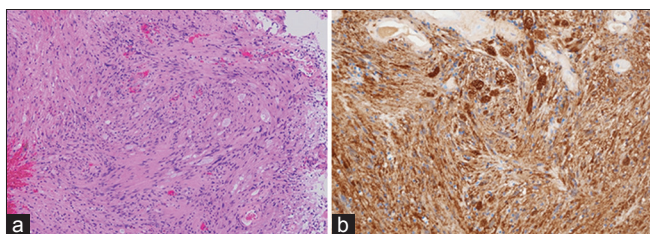


Figure 3: Histopathological examination of an intradural extramedullary tumor. (a) Photomicrograph showing a bundle of spindle-shaped cells, some of which have a shelf-like arrangement of nuclei. Hematoxylin and eosin staining, ×100. (b) Immunohistochemical staining for S-100 protein showing strong and diffuse positive staining of the tumor cells. ×400.

requiring shunt placement.^[7] In four cases, including ours, NPH shunting performed before tumor removal resulted in the rapid progression of spinal cord deficits over an average period of just 4.3 postoperative months [Table 1: cases 1, 2, 3, and 4].^[5,6] We, therefore, conclude that NPH shunting be avoided before spinal tumor removal to avoid exacerbating tumor-related spinal cord deficits; further, symptoms spontaneously resolve following tumor excision alone without shunting in the majority of these patients.

CONCLUSION

A 72-year-old female with NPH became symptomatic 1 year later from a C5-C6 spinal intradural extramedullary schwannoma. This study demonstrates that before shunting for NPH, patients should undergo holospinal MRI studies to rule out accompanying spinal lesions that may be contributing to/responsible for NPH-like symptoms/signs due to their production of high-protein levels with resultant interference with CSF flow dynamics.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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