Cervical Neurinoma associated with Hydrocephalus: Case Report

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ABSTRACT

A 64-year-old woman with a 3-month history of gait disturbance and left leg numbness followed by headache, dementia and urinary incontinence. Head computed tomography (CT) revealed marked ventriculomegaly and magnetic resonance imaging with gadolinium contrast showed an enhanced mass at C2-3. Excision of neurinoma resulted in improvement of both gait and memory disturbance with no urinary incontinence. Follow up head CT demonstrated ventricular size became smaller without periventricular lucency. In this case, mechanical obstruction of the cerebrospinal fluid (CSF) pathway resulted in alteration of the spinal subarachnoid space function as a “buffer reservoir” to cranial CSF circulation, and this can play a critical role, particularly in patients with normal CSF protein concentrations.

Key words: cervical spine, neurinoma, hydrocephalus, ventriculomegaly

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Introduction

The pathophysiology of the alteration of cerebrospinal fluid (CSF) circulation is understandable in cases of malignant lesions, which have a natural tendency to spread in the subarachnoid pathways. Hydrocephalus appeared to be directly related to neoplastic meningeal infiltration and or direct tumor extension [16,18]. It is generally assumed that the spread of tumor in the subarachnoid space creates an increase in the flow resistance, explaining the ventricular enlargement and elevated intracranial pressure [4].

On the other hand, the relationship between benign lesions, particularly cervical spinal neurinoma and hydrocephalus is much more controversial. Several pathophysiological explanations have been proposed, such as increase CSF viscosity because of elevated fluid protein content, blockage of the spinal subarachnoid pathways of CSF resorption, spinal subarachnoid hemorrhage, localized arachnoiditis, congenital abnormality, and coincidence [3,8,18]. We present a case in which a patient developed hydrocephalus with C2-3 neurinoma and normal CSF protein concentration, then discuss the proposed mechanisms for the production of intracranial symptoms.

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Case Report

A 64-year-old woman had noted a gradual onset of gait disturbance and numbness in her left leg since three months before admission. Then, she began to have urinary incontinence and memory disturbance, and was admitted to our hospital. Magnetic resonance imaging with gadolinium contrast demonstrated an enhanced intradural extramedullary mass of 10 x 20 mm at C2-3 level. Head CT demonstrated marked enlargement of ventricular systems with apparent periventricular lucency but no focal lesions or obstruction to CSF flow (Fig.1 and 2A). Neurological examination revealed dementia, spastic gait, right biceps and right knee jerk hyperreflexia, hypalgiesia at right S1 and L4 dermatome. There were no papilledema, no signs of motor weakness, grasping power was 12 kg on the right and 10 kg on the left hand. Lumbar puncture before operation for CSF pressure measurement was not done to avoid neurological deterioration, because MR imaging showed obstruction at C2-3 level. We suspected the hydrocephalus associated with this cervical tumor.

The patient underwent tumor removal in the prone position with her head fixed on Sugita frame. Somatosensory evoked potential (SEP) and motor evoked potential (MEP) were prepared for intraoperative monitoring. Unilateral laminotomy was performed at C2 and C3 on the left side by using ultrasonic bone scalpel (SONOPET, MdM Co Tokyo Japan). The bulging dura was incised in a curvilinear fashion. A tumor of 10 x 20 mm in the intradural-extramedullary space was located posteriorly to the dentate ligament. The mass was yellowish, and compressing the spinal cord and nerve roots anteriorly and toward right side. Both intraoperative frozen section and the fixed staining sections demonstrated the Antoni type A cells with ‘palisading’ patterns. So, final pathological diagnosis was neurinoma. The rostral part of tumor which was adhered to the cord was successfully dissected and the tumor was totally removed. SEP and MEP showed no significant changes during all procedures.

CSF obtained from the subarachnoid space around the tumor and before the tumor removal during the operation contained 31 mg/dL of protein. She has been followed up for 6 months, on examination showed no gait disturbance, no urinary incontinence or dementia. Head CT follow up on the third, fourth and sixth month revealed similar view of smaller
Discussion

In this case, we found the triad symptoms of normal pressure hydrocephalus (NPH) which is usually described as gait disturbance, dementia and urinary incontinence [2,14,18]. However, it is not simple to diagnose NPH only by symptoms. Lumbar puncture for CSF pressure measurement was not done because of spinal obstruction to avoid neurological deterioration. We can not diagnose definitely as NPH since the measurement of intracranial pressure should be done as one of the minimum requirements for the diagnosis, so we can only mention it as the most suspected diagnosis. Hypertensive hydrocephalus could demonstrate the similar clinical and radiological findings. On head CT before operation, the patient had apparent periventricular lucency with normal tension. After removal of spinal tumor the patient had a significant improvement in patient’s disorientation, gait disturbance and urinary incontinence.

Any theories proposed to explain the pathogenesis of hydrocephalus associated with spinal tumors must take into account the following observations: Only 1% of patients with spinal cord tumors will present with signs and symptoms of hydrocephalus [3]. Twelve cases of neurinoma associated with hydrocephalus were reported in the literature. Most common location was thoracolumbar region and have elevated CSF protein concentration [3, 6-8, 11-13, 15, 17, 20, 21]. Only one literature was reported hydrocephalus with no elevation of CSF protein, but its pathological relationship still unexplained [7].

Many current theories are not reconcilable with the evidence we found during operations. The suggestion that protein released from the tumor into CSF and causing the elevated CSF protein level [3, 6, 8, 11] leads to hydrocephalus is unlikely in our case, because the CSF obtained from around the tumor was clear and contained normal CSF protein concentration. The proposal that tumor-induced subarachnoid hemorrhage causing hydrocephalus [6, 17] is inconsistent with the fact that we found no changes in the spinal fluid or no evidence of previous hemorrhage episodes. Localized arachnoiditis has been postulated as the cause of hydrocephalus [6] but this also has never been detected during tumor removal. Furthermore, clinically there were no meningeal signs or infection. Finally, the suggestion
that tumor isolation of spinal arachnoid granulations as absorptive pathway of CSF leads to hydrocephalus [21] ignores the fact that indeed this pathway has a small contribution on CSF absorption and no evidence of spinal arachnoid granulations around the tumor or from histopathological sections in our case.

A more plausible theory is mechanical explanation based on experimental study by Martins AN et al.[9] They emphasized that the spinal dural sac serves as an elastic reservoir to buffer the physiological variations of CSF pressure. They observed the contrast material in the subarachnoid space during myelography by changing the CSF volume by withdrawals and rejections of fluid. The spinal dural sac enlarged with increases in volume of CSF. With increases in intra-abdominal and intrathoracic pressure during Valsalva manoeuvre, the thoracolumbar sac partially collapsed, while the cervical sac enlarged. From these observations, they concluded that the spinal dural sac served an ‘elastic reservoir’ to buffer variations of CSF pressure[4, 9]. 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 ventricular dilatation [4]. In our case, mechanical obstruction of CSF pathway which resulted in alteration of spinal dural sac function as a ‘buffer reservoir’ seemed to be the most probable cause for hydrocephalus.

Another issue still exists, the ‘minor pathway’ of spinal subarachnoid space could be predominant in CSF circulation [5]. The role of central canal in the spinal cord and the CSF circulation could not be ruled out in elder person. In those person, spinal subarachnoid space could have wider function more than a buffer. It can play not only a buffer for the increased CSF pressure but also it could be a major CSF pathway.

In conclusion, hydrocephalus associated with cervical neurinoma and normal CSF concentration is relatively rare. The alteration of buffer function and CSF circulation pathway of spinal subarachnoid space due to mechanical obstruction is postulated to play a critical role in causing hydrocephalus in this case.

References

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Reviewer’s comment : Akira Matsumura, M.D., Ph.D.
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This is a unique case report of a spinal neurinoma with normal pressure hydrocephalus (NPH) associated with normal CSF protein. As authors have described in previous literature, the combination of neurinoma and NPH is usually associated with high CSF protein.

It would have been of great interest if RI cisternography or other CSF dynamics studies had documented in this case, to add further information and to clarify the etiology of the NPH. Nevertheless the author’s hypothesis of a “buffer reservoir” theory is an interesting and possible cause of the NPH in this particular case. Based on this valuable case report, further case studies should be documented to clarify the nature of NPH in spinal neurinoma without CSF protein elevation.

Reviewer’s comment : Nobuhito Morota, M.D.
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This is a very unique case report because the author discussed not only the relationship between the spinal cord tumor and hydrocephalus but also the role of spinal subarachnoid space for the CSF circulation. Hydrocephalus is a rare association with the spinal cord tumor, especially with the neurinoma which does not present the high CSF protein. The author attributed hydrocephalus to the loss of butter function in the spinal subarachnoid space by tumor obstruction in the CSF pathway.

It should be reminded that controversies remain for the actual site of CSF production and absorption. The CSF circulation that we know as the major pathway may be different in young or elder people. The minor pathway may play more important role in those generations and could cause hydrocephalus once the communication was disrupted. Individual difference in terms of the CSF circulation and the level of the tumor (the higher the level of CSF obstruction, the greater the loss of spinal buffer space) would also have influenced for developing hydrocephalus in this particular case.

I appreciated the author's deep insight for the mechanism of hydrocephalus associated with spinal cord tumor. It is a pity that the author did not measure the CSF pressure above and below the tumor before the dural opening. I guess there was a pressure gradient between the both spaces if the author’s speculation was right.