Cervical Subependymoma Presenting as an Extramedullary Tumor

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A rare case of cervical subependymoma in a 45-year-old man is described. The tumor appeared as an extramedullary mass lesion, but a discrete, less well-demarcated portion was observed in the anterolateral part of the cord at the C3-C7 level. Previous reports of spinal subependymomas are reviewed, and nosological possibilities of extramedullary presentation are discussed.

KEY WORDS: Extramedullary tumor · Spinal cord · Spinal tumor · Subependymoma.

Introduction

Subependymoma is a rare, benign glial tumor observed mainly in the 4th or lateral ventricles.11,12,13,14,21 It is thought to be derived from the bipotential subependymoma cells in the subependymal layer, and it grows in an expanding rather than infiltrative manner.12,13,14,21 Most of the intraventricular subependymomas are subclinical and thus incidentally encountered at autopsy, whereas the spinal ones are inevitably accompanied by myelopathy and are often diagnosed clinically as ependymomas or astrocytomas.9 Only six cases of symptomatic spinal subependymoma have been reported in literature.2,15-17. These tumors usually originate near the central canal, presenting as intramedullary tumors.2,14,17 Extramedullary spinal subependymoma has been reported only two cases.1,7 We report an additional case of the extramedullary presentation of a spinal subependymoma and discuss this condition.

Case Report

A previously healthy 45-year-old man was admitted to our hospital because of slowly progressive tingling sensation on the right hand and increased deep tendon reflexes on the right side. A plain x-ray film showed a normal finding of the cervical spine (Fig. 1). A cervical MRI showed revealed an elongated high T2 and low T1 signal intensity mass in C3-C7 (Fig. 2). Radiological impression was a tumorous condition such as astrocytoma, vascular malformation and transverse myelitis with hemorrhage. Five days after admission, laminoplasty from C3 to C7 was performed and the tumor was observed as an extramedullary lesion between the right anterior and lateral nerve roots that was compressing the spinal cord to the left (Fig. 3). During tumor extirpation from rostral to caudal, we found that the tumor extended ventrally in a bead-like fashion and was attached in a small region to the spinal cord, showing no clear demarcation between the cord and the tumor. Except for this part, the tumor could be radically excised. After the operation, the tingling sensation on the right hand has worsened. This symptom gradually improved and 2 months after the operation, he is enjoying life without any evidence of tumor recurrence. The histological findings...
Discussion

In review of the literature, including our own, the age of the patients with spinal subependymoma ranged from 16 to 49 years (average 34.1), which was younger than the age of the patients with intracranial subependymomas reported in the literature. There was a marked male proponentance (M:F = 6:1). The duration of the symptoms ranged from 1 to 8 years (average 4.8 years). The duration of the symptoms tended to be shorter when compared with the adult spinal tumor series of Cooper and Epstein, with 8.3 years for benign astrocytoma and 15 years for ependymoma. The relatively short history may well be due to the expanding nature of the tumors, as observed in other studies of the duration of symptoms with infiltrating tumors. In our patient, the tumor was an extramedullary lesion associated with a pedunculated connection to the spinal cord. The tumor was extramedullary with dense adherence to the spinal cord. To explain the unusual extramedullary presentation of the subependymomas in these cases, three possibilities are hypothesized. (a) The subependymoma originates near the central canal, where subependymal biopotential cells usual exist, breaks through the spinal cord into the free subarachnoid space, and increases its size in the extramedullary space. (b) The tumor originates in the marginal gray matter, growing as a pedunculated tumor. (c) The tumor originates from leptomeningeal heterotopic glial tissue and then attaches to the surface of the spinal cord. The first hypothesis seems...
Cervical Subependymoma

Fig. 7. After infusion of the contrast media, no definite abnormality enhancing lesion is seen in the cervical spinal cord. A : Sagittal view. B : Axial view of C 4–5 level.

to be unlikely because subependymomas usually show intramedullary expansive growth and in fact, no massive intramedullary tumor was found in our case. In the second hypothesis, the subependymal cells migrate from the central portion of the spinal cord to the marginal zone during embryological development and then become the source of the tumor. The third possibility, the heterotypic cell origin theory, was first described by Kernohan et al and has been supported by other authors.

Adherence of extramedullary tumors to the spinal cord may occur secondarily. Ho described the simultaneous occurrence of subependymoma and leptomeningeal heterotopia. In the aspect of these three possibilities, we think that occurrence of subependymoma was more higher than the thoracic lesion than the cervical lesion. We did not perform CSF study and radiation therapy because no evidence of metastasis was found on radiologic examination, especially in MRI and the operative finding. And we think that preoperative CSF study is risky procedure because of possibility of herniation. But we think that intraoperative CSF study is necessary. Our intraoperative findings indicate that the tumor originated in the cord itself because the tumor was firmly attached to the anterolateral aspect of the spinal cord and had no clear demarcation from the spinal cord. This suggests that the tumor originated from within the cord rather than as simple adhesion to the cord.

As Kernohan and Hughes stated, the actual incidence of spinal subependymoma is probably masked by the tendency to diagnose subependymomas either as astrocytoma or as ependymoma. Subependymoma should be reported as an independent entity even in spinal processes. Because it grows by expansion and rarely shows recurrence, a successful surgical procedure and clinical outcome may be expected. In our case, the patient's symptom, such as right hand tingling sensation gradually decreased and after a 1 year of follow up, any evidence of tumor recurrence was not found on follow up MRI (Fig. 7). On radiological studies, there was no specific sign of sub-ependymomas. But MRI showed that the tumor was sharply demarcated and had eccentric features. A partial illdefined portion was found. This finding suggested that this tumor was extramedullary. When confronted with a complete intraventricular lesion or with a spinal lesion causing little or no edema which is minimally enhancing or nonenhancing and is sharply demarcated, one must consider the diagnosis of subependymoma.

Conclusion

Spinal subependymoma usually originate near the central canal, presenting as intramedullary tumors. Extramedullary spinal subependymoma is very rare. Only two cases extramedullary spinal subependymoma have been reported. Our case in this paper is the first one since the first case was reported in 2001.

References