

A Case of Rapidly-Progressing Cervical Spine Subependymoma with Atypical Features

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Abstract:

This was a study of the case of a 60-year-old woman who presented with a six-month history of headache and numbness radiating to the right arm. MRI revealed a fusiform intramedullary spinal tumor spanning C2 to C5 at the hospital where she first presented. As her right upper limb weakness had presented gradually, she visited our hospital after one and a half years. Neurological examination revealed muscle weakness in the right deltoid, but no sensory disturbance.

The patient underwent a C2-C6 total laminectomy and posterior midline myelotomy from the posterior median fissure of the spinal cord. The intraoperative histological diagnosis was glioma.

Pathological findings in low magnification demonstrated clusters of small uniform nuclei embedded in a dense and fibrillary matrix in hematoxylin-eosin staining (H.E.). On immunohistochemical staining, the tumor cells were weakly positive for glial fibrillary acidic protein (GFAP), but negative for the epithelial membrane antigen (EMA). The histopathological findings were consistent with the diagnosis of a subependymoma. However, the MIB-1 labeling index was of moderately high level up to approximately 8%.

In this case, we performed total resection because the tumor had rapidly increased in size and was of atypical form in histological findings.

It should be minded that some of subependymomas have a possibility of rapidly increasing in size with progressing neurological deficits.

Keywords:

MIB-1 labeling index, subependymoma, intramedullary spinal tumor

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Introduction

Subependymoma is a rare benign tumor that can occur throughout the central nervous system. In the spinal region, the most frequent site is the cervical intramedullary region (22.9%)^{1,2,4-16)}. Histologically, subependymoma corresponds to the World Health Organization Grade I³⁾.

Usually, mitotic activity is absent or low, and the index of Ki67/MIB-1 from immunohistochemical studies is under 1%¹⁷⁾.

We report a case of cervical subependymoma that rapidly increased in size in a year and a half, with progressing neurological deficits, and which had atypical pathological features.

The patient was informed that the data concerning the case would be submitted for publication and patient consent was provided.

Case Report

A 60-year-old woman presented with a six-month history of headache and numbness radiating to the right arm. MRI revealed a fusiform intramedullary mass at the hospital where she first presented. However, it was not specifically treated. As her right upper limb weakness had presented gradually, she visited our hospital after one and a half years. Neurological examination revealed muscle weakness in the right deltoid.

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Figure 1. (a) First MRI. (b) Second MRI on T2-weighted imaging.

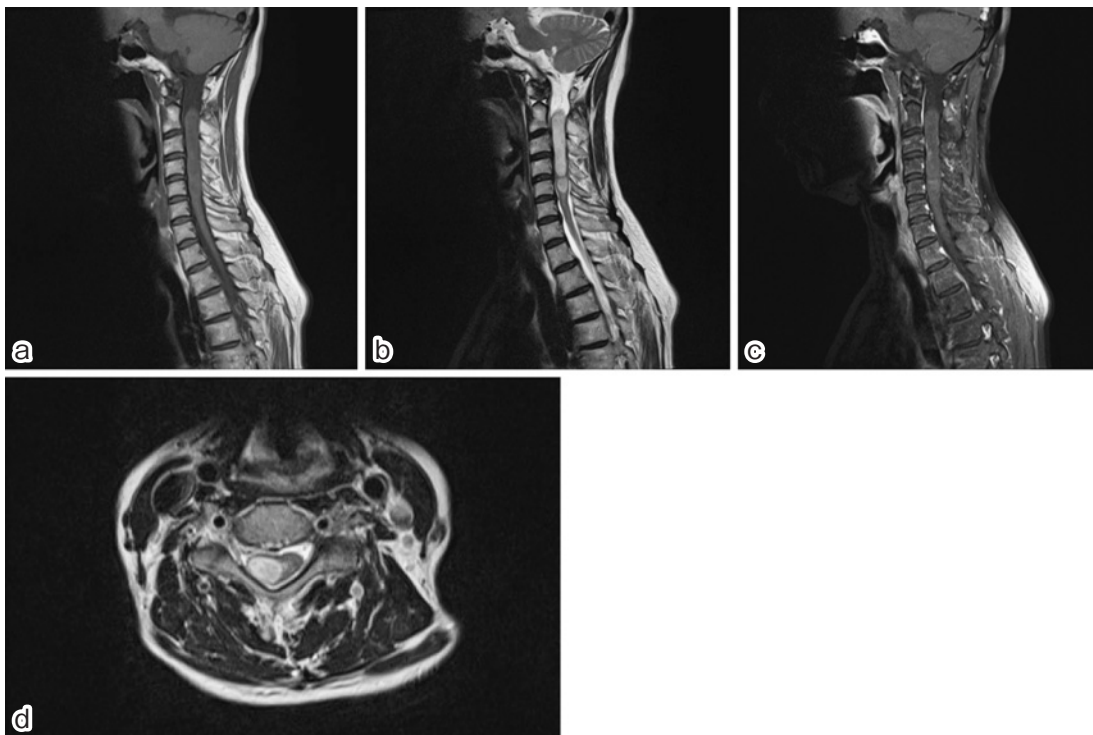


Figure 2. MRI (a) T1-weighted imaging. (b) T2-weighted imaging. (c) Enhancement imaging. (d) Axial view on T2-weighted imaging.

The tumor mass had progressed to the rostro-caudal side (Fig. 1). The two tumors showed low signal intensity on a T1-weighted MRI and high signal intensity on a T2-weighted MRI. Heterogeneous enhancement was observed with gadolinium administration. The axial MRI showed that the mass extended along the right antero-lateral surface of the spinal cord (Fig. 2).

The patient underwent a C2-C6 total laminectomy and

posterior midline myelotomy (Fig. 3). The tumor was located laterally to right side, and right dorsal root entry zone was swelling and curved, indicating difficulty of safe approach from around the right dorsal root entry zone. Therefore, we selected a midline approach to the tumor in order to minimize injury of the dorsal cord. The tumor was totally resected in microscopic view. However, a small amount of residual tumor was detected in the postoperative MRI, indi-

cating nearly total resection by the initial operation.

The intraoperative histological diagnosis was glioma. Pathological findings of the tumor tissue in low magnification demonstrated the distinctive clusters of small uniform nuclei embedded in a dense eosinophilic fibrillary matrix of glial cell processes with frequent occurrence of microcysts (Fig. 4a). Tumor cell nuclei appeared isomorphic and some tumor cell nuclei were surrounded by halos, and some tumor nuclei arranged around the micro lumen-like ependymoma (Fig. 4b). However, this tumor lacked cellularity, spindle cells, abundant eosinophilic cytoplasm, and perivas-

cular pseudorosette. The tumor demonstrated immunoreactivity for GFAP (glial fibrillary acidic protein) in fibrillary matrix (Fig. 4c), negative for epithelial membrane antigen (EMA), and 8% of nuclei of the tumor cells were positive for Ki-67 labeling (Fig. 4d). The characteristic microcystic change, clustered isomorphic nuclei, and dense fibrillarity produced by glial processes readily distinguish subependymoma from other gliomas.

Postoperatively, the patient developed a right upper and lower limb monoparesis and dysesthesia. At six-month follow-up, the motor function subsequently improved. MRIs showed no significant changes in the small remnant tumor.

Discussion

Scheinker first described this type of tumor in 1945¹⁸. Boykin proposed that it originated from subependymal astrocytes and named it subependymal glomerate astrocytoma¹⁵. To our knowledge, 70 cases have been presented as spinal subependymomas in previous papers^{1,2,4-16}. There were 16 cases of intramedullary cervical cord subependymomas; however, there were no reports regarding the process of tumor progression. Our case was able to follow the tumor growing for one and a half years, incidentally, and found rapid progression.

Regarding MRI findings on subependymomas, the usual finding on MRI was of no or scarce contrast enhancement, but this case showed heterogeneous enhancement¹².

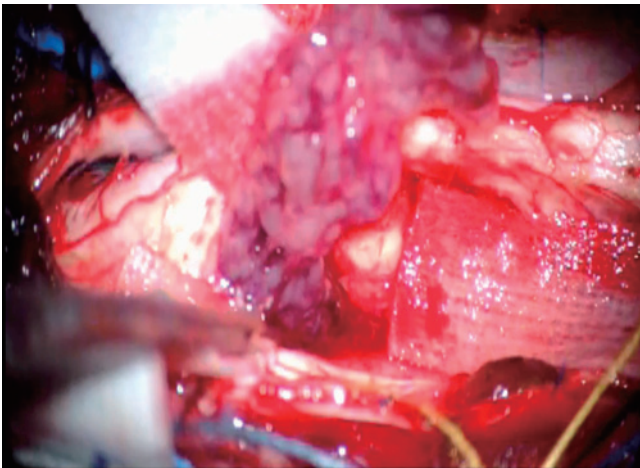


Figure 3. Tumor findings during the operation.

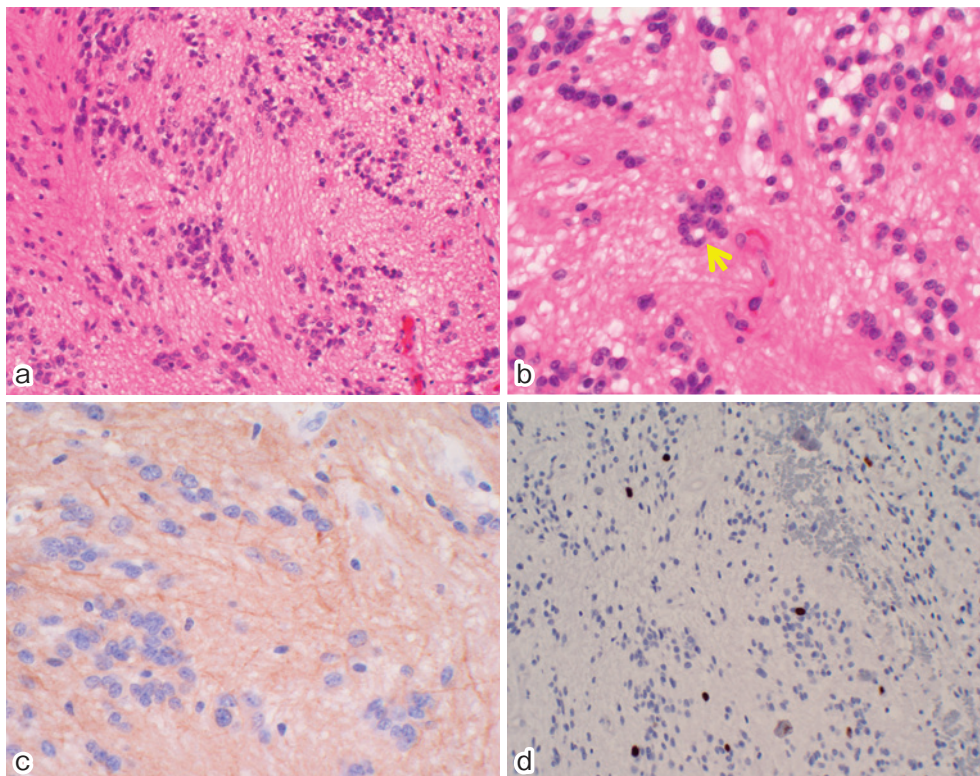


Figure 4. Histological findings. (a) H&E staining $\times 100$. (b) H&E staining $\times 400$; yellow arrow indicates micro lumen consisted by the nucleus of the tumor. (c) GFAP staining $\times 400$. (d) MIB-1 staining $\times 100$.

Mitoses are very rare or absent, with Ki-67 (i.e., MIB-1)-staining studies revealing labeling indices less than 1%. However, the MIB-1 labeling index was moderate high level up to approximately 8% in this case. Paryson mentioned that subependymomas accompanied by clinical symptoms have slightly higher Ki-67 indices than tumors discovered at autopsy, suggesting that even a slight increase in proliferative rate might affect clinical behavior¹⁷.

Regarding treatment, the efficacy of radiotherapy remains controversial^{21,24,25}.

Typical subependymomas have a good prognosis with surgical resection^{13,20,21}. Wu et al. mention that surgical treatment demonstrated that aggressive surgery for this tumor may cause either worsening of an existing deficit or the development of new deficits¹.

In this case, we performed nearly total resection because the tumor had rapidly increased in size and was of atypical form in histological findings.

It should be minded that some subependymomas have the possibility of rapidly increasing in size with progressing neurological deficits.

Conflicts of Interest: The authors declare that there are no relevant conflicts of interest.

Author Contributions: Hirosuke Nishimura wrote and prepared the manuscript, and all of the authors participated in the study design. All authors have read, reviewed, and approved the article.

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