

Two Cases of Spinal Tanycytic Ependymoma Associated with Neurofibromatosis Type 2

Xiao-Gang Tao, Zong-Gang Hou, Shu-Yu Hao, Jun-Ting Zhang, Bai-Yun Liu

Department of Neurosurgery, Beijing Tiantan Hospital, Capital Medical University, Beijing 100050, China

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INTRODUCTION

Tanycytic ependymoma is a rare subtype of the World Health Organization Grade II ependymoma most commonly found in the cervical and thoracic segments of the spinal cord. Spinal tumors in neurofibromatosis type 2 (NF-2) can be intramedullary, extramedullary, or associated with the nerve roots, and there may be multiple tumors at multiple locations in the spine. The histology of these tumors includes meningiomas, schwannomas, ependymomas, and neurofibroma.^[1] However, spinal tanycytic ependymomas associated with NF-2 are extremely rare, with only four cases reported in the English literature.^[1-4] In this study, we report two rare cases of spinal tanycytic ependymoma associated with NF-2 and provide a review of the literature.

CASE 1

A 17-year-old female patient was found to have NF-2 a year ago when magnetic resonance imaging (MRI) demonstrated bilateral acoustic neuromas. She underwent gamma knife radiosurgery for the right acoustic tumor. Neurological examination disclosed a loss of touch perception and a decrease of pain and cold sensations on the right side below the T2 dermatome. T1- and T2-weighted MRI images revealed a solid-cystic lesion of the cervical and thoracic spinal cord, which spread from the level of the C1–T3 vertebrae with areas of isointense signals and moderate enhancements [Figure 1a-1d]. A gross total resection of the lesion was performed using a posterior midline approach. The postoperative course was uneventful. In the 2-week and 3-month follow-up MRIs, there was no evidence of tumor recurrence [Figure 1e-1h]. Histopathologically, the tumor showed spindle cells with oval and elongated nuclei with occasional hemosiderin deposits present. In the immunohistochemical study, tumor cells were positive for the

glial fibrillary acidic protein (GFAP). The Ki-67 labeling index was 1%, indicating low proliferative activity. Ultrastructurally, the tumor showed spindle cells arranged in bundles with a scant extracellular matrix. Intercellular junctions, numerous slender surface microvilli, and microvilli-lined lumina were observed [Figure 1i-1j]. A follow-up at 8 months postsurgery did not reveal any recurrence of the lesion.

CASE 2

Genetic studies confirmed the presence of a mutation in the *NF-2* gene in a 43-year-old woman previously diagnosed with NF-2. The patient had a previous history of multiple tumors of the nervous system as a result of NF-2. Two months ago, the patient presented with decreased muscle strength of the right foot and occasional urinary and fecal incontinence. An MRI was performed, which showed that the T11-12 lesion had grown approximately 3.0 cm. Our main diagnosis in the differential for this lesion was a recurrent schwannoma. A gross total resection of the lesion was performed using a posterior midline approach. After surgery, the patient recovered and was asymptomatic at a 4-year follow-up. The pathologic examination of the tumor revealed elongated tumor cells with fibrillar processes arranged in a fascicular pattern. Inconspicuous perivascular pseudorosettes were noted. The Ki-67 labeling index was 1.5%.

DISCUSSION

Ependymoma is the most common spinal tumor in patients

Address for correspondence: Dr. Bai-Yun Liu,
Department of Neurosurgery, Beijing Tiantan Hospital,
Capital Medical University, Beijing 100050, China
E-Mail: liubaiyun1963@163.com

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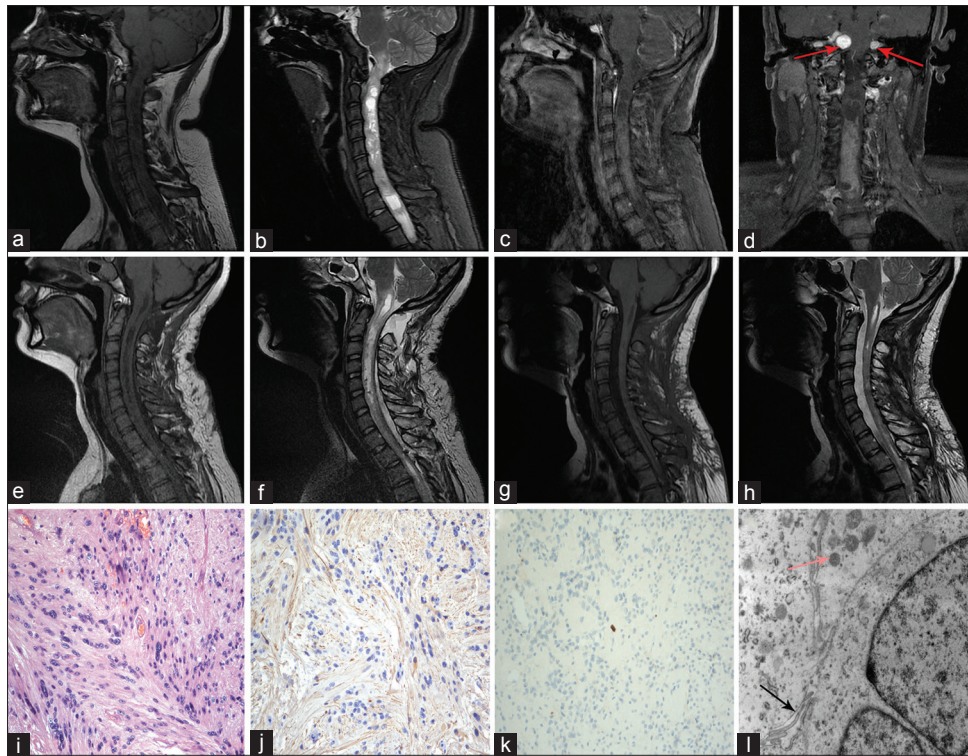


Figure 1: T1- and T2-weighted MRI images of the spinal cord show an isointense tumor in case 1 (a and b). Contrast-enhanced sagittal and coronal images show a slightly hyperintense tumor with bilateral acoustic neuromas (red arrow) (c and d). Sagittal T1- and T2-weighted images of the cervical spine after C1–T3 laminectomies and tumor resection demonstrating GTR with no residual mass in the 2-week and 3-month follow-up MRIs (e-h). Red arrows indicate bilateral acoustic neuromas. Histological sections showing spindle cell neoplasm of moderate cellularity arranged in interlacing fascicles (i). The tumor cells are diffusely and strongly positive for GFAP (j). The Ki-67 labeling index was low (1%) (k). Electron micrograph of the tumor showing intercellular junctions (black arrow) and numerous slender surface microvilli (pink arrow) (l). Original magnification in (i-k) is $\times 200$, (l) is $\times 10,000$. GTR: Gross total resection; MRI: Magnetic resonance imaging; GFAP: Glial fibrillary acidic protein.

with NF-2. However, reports of spinal tancytic ependymoma associated with NF-2 are extraordinarily rare. Using the search terms “tancytic ependymoma” and “spine”, we identified 27 reports of 34 patients with histologically proven diagnoses of tancytic ependymoma. Patient ages ranged from 10 to 76 years, with a mean of 40 years and a female-to-male ratio of 1.25:1. The most common location was the cervical spine (15/34), followed by the thoracic spine (14/34), and cauda equina (5/34). The diagnosis of tancytic ependymoma is based mainly on histological features of tancytic tumor cells and immunohistochemical features. The differential diagnosis of tancytic ependymoma includes schwannoma and pilocytic astrocytoma, which makes the diagnosis more challenging and difficult. Histologically, the characteristic “salt and pepper speckled” appearance of nuclei in tancytic ependymoma distinguishes it from astrocytoma. Furthermore, tancytic ependymoma normally shows a strong immunoreactivity for GFAP, whereas schwannomas only focally express GFAP.^[1] Electron microscopy can aid in distinguishing tancytic ependymoma from other tumors if the diagnosis is in doubt. The primary therapeutic option for spinal tancytic ependymoma is complete resection with no microscopic residual tumor. When complete resection is performed, postoperative radiotherapy is not necessary. We continue to observe this patient carefully and consider reoperation or adjuvant radiotherapy if recurrence occurs.

The Ki-67 labeling index of tancytic ependymoma was lower than other subtypes of ependymoma, which indicated that tancytic ependymoma has a better prognosis. However, NF-2 is associated with the development of various tumors in different areas of the body. Therefore, routine periodic surveillance with MRI and long-term follow-up of patients suffering from spinal tancytic ependymoma associated with NF-2 is mandatory after treatment.

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Conflicts of interest

There are no conflicts of interest.

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