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CASE REPORT

Tamio Ito · Yoshimaru Ozaki · Jyoji Nakagawara
Hirohiko Nakamura · Shinya Tanaka · Kazuo Nagashima

A case of cervicomedullary junction tanycytic ependymoma associated with marked cyst formation

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Abstract Tanycytic ependymomas are a subtype of ependymomas that were formally recognized as a new pathological entity in the latest World Health Organization (WHO) classification of 2000. They occur mostly in the spinal cord. Only a few reports have analyzed the proliferative potentials of these tumors; however, it has been reported that the MIB-1 labeling index of tanycytic ependymoma is lower than that of other subtypes of WHO grade II ependymomas. We report a rare case of cervicomedullary junction tanycytic ependymoma associated with marked cyst formation. A 62-year-old man had a history of progressive gait disturbance, diplopia, and swallowing disturbance over a one-month period prior to admission. Magnetic resonance imaging (MRI) showed a cystic mass with a mural nodule at the cervicomedullary junction with Gd-DTPA enhancement. Cyst-subarachnoid shunt was performed using a far lateral approach. After 6 years, however, the man was readmitted to the hospital because of reaccumulation of the cyst. Partial removal of a mural nodule and a cyst-subarachnoid shunt were performed simultaneously by a midline suboccipital approach. The pathological diagnosis was tanycytic ependymoma. Postoperatively, the patient recovered well and was discharged from the hospital without further treatment. Most of the tumor cells had small, round nuclei; pleomorphism was minimal. The cytoplasm was dilated. The tumor cells were positive for EMA and s-100, and negative for CD-34. GFAP was not determined due to difficulty caused by background glial processes. The MIB-1 labeling index was less than 1%. Ultrastructurally, the tumor cells had ependymal cell features, such as desmosomes and microvilli. Based on these findings, the pathological diagnosis was tanycytic ependymoma.

T. Ito (✉) · Y. Ozaki · J. Nakagawara · H. Nakamura
Department of Neurosurgery, Nakamura Memorial Hospital,
Minami-1 jo Nishi-14 chome, Chuo-ku, Sapporo 060-8570, Japan
Tel. +81-11-231-8555 (ext. 311); Fax +81-11-231-8387
e-mail: titoh@med.nmh.or.jp

S. Tanaka · K. Nagashima
Department of Laboratory of Molecular and Cellular Pathology,
Hokkaido University Graduate School of Medicine, Sapporo, Japan

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Introduction

Tanycytic ependymomas are a subtype of ependymomas that were formally recognized as a new pathological entity in the latest World Health Organization (WHO) classification of 2000.[1] These tumor cells have morphological characteristics resembling tanycytes. Therefore following the first report in 1978, by Friede and Pollak, these tumors were referred to as a tanycytic variant of ependymoma.[2] There are only a few reports that have examined the proliferative potentials of these tumors; however, it has been reported that the MIB-1 labeling index of tanycytic ependymoma is lower than that of other subtypes of WHO grade II ependymomas.[1] While the tumors occur mostly in the spinal cord, we report a rare case of cervicomedullary junction tanycytic ependymoma associated with marked cyst formation, which recurred several times.

Clinical summary

A 62-year-old man was admitted to Nakamura Memorial Hospital on March 24, 1997, with a history of progressive gait disturbance, diplopia, and swallowing disturbance of one month's duration. MRI demonstrated a cystic mass with a mural nodule at the cervicomedullary junction with Gd-DTPA enhancement (Fig. 1A,B). For the purpose of communication between the cyst and subarachnoid space, a cyst-subarachnoid shunt was performed using a far lateral approach on May 7, 1997. The postoperative period was uneventful and the man was discharged from the hospital on July 22, 1997. Because magnetic resonance imaging (MRI) on November 7, 2002, showed reaccumulation of the cyst and the patient had developed gait disturbance and diplopia, partial removal of the tumor and opening of the

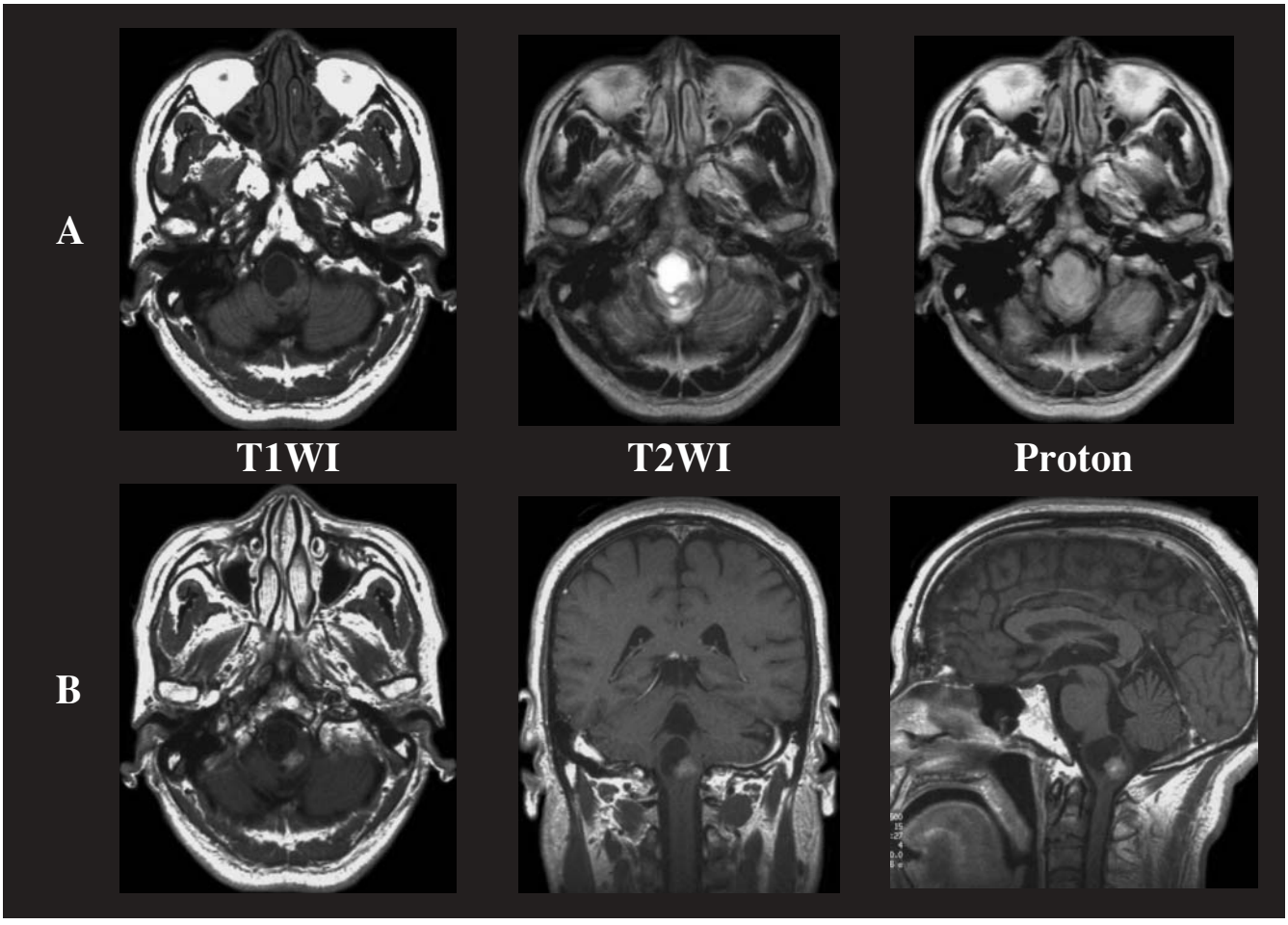

Fig. 1. Pre-first operative MRI (**A** Plane, **B** Gd-GTPA enhancement), showing a cystic tumor with a mural nodule in the cervicomedullary junction

cyst were performed using a midline suboccipital approach on February 1, 2003. Postoperatively, the patient recovered well due to shrinkage of the cyst, but a pathological diagnosis of the tumor was not obtained because the size of the removed tumor was too small. However, the man was readmitted to the hospital on July 1, 2003, with a history of progressive gait disturbance, diplopia, and swallowing disturbance. MRI demonstrated regrowth of both the cyst and the mural nodule with Gd-DTPA enhancement (Fig. 2A). Partial removal of the tumor and a cyst-subarachnoid shunt were performed simultaneously using a midline suboccipital approach. Postoperative Gd-MRI showed shrinkage of both the cyst and the small residual tumor (Fig. 2B). The postoperative period was uneventful. The pathological diagnosis was tanycytic ependymoma and the MIB-1 index was lower than 1 %. The patient was therefore discharged from the hospital without further treatment and is now recurrence-free one year after the third operation.

Pathological findings

Most of the tumor cells had small, round nuclei, and pleomorphism was minimal. The cytoplasm was dilated (Fig. 3A,B). The tumor cells were positive for EMA and s-100 (Fig. 4B,C), and negative for CD-34 (Fig. 4D). GFAP was not determined due to difficulty caused by the background glial processes (Fig. 4A). The MIB-1 labeling index was less than 1% (Fig. 4E). Ultrastructurally, the tumor cells had ependymal cell features such as desmosomes and microvilli (Fig. 5A,B). Based on these findings, the pathological diagnosis was tanycytic ependymoma.

Discussion

Tanycytic ependymomas are a subtype of ependymomas that were formally recognized as a new pathological entity in the latest WHO classification of 2000.[1] However, this name has been used for a long time, because the tumor cells have morphological characteristics resembling tanycytes. In

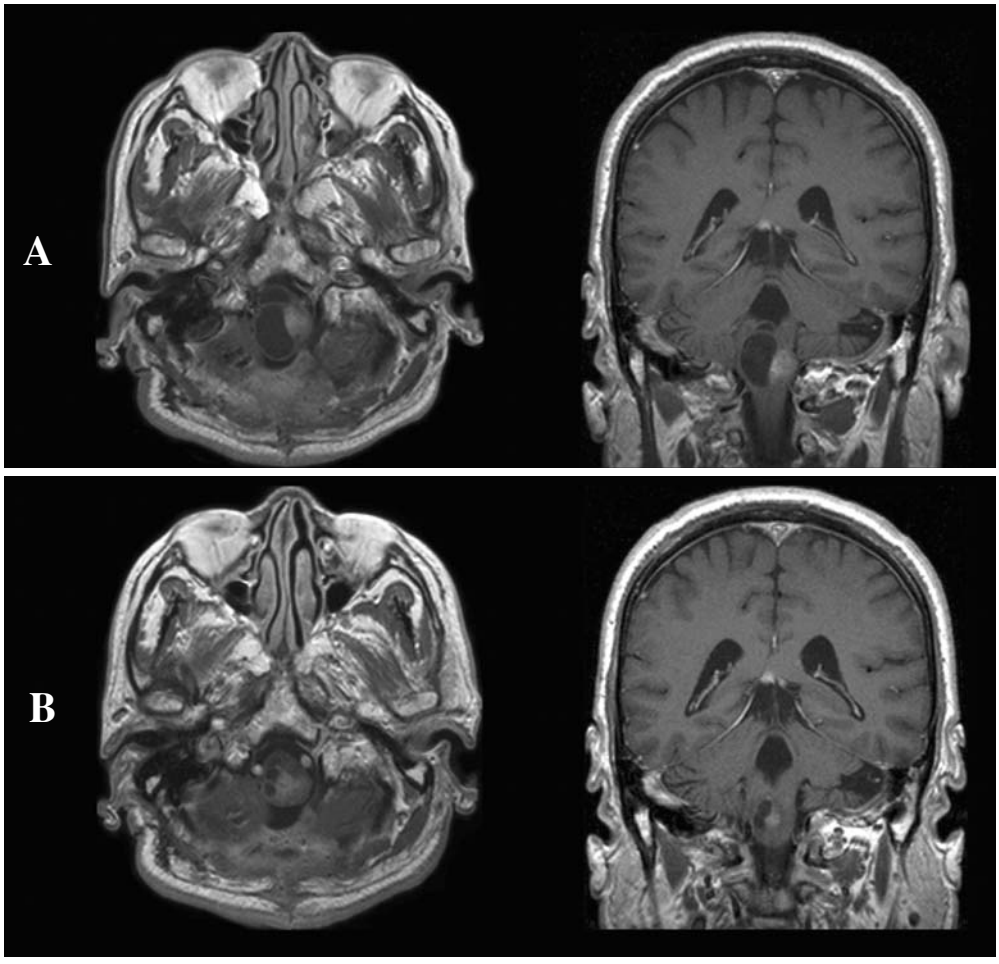

Fig. 2. **A** Pre-third operative Gd-MRI, showing reaccumulation of the cyst and a slowly growing mural nodule. **B** Post-third operative Gd-MRI, showing shrinkage of the cyst and partial removal of the tumor

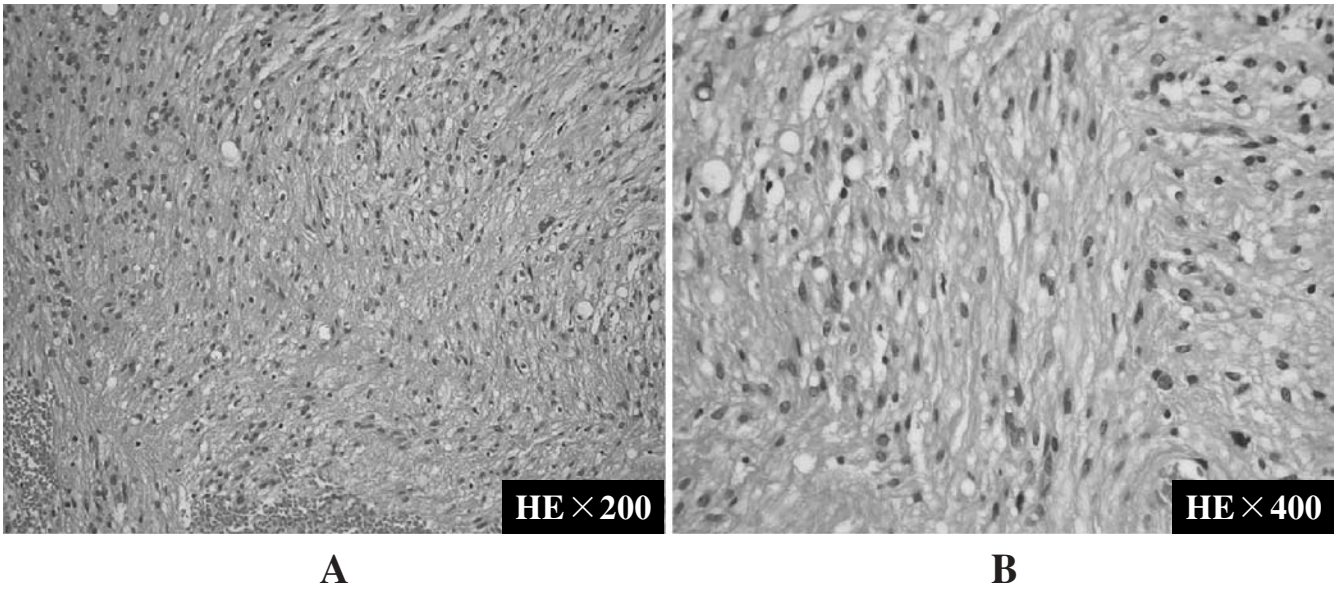

Fig. 3. Histological examination of the tumor, showing elongated cells with small, round nuclei and dilated cytoplasm. H&E stain, **A** × 200, **B** × 400

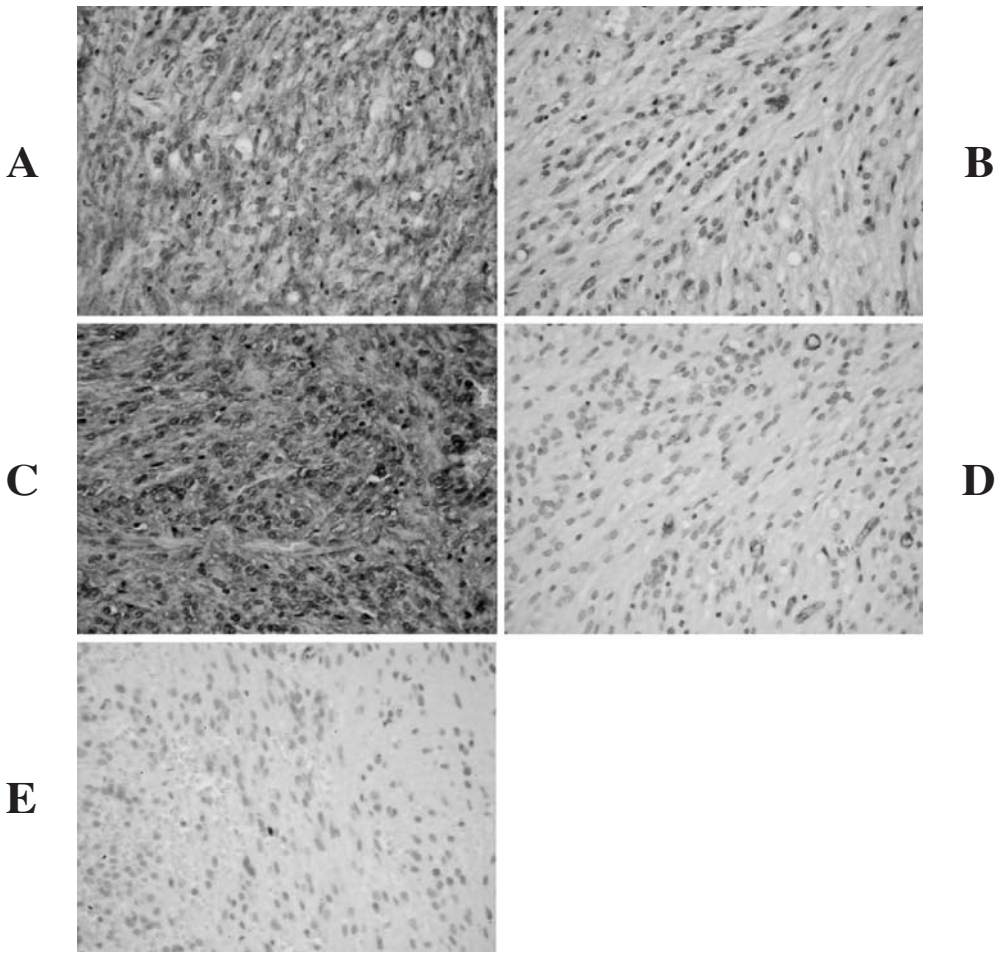

Fig. 4A–E. Immunohistochemical examination of the tumor. GFAP was not determined due to difficulty caused by background glial processes (**A**). EMA and s-100 were positive (**B, C**), and CD-34 was negative (**D**). The MIB-1 labeling index was lower than 1% (**E**)

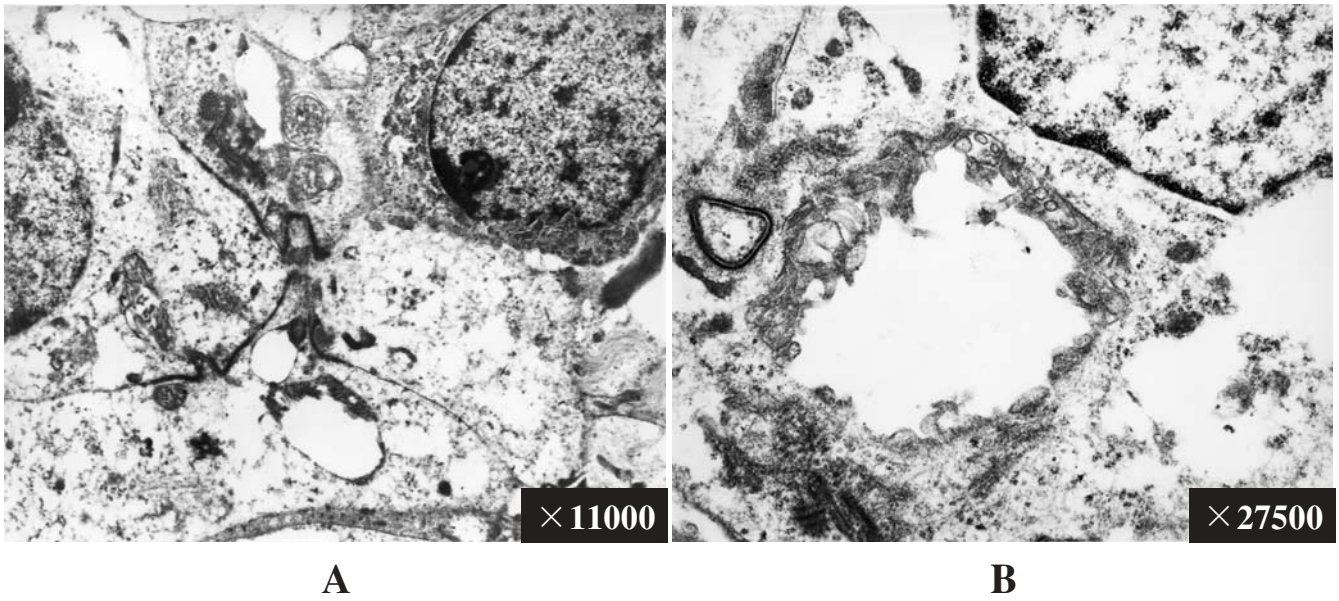

Fig. 5. Electron micrographs of the tumor, showing ependymal cell features, such as desmosomes and microvilli (**A** × 11 000, **B** × 27 500)

Table 1. Reported cases of tancytic ependymoma

Author		Age/sex	Location	Gd	Cyst
1978	Friede/Pollak	38/F	C6-T3		
		46/F	T7-L2		
		45/M	C1-T1		
		17/M	C1-3		
		36/F	C1-6		
		35/F	T10-11		
1986	Spaar	32/M	Cervical		
1997	Langford/Barre	52/F	T6-7		
1999	Daneyemez	42/M	lateral v.		+
2001	Ueki	18/F	C7-T2	+	+ (syr)
2001	Kawano	45/M	T3-4	+	–
		36/F	C7-T2	+	+
		55/F	C3-6	+	+
2004	Present case	62/M	CMJ	+	+

v, ventricle; syr, syrinx; CMJ, cervicomedullary junction

1978, Friede and Pollak referred to these tumors as a tanycytic variant of ependymomas.[2] It is thought that tanycytes are a special subtype of ependymal cells that are found in the ventral region of the third ventricle. As for their function, it is thought that tanycytes have neuroendocrine functions, mediating between the cerebrospinal fluid and hypothalamic neuronal cells.[3]

These elongated cells with rich fibrillary processes resemble pilocytic astrocytomas, schwannomas, subependymomas, and fibrous meningiomas.[4–6] As for immunochemical staining, tanycytic ependymomas are positive for GFAP, s-100, and vimentin, and generally negative for EMA. However, some tumor cells may be positive for EMA in a scatterd and granular fashion. In our case, GFAP was not determined due to difficulty caused by the background glial processes, but EMA and s-100 were negative. For the diagnosis of tanycytic ependymoma, it is most important to note that the tumor cells have epithelial features, as seen on electron microscopy. Ultrastructurally, the tumor cells have ependymal cell features, such as desmosomes and microvilli, as was the case in our patient. There are only a few reports that have examined the proliferative potentials of these tumors. Kawano et al. reported MIB-1 indexes of 0.5%, 1.1%, and 2.3% in their three spinal tanycytic ependymoma cases.[5] In Suzuki's report, the MIB-1 index of the clear-cell types was higher and that of tanycytic types was lower in the 29 grade 2 ependymoma cases.[7] The MIB-1 index of the tumor in our patient was also less than 1%, so it seems that the MIB-1 index of tanycytic ependymomas is generally low, but further examination will be necessary.

As for tumor locations, Kawano stated that more than half of the tanycytic ependymomas (11/19) occurred in the spinal cord, based on a review of reported cases.[5] Of these 11 cases, six occurred in the cervical region and five in the thoracic region.[4–6,8,9] There was a rare case in the lateral ventricle;[4] however, it is also rare that the tumor occurred in the medulla in our patient.

There are only a few reports of cyst formation, but four of five tanycytic ependymomas (4 spinal cord, 1 lateral ventricle) had cyst formations (Table 1),[4–6] and there was a mural nodule in one of Kawano's cervical cases.[8] There was also a mural nodule in our case, so it may be that tanycytic ependymomas tend to cyst formation.

Regarding the surgical treatment of tanycytic ependymomas, it is important that they are removed totally, because these tumors frequently recur by cyst formation.

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