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Case Report

Tanycytic ependymoma arising from the right lateral ventricle: A case report and review of the literature

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A 38-year-old man presented with a year-long history of worsening headache. Neuroradiological findings showed that a solid cystic mass occupied the right lateral ventricle. Histologically, the tumor composed of nuclear dense zones consisting of a cluster of spindle cells and fibrillary zones consisting of streaming of cell processes. The tumor cells showed the characteristics of monopolar or bipolar processes. Some tumor cell processes extended to the vessel wall and formed ill-defined perivascular rosettes. No mitoses or necrosis were found. The cells presented positive for GFAP, S-100 protein, vimentin, Nestin and neurofilament, and dotlike positive for epithelial membrane antigen, but negative for Syn and NeuN. Four cases of tanycytic ependymoma arising from the lateral ventricle have been reported in literature. Histological differential diagnosis includes spindle-shaped neuroepithelial tumors, such as pilocytic astrocytoma, fibrillary astrocytoma and schwannoma. Tanycytic ependymoma has slightly better prognosis than other ependymoma subtypes.

Key words: brain neoplasms, ependymoma, immunohistochemistry, tanycyte.

INTRODUCTION

Tanycytic ependymoma (TE) is a rare subtype of ependymoma deriving from tanycyte.[1–3] The tumor is composed of clusters of elongated cells forming nuclear dense zones and streaming cell processes forming fibrillary zones. Typical ependymal rosettes are absent, and perivascular rosettes are vague. The pathological diagnosis of TE is confused with other neuroepithelial tumors consisting of spindle cells. TEs show a predilection for the spinal cord, a few for the supratentorial region. This paper reports a case of TE arising from the right lateral ventricle and reviews relevant literatures.

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CASE REPORT

Clinical history

A 38-year-old man was admitted to our neurosurgery department in October 2006 because of intermittent headache for 1 year which had worsened with nausea for the past month. The patient had fitful headache without obvious causes over the previous year, of about a half-hour duration every time. The neurological examination showed no motor or sensory weakness. There were no abnormalities noted in the general physical examination, and no history of hypertension and diabetes, and no familial inherited disease.

Neuroradiological findings

A brain MRI showed a $3.5 \times 2.3 \times 2.4$ cm well-defined solid-cystic (mainly solid) mass in the anterior horn of the right lateral ventricle. The mass showed low intensity on T1-weighted images and high intensity on T2-weighted images (Fig. 1A,B). The tumor was enhanced irregularly by gadolinium-diethylene triaminopenta-acetic acid (Gd-DTPA) (Fig. 1C,D). A clinicoradiologic diagnosis of ependymoma was made.

Surgery

Under general anesthesia, after turning the right coronal bone flap and incising the frontal lobe, the tumor was seen as being attached to the right lateral ventricle. The tumor had a rich blood supply, and its size was $3.5 \times 2.3 \times 2.4$ cm.

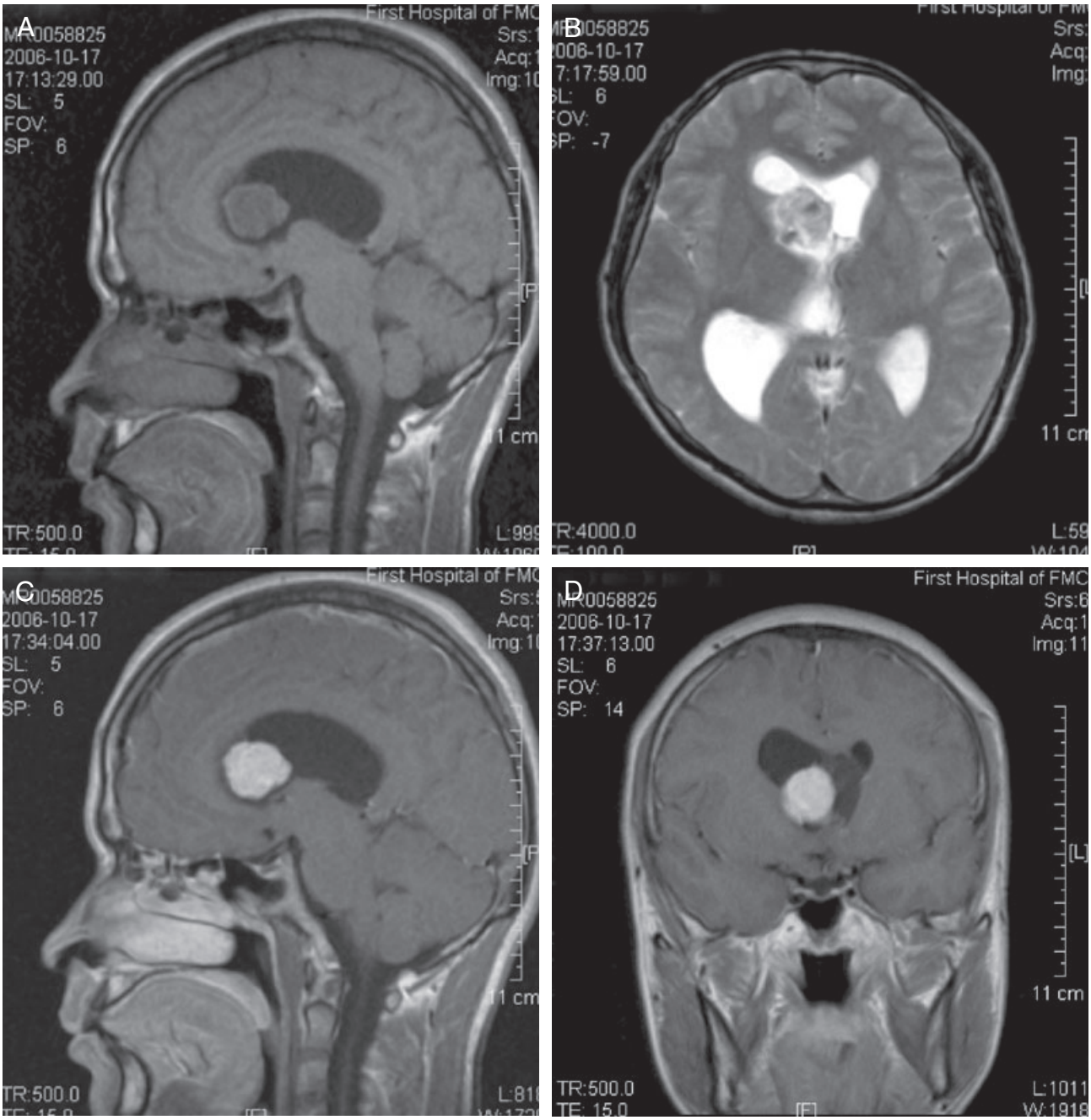

Fig. 1 MRI showing a 3.5 × 2.3 × 2.4-cm well-defined solid cystic (mainly solid) mass in the anterior horn of the right lateral ventricle. (A) Sagittal T1-weighted image shows the tumor located on the anterior horn of lateral ventricle. (B) Axial T2-weighted image shows heterogenous signal. (C) Sagittal T1-weighted image enhanced by Gd-DTPA. (D) Coronal T1-weighted image enhanced by Gd-DTPA.

It was disconnected along its boundary then dissected completely under neurosurgical microscope.

Pathological changes

Tumor fragments were 2 × 3 × 1 cm. A cross-section of the tumor presented as fresh fishmeat-like, gray and fragile. A few areas showed calcification.

Microscopically, the tumor had nuclear dense zones consisting of a cluster of spindle cells and fibrillary zones consisting of streaming of cell processes (Fig. 2A). The tumor cells were monopolar or bipolar long spindles and resembled tanycytes. Some tumor cell processes extended to the vessel wall and formed ill-defined perivascular rosettes. No classic perivascular rosettes and true ependy-

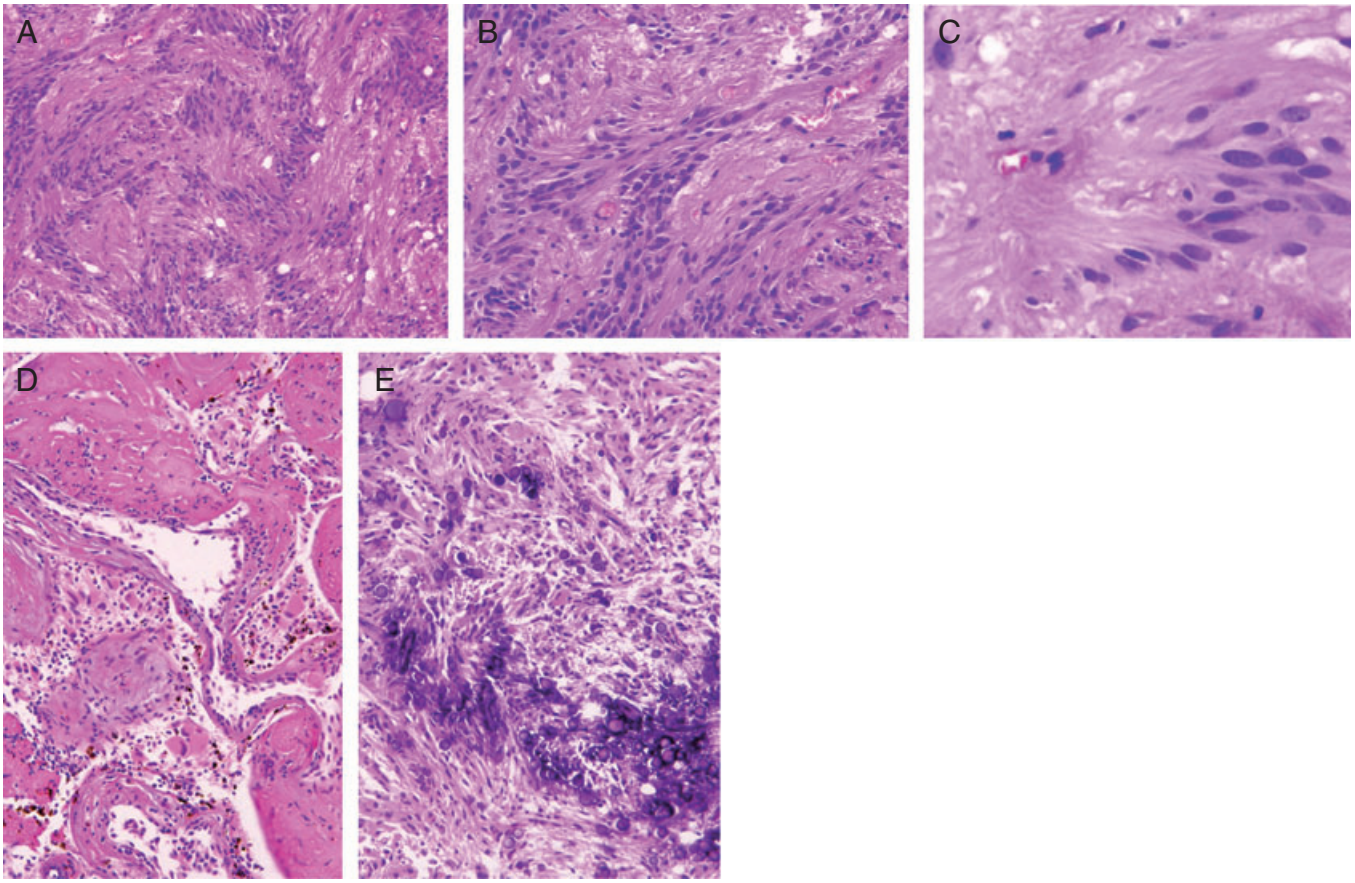

Fig. 2 Histological features, HE-stained. The tumors consisted of monotonous proliferation of long spindle monopolar or bipolar cells. The tumor has nuclear dense zones and fibrillary zones (A × 100). Focally, these spindle tumor cells showed a vague arrangement around blood vessels (B × 200). Most of the tumor cell nuclei had smooth contours and contained fine chromatin. Nuclear pleomorphism was minimal and exhibited the salt-and-pepper speckling of other ependymomas (C × 400). Hyalined vessel walls (D × 100) and calcospherites (E × 100) were seen in several areas.

mal rosettes were found (Fig. 2B). The nuclei showed round-to-oval with slight pleomorphism and exhibited the salt-and-pepper speckling of other ependymomas (Fig. 2C). No mitoses or necrosis were appreciated. Part of the tumor presented degenerative changes including hyalined vessel walls and calcospherites.

Immunohistochemical staining showed that GFAP and neurofilament (NF) were positive, S-100 protein, vimentin and Nestin were strongly positive, EMA was dotlike positive (Fig. 3), but Syn and NeuN were negative. The MIB-1 labeling index was <0.5%.

Postoperative treatment and follow-up

The patient was discharged from the hospital on postoperative day 5, neurologically intact. He was further treated with a course of radiotherapy, from DT40Gy/20f to DT54Gy/27f. The patient has remained stable for 8 months without recurrence.

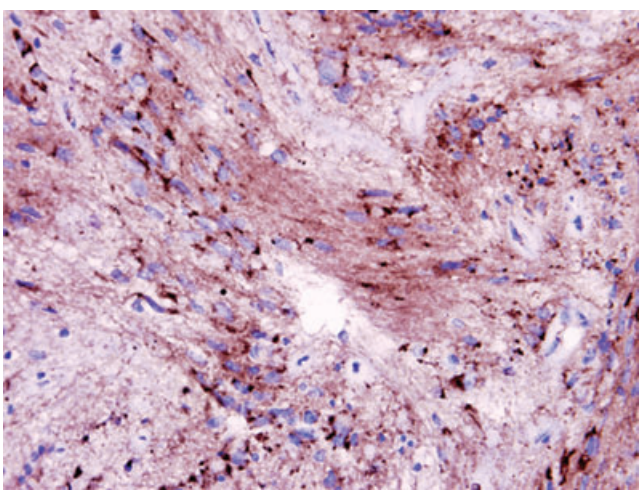

Fig. 3 Immunostaining for epithelial membrane antigen exhibited a dotlike intracytoplasmic positivity.

Table 1 Location and sex distribution of tanycytic ependymomas

	Male (%)	Female (%)	Total (%)
Supratentorial			
Extraventricle	4 (13.8%)	0 (0)	4 (13.8%)
Intraventricle	4 (13.8%)	2 (6.9%)	6 (20.7%)
Spinal cord	8 (27.6%)	11 (37.9%)	19 (65.5%)
Total	16 (55.2%)	13 (44.8%)	29 (100%)

DISCUSSION

Twenty-nine TEs have been reported.[4–19] Patient ages at diagnosis ranged from 6 to 62 years (mean, 36.0; median, 39 years). TEs occur frequently in spinal cord (65.5%, 19/29) followed by intraventricle (20.7%, 6/29) and supratentorial extraventricle (13.8%, 4/29) (Table 1). No obvious gender difference was found, male patients being slightly more than female and M : F ratio being 1.2 : 1.0. Interestingly, all four patients reported suffering from supratentorial extraventricular mass were male.[3–6] Including the present case, four cases of TEs have arisen from the lateral ventricle (Table 2).[7–9] Neuroradiologically, they are cystic or solid-cystic masses involving occasional brain tissue in the vicinity of or in the third ventricle.

Origin and genetics of TEs

The origin of ependymoma is a matter of debate. However, there seems to be an agreement that the ontogeny of ependymoma is radial glia,[7,20] which is the stem cell of tanycyte and glia. Tanycytes are interspersed among the columnar ependymal cells. Their morphologically distinguishing feature is that they have a long non-branching cytoplasmic process, which extends for a variable distance toward the surface of blood vessels, neurons, or the pia mater.[21] Friede *et al.*[3] coined the term "tanycytic ependymoma" in 1978 for the similarity to tanycytes and reported six cases. Recent studies on this rare tumor indicated that they originated from tanycytes. TE has been classified as a variant of ependymoma by the 2000 and 2007 WHO classification. Studies suggested that tumors with histomorphologic and ultrastructural characteristics similar to tanycyte also include pilocytic astrocytoma, myxopapillary ependymoma, astroblastoma,[22,23] subependymoma, and so on. These tumors have similar biological behavior and prognosis, so Lieberman *et al.*[24] suggested that they would be described as the same type, and a new diagnostic term "tanycytoma" was proposed.

Genetic features of TE remain unclear. Two cases reported were in relation with neurofibromatosis (NF) type 2, which indicated NF2 gene may be involved in the tumorigenesis.[10,11] One case has a family history of ependymoma.[12]

Histopathology

Generally, TE is a well-demarcated mass. Microscopically, the tumor has nuclear dense zones consisting of a cluster of spindle cells and fibrillary zones consisting of streaming of cell processes. It generally has no ependymal rosettes and ill-defined perivascular pseudorosettes, which Kawano *et al.*[13] defined as a "pure type." Another type was "mixed type" that contains both true rosettes and perivascular pseudorosettes. Monopolar or bipolar long spindle tumor cells are seen as streams of piloid or hair-like, and resemble tanycytes. The nuclei showed round-to-oval with slight pleomorphism and exhibited the salt-and-pepper speckling of other ependymomas. Mitoses and necrosis were not appreciated. Part of the tumor presented degenerative changes including hyalined vessel walls and calcospherites.

Immunohistochemical staining generally showed reactive for GFAP, S-100 protein and vimentin.[17] Nestin is reported to be mid-to-strongly positive. EMA staining exhibited diffuse distribution of dotlike positivity.[25] Syn and NeuN were negative immunoactivity. The MIB-1 labeling index was from 0.3% to 4.6%.[4,11,13,19]

Electron microscopy (EM) can aid in distinguishing TEs from other tumors if diagnosis is in doubt. EM of these tumors shows characteristic ependymal features including intracytoplasmic intermediate filaments, prominent intercellular junctions, numerous slender surface microvilli, and microvilli-lined lumina, but no basement membrane in aggregated tumor cells.[4,6,8,15]

Differential diagnosis

The diagnosis of TE is based mainly on histological features of tanycytic tumor cell and immunohistochemical features. The differential diagnosis of TE includes those spindle cell neuroepithelial tumors, such as pilocytic astrocytoma, fibrillary astrocytoma and schwannoma.[7,11,14,26] TE may be confused with pilocytic astrocytoma, but the latter shows a biphasic pattern, Rosenthal fibers and/or granular bodies. Fibrillary astrocytoma is identified by characteristic morphologic feature of microcysts and mucinous degeneration, which are not seen in TE. Schwannoma may raise a problem. However, TE is a compact mass of elongated cells without loose areas (Antoni B areas) and Verocay bodies as seen generally in schwannomas. TEs and astrocytomas often show immunopositivity for GFAP, vimentin, S-100 protein and Nestin, whereas schwannomas are negative for GFAP and vimentin, and pilocytic and fibrillary astrocytomas are negative for Nestin. Ultrastructural studies are recommended and are very useful in identifying TE if the differential diagnosis is in doubt.

Table 2 Tanycytic ependymomas of the lateral ventricle

Author/year	Age (years)/sex	Presentation	Radiographic features
Danyemez *et al.* (1999)[7]	42/male	Leg weakness, dysarthria, and headache for 6 months	3 × 3 × 2 cm cystic mass of right lateral ventricle
Hayashi£ *et al.* (2000)[8]	51/male	Long-standing epilepsy, imaged after a fall	Large cystic mass of right frontal lobe, arising from right ventricle
Ragel£ *et al.* (2005)[9]	55/female	Dizziness with worsening dysequilibrium	3 cm solid cystic mass of the left lateral ventricle and the third ventricle
Present	38/male	Relapse headache for 1 year	3.5 × 2.3 × 2.4 cm solid cystic mass of the right ventricle

Treatment and prognosis

An effective treatment plan is a complete resection followed by radiologic surveillance.[9] Repeating resection or radiation treatment will be recommended in the event of recurrence. Many studies have shown that ependymomas have a good prognosis. Supratentorial ependymomas also have a good prognosis despite recurrence.[27] Suzuki *et al.*[19] suggested that MIB-1 labeling index and p53 expression of TEs were lower than other subtypes of ependymomas, which indicated that TEs have a better prognosis than other ependymomas. Follow-up data demonstrated that 5- and 10-year survival rates in TE were obviously higher than other subtypes.

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