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Central neurocytoma: a clinical, radiological and pathological study of nine cases

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Abstract

Purpose—Central neurocytoma is a rare intraventricular brain tumor that affects young adults and presents with increased intracranial pressure secondary to obstructive hydrocephalus. Typically, it has a favorable prognosis after adequate surgical intervention, but in some cases the clinical course is more aggressive. In this report, we describe the diagnosis and treatment of central neurocytoma in a series of patients at our institution.

Patients and Methods—Our series of nine patients (M:F=2:7, mean age, 28.2 years) with ventricular tumors showed typical radiological, histologic and immunohistochemical features of central neurocytoma. Most patients received craniotomy with removal of the tumor through transcallosal or transcortical approach. The surgical and histopathologic data of these patients were reviewed and analyzed.

Results—The prognosis is generally favorable. Although most patients were alive and well at the last follow-up, two developed recurrence. Typical histologic features of recurrent neurocytoma include high proliferative activity (MIB-1 labeling index: 2.0–6.8%), prominent vascular proliferation and remarkable synaptophysin expression. Two patients (non-recurrent) died during follow-up due to sepsis or central failure. The MIB-1 labeling indices were as high as 2.2–5.4% for these two patients.

Conclusion—Although central neurocytoma is generally a benign neoplasm, some variant forms of recurrence are also present. Complete resection provides favorable long-term prognosis in most cases. Recurrent tumors are often local and the patients seem to recover well after a second resection followed by radiotherapy. Histologic features such as tumor proliferation (MIB-1 labeling index), vascular proliferation, and synaptophysin expression are often prominent in the recurrent tumor. We recommend that these histologic features be considered for tumor recurrence during treatment and follow-up of these patients.

Keywords

central neurocytoma; MIB-1 labeling index; proliferation; recurrence

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Introduction

In 1982, Hassoun et al. [1] reported two cases of central neurocytomas, defining these tumors as a distinct pathological entity. This report resulted in increased recognition of central neurocytomas and a number of case reports and series have been published since [1,2,3,36, 39]. Despite their increased recognition, central neurocytomas remain rare neoplasms of the central nervous system. Overall, they compromise only 0.25–0.5% of brain tumors [2]. The main features of central neurocytoma include: 1) a lateral ventricular location, 2) occurrence in young adults, 3) characteristic radiological findings, 4) resemblance to oligodendroglioma or ependymoma on light microscopy, 5) a neuronal origin seen on electron microscopic and immunohistochemical examination, and 6) a favorable prognosis with benign biological behavior [2,3]. The diagnosis and management of this tumor remains controversial. In this report, we describe the diagnosis and treatment of central neurocytoma in a series at our institution.

Patients and methods

Between January 1992 and May 2003, 9 patients (0.31% of all intracranial tumors at the same period) were diagnosed as central neurocytoma by clinical symptoms, radiological findings and histopathological examinations including light microscopy, immunohistochemical staining for neuron-specific enolase (NSE) and synaptophysin at Taichung Veterans General Hospital. To evaluate the proliferative potential of the tumors, the specimens were immunostained with MIB-1 antibody, which is specific for detecting Ki-67 antigen present in all proliferative cells. Local control and overall survival were analyzed using the Kaplan–Meier survival curve.

Results

Basic clinical data of the patients are as shown in Table 1. The male to female ratio was 2:7 and their ages ranged from 17 to 45 years (average: 28.2 years). Most of the patients developed symptoms before 30 years of age. Only three patients were older than 30 years. Symptoms including raised intracranial pressure (ICP) in all patients, general seizure, general weakness and blur vision were found. The duration of symptoms varied from one month to one year (average 4.7 months).

Radiographic Studies

The CT scans generally revealed that the tumors were round, relatively well circumscribed, and iso- or slightly hyperdense in the supratentorial intraventricular area. Cystic changes were present in five cases and calcifications were found in four. Images obtained using contrast materials showed relatively homogeneous enhancement in all cases. In the T1- and T2-weighted MR images, the tumor mass showed isodense to hyperdense signal intensity. All cases revealed moderate to strong enhancement of tumor with gadolinium–diethylenetriamine pentaacetic acid. The tumor was confined to the lateral ventricle in four cases and extended from lateral into the third ventricle through the foramen of Monro in another five cases. (Table 1) Cerebral angiography was not performed.

Treatment and Outcome

The treatments used and outcome of the patients are shown in Table 2. Eleven surgical procedures were performed in the nine patients. Two patients (Cases 7 and 8) underwent a second operation due to recurrence. The approaches used were anterior transcallosal in three cases, frontal transcortical in five and stereotactic biopsy in one (Table 1). During each

operation, demarcation of the mass was relatively sharp except in the area of attachment, which may be the origin of the tumor. The attachment sites of the tumors were the septum pellucidum in seven cases, the lateral wall of the lateral ventricle in one, and the roof of the lateral ventricle in one. Grossly total removal of the tumor was performed in seven and subtotal removal was performed in two cases (Cases 2 and 3). Profuse intraoperative bleeding was the main cause of subtotal removal. Postoperative fractionated radiotherapy (5000–5500 cGy) was administered to three patients. No patient received adjuvant chemotherapy. All patients were followed up with computed tomography (CT) or magnetic resonance (MR) imaging.

After the first total resection of the tumor, two patients (Cases 7 and 8) developed a recurrent tumor 28 and 108 months, respectively. They received a second total resection and postoperative radiotherapy with doses of 5000–5500 cGy. The patients had mild neurological deficits during 43–45 month follow-up. However, the patient experienced a recurrence and eventually had good clinical outcome. Another two patients (Cases 2 and 3) died within 6 months after operation, one with sepsis after subtotal resection and the other with central failure caused by tumor progression and increased ICP. The remaining 5 patients are free of recurrent disease at 52 to 190 months (median 99 months) postsurgery, 4 with good recovery and one with moderate neurological deficits.

Tumor Histology

Light Microscopy showed that all tumors except one exhibited essentially the same features, which are characteristic of these lesions: sheets of monotonously small- to medium-sized neoplastic cells with uniform round-to-oval nuclei and inconspicuous nucleoli. The chromatin pattern was vesicular. The cytoplasm was clear or eosinophilic with indistinct border. Capillary networks were well developed and divided the tumor cells into groups. Some cases showed coagulation-type necrosis. No nuclear pleomorphism or mitosis was seen. Results of Immunohistochemistry of the tumors are shown in Table 3. In all 9 cases, NSE was strongly positive and showed a diffusely granular pattern. Synaptophysin was positive in 7 patients, including the two with tumor recurrence. Chromogranin was negative in all patients and GFAP was only positive in a few cells in one recurrent tumor. The GFAP-positive cells were thought to be entrapped nonneoplastic astrocytes because they had dendritic processes. The MIB1 labeling index (LI) varied from less than 0.1% to 6.8%. Patients with tumor recurrence (Cases 7 and 8) seemed to have higher MIB1-LIs in the first resected specimen than those of non-recurrent tumors (Patients 1, 4, 5, 6, 9). Patients 2 and 3 were not included in the non-recurrent group because they expired early after operation due to sepsis or increased ICP. These two patients also have higher MIB1-LIs as those with tumor recurrence. The results suggested that central neurocytoma with relatively higher proliferative activity was associated with more rapid tumor progression and clinical course.

Discussion

Historical Perspectives and Clinical Presentation

Central neurocytoma first was described in 1982 by Hassoun et al. as a midline neuronal tumor characterized by its intraventricular location, greater frequency among young adults, specific radiologic features, and good postoperative prognosis due to its benign clinical course [1,2,4,6,9,36,39,43]. Neurocytoma can arise from the septum pellucidum, fornix, or the walls of the lateral ventricles (subependymal layer). It may derive from bipotential precursor cells of the periventricular germinal matrix, which are capable of both neuronal and glial differentiation, but maintain a low proliferative potential after birth [4,42,43,44]. Central neurocytoma has a long clinical course. Patients frequently present with headache and visual changes, and the duration of clinical symptoms and signs typically is less than 6 months. Most of these symptoms are attributed to increased intracranial pressure secondary to obstructive hydrocephalus. Most

authors report that central neurocytomas affect young adults without predilection for either sex [43]. However, in our series there were two patients with age over 40 years and female patients predominated. Consequently, central neurocytoma must be considered in the differential diagnosis of anteriorly located lateral ventricular tumors, even in the elderly patients.

Radiology

CT scans typically demonstrate an iso- or slightly hyperdense mass within the body of the lateral ventricles near the foramen of Monro. Areas of hypodensity represent cystic degeneration. Approximately 51% of central neurocytomas demonstrate calcification on CT images [2]. As these tumors are thought to arise from septal nuclei, they are centered at the midline, making the septum pellucidum not seen well in CT scans. The tumors usually have a broad-based attachment to the superior and lateral wall of the ventricle. Obstruction of the interventricular foramen of Monro by the tumor mass usually results in hydrocephalus. Contrast enhancement is mild to moderate for most central neurocytomas. MR imaging usually reveals a mass that is isointense in T1-weighted images [6,7,39]. In T2-weighted images, most central neurocytomas are relatively isointense with cortex. There is usually moderate enhancement after the administration of gadolinium. Angiography shows homogeneous vascularization but is rarely performed for central neurocytomas [8,9]. Based on radiological images, differential diagnosis of central neurocytoma depends on the exact tumor location and age of the patient. Tumors in the lateral ventricle in young adults include oligodendrogliomas, subependymal giant cell astrocytomas, ependymomas, and low grade or pilocytic astrocytomas. Astrocytomas and ependymomas may occur anywhere within the ventricular system, but usually lack intratumoral cysts and calcifications. The intraventricular oligodendroglioma is usually located within the body of the lateral ventricle. However, intratumoral calcifications in oligodendrogliomas are typically large and irregular [10]. A typical central neurocytoma is located in the supratentorial ventricular system of which the anterior half of the lateral ventricle is the most frequent location [5]. Extension into the third ventricle occurs in 26% of central neurocytomas. Isolated third or fourth ventricular occurrence and extraventricular sites are rarely reported [9–14]. There are also rare cases of central neurocytomas with craniospinal dissemination [15]. More recently, proton magnetic resonance spectroscopy (MRS) of central neurocytomas has been reported. Typical patterns include elevated choline, decreased creatine, and NAA [16,17,36,37,38,41].

Neuropathology

Gross pathology for a central neurocytoma is that of a lobulated, well-circumscribed, gray-colored mass. Necrosis and cyst formation are frequently seen and some neurocytomas are very vascular (Fig. 2A). Intratumoral hemorrhage is not typical. The histopathologic appearance of a central neurocytoma can be similar to that of an oligodendroglioma. Both neoplasms have small uniform cells with rounded nuclei and scant cytoplasm resembling perinuclear halos (‘fried egg’ appearance), and many of the intraventricular tumors diagnosed as oligodendrogliomas may indeed represent central neurocytomas [18,36,39,43]. However, alternating fibrillary and cellular areas, scant mitotic activity and a tendency to form ill-defined rosettes in the latter are features against oligodendroglioma [19]. Immunoreactivity against neuron-specific enolase (NSE) and synaptophysin confirms the neuronal nature of the neoplasm. Glial fibrillary acidic protein (GFAP) staining can be found in central neurocytomas. It is unclear whether the GFAP-positive cells represent neoplastic cells or reactive astrocytes. It has been suggested that central neurocytomas originate from bipotential (neuronal and astrocytic) progenitor cells in the periventricular region that persist into adulthood [20,39].

Anaplasia, proliferative potential and biological behavior

Limited data reported previously showed that central neurocytomas have low proliferative potential [3,43,44]. However, anaplasia has been demonstrated in recent literatures. The influence of anaplasia on prognosis is uncertain [6,18–21,43–44]. Enhancement of GFAP positivity and vascular proliferation might suggest a more malignant course [22]. It is not clear if tumors with anaplasia have a higher relapse rate or if they need additional treatment. Yet some central neurocytomas appear to be more aggressive despite benign histology. In addition, they can even disseminate along the CSF pathways [22,23]. In an attempt to clarify the biological behavior of central neurocytomas, the role of proliferation markers, such as Ki-67, has been investigated. Ki-67 antigens react with MIB-1 monoclonal antibodies and a MIB-1 labeling index (MIB1-LI, represents a percentage of proliferative tumor cells) of >2% is a relevant prognostic factor of central neurocytoma for both local control and overall survival [24,36,43–44]. In our series, two patients had a symptomatic relapse and two patients died of progressive tumor growth and surgical complication. The MIB1 LIs in these four patients were easily found > 2% and only one had histologic atypia. It suggests that a clinically more aggressive subgroup of central neurocytomas occurs with elevated proliferative potential that may be revealed by MIB1 LI. Nevertheless, with longer follow-up, it is possible that some of the tumors with low MIB-LI might relapse (as case 8).

Treatment options and outcome

A. Surgery—For most patients with a newly diagnosed intraventricular mass, the first choice of treatment is surgery. The goals of surgery are to re-establish CSF pathways, to maximize a safe resection, and to provide tissue for accurate diagnosis. Although many patients present with signs and symptoms of increased ICP secondary to obstructive hydrocephalus, only rarely is it necessary to place a preoperative shunt. If the patient's neurologic status rapidly declines and there is evidence of hydrocephalus, one should consider external ventricular drainage as a temporizing maneuver prior to definitive microsurgery. After surgical resection of the tumor and re-establishment of CSF pathways, the need for permanent shunting might be avoided, unless the patient continues to have hydrocephalus. In addition, a third ventriculostomy can be useful in patients with noncommunicating hydrocephalus [25]. The surgical approaches for these lateral ventricular tumors include transcortical–transventricular and interhemispheric transcallosal–transventricular routes. Since most neurocytomas are centered on the septum pellucidum, the transcallosal approach typically gives the greatest flexibility for operating on both the right and the left sides of the ventricle. As the tumor is located exclusively on one side, transcortical approach is more reasonable to perform. If the tumor predominantly resides superiorly on the unilateral lateral ventricle, a contralateral interhemispheric route can give the best angle to approach the tumor.

In the latest operations, we routinely use image-guided surgical systems for operating on central neurocytomas. Prior to the start of surgery, the dilated obstructed ventricle can be cannulated with the assistance of an image-guided system, and the ventricle can be drained to reduce intracranial pressure and resistance against medial hemispheric retraction. Once the ventricle is entered and the tumor identified, visualization of normal ventricular landmarks are the key to executing an expedient and safe resection. For larger tumors, an internal debulking with an ultrasonic aspirator is performed and the margins of the tumor are folded in on themselves. As this is done, normal ventricular landmarks such as choroid plexus, foramen of Monro, and ependymal veins come into view. It is important to avoid intruding into the subependymas during tumor resection by taking the ependymal veins and the choroid plexus as important guides of depth. Central neurocytomas are very vascular at times. As more of the tumor is removed, blood supply from the choroidal vessels would be encountered and controlled. Large veins within the body of a big tumor are also common and should be suspected based on review of preoperative imaging studies. After completion of tumor resection, an external ventricular

drain should be placed for CSF drainage until the fluid is clear to avoid delayed hydrocephalus related to intraventricular hemorrhage [5].

B. Radiotherapy and radiosurgery—Since central neurocytomas are usually benign with low proliferative potential, radiotherapy is not theoretically necessary. However, there are several reports claiming that postoperative radiotherapy for central neurocytoma leads to the disappearance or shrinkage of residual tumors [26–28,42–43,45]. Radiotherapy after grossly total resection remains controversial since most patients have long-term tumor control without radiotherapy. Rades et al. [29] reviewed the literature on 310 patients with central neurocytomas and demonstrated that adjuvant radiotherapy after complete resection did not result in improved local control or increased survival. The use of radiation for the residual tumors after subtotal resection is also controversial. Rades et al. [29] found that incomplete tumor resection followed by radiotherapy was superior to incomplete resection alone with significantly better local control, but there was not a significant improvement in survival. Schild et al. [30] demonstrated that the 5-year local control rate for residual central neurocytoma was 100% with radiation compared to 50% without radiation ($p < 0.02$). Kulkarni et al. [27] followed up seven patients who were treated with whole brain irradiation after stereotactic biopsy. There were no procedure-related morbidities, and, overall, one patient had disseminated intracranial disease at 15 months while the other 6 months were symptom-free and had local control at 78 months. Radiosurgery seems more advantageous than adjuvant radiotherapy in the treatment of central neurocytomas because these tumors are mostly well circumscribed in the ventricles. Additionally, unlike larger field of brain irradiation for adjuvant radiotherapy, radiosurgery spares regional structures including the fornix, thalamic and basal ganglia nuclei, and the deep frontal lobe from delayed radiation effects [31]. Even with limited follow-up, it appears that for small, residual, or recurrent tumors, radiosurgery is a reasonable treatment option [36,40,42–45].

C. Chemotherapy—Reports on chemotherapy for central neurocytomas have been more limited. Various combinations of carmustine, lomustine, prednisolone, vincristine, and cisplatin have been used to treat central neurocytomas, but the responses to these agents have not been well documented [32–35]. Chemotherapy may be beneficial from its potential to cause less permanent damage to the brain. In our series, all 9 patients did not receive chemotherapy.

Conclusions

Generally, central neurocytomas have a favorable prognosis, but in some cases the clinical course is more aggressive. Histological features of anaplasia do not predict the biologic behavior of the tumor. The proliferation index MIB1-LI may be useful in predicting tumor relapse. The most important therapeutic modality remains surgery. A safe maximal resection confers the best long-term outcome. Recurrences are mostly local and the patients often recover well after a second resection followed by radiotherapy. We recommend that neurosurgeons consider the possibility of recurrence of central neurocytomas based on the histologic features, especially proliferation index (MIB1-LI), during treatment and follow-up of these patients.

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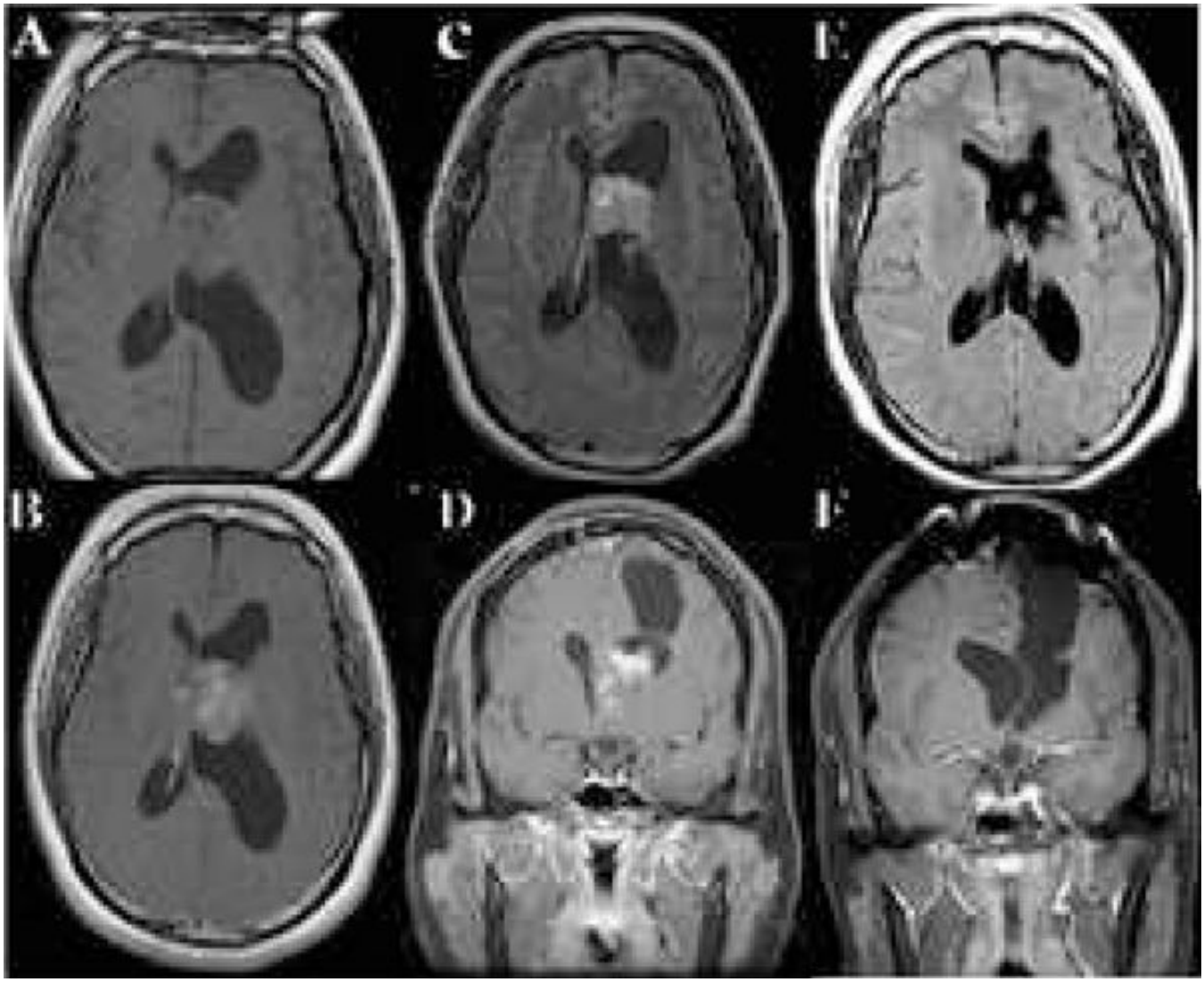

Fig. 1.

Preoperative axial T1-weighted magnetic resonance (MR) images of a patient with central neurocytoma. **A.** Before contrast enhancement; **B.** After contrast enhancement, a large mildly enhanced mass with “soap-bubble” appearance was shown in the left lateral ventricle and attaching to the septum pellucidum. Contrast-enhanced T1-weighted magnetic resonance images 28 months after the first resection demonstrated a recurrent central neurocytoma in Patient 7 as shown in Table 1. **C.** axial view; **D.** coronal view. Postoperative contrast-enhanced T1-weighted magnetic resonance images 43 months after total removal of the recurrent tumor and fractionated radiotherapy showed no enhanced mass in the left lateral ventricle. **E.** axial view; **F.** coronal view.

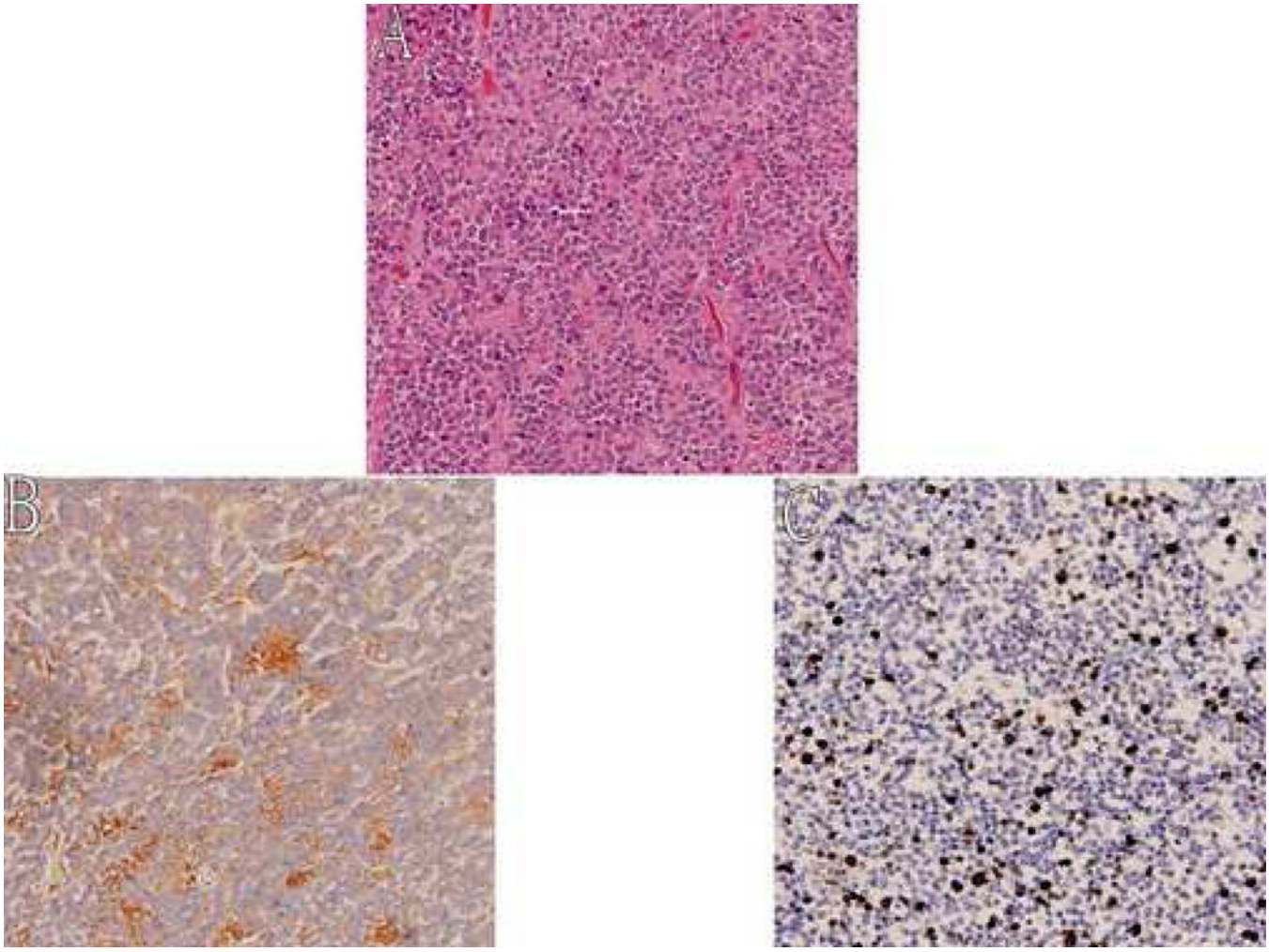

Fig. 2.

Fig. 2A. Photomicrograph (hematoxylin and eosin stain, 200x) of central neurocytoma. Numerous round nuclei with delicate chromatin and indistinct cell border can be seen in the fibrillary and well-vascularized matrix. Some cells have a perinuclear halo. Mild nuclear pleomorphism and mitosis scattered throughout the section. **2B**. Photomicrograph (100x) showing strong immunoreactivity of synaptophysin in central neurocytoma. Synaptophysin in the tumor presented in a diffuse fibrillar pattern. **2C**. Photomicrograph (200x) revealing immunoreactivity of Ki-67 in the first-resected central neurocytoma of patient 7. The proliferation index (MIB1-LI) is evaluated as 6.8%.

Table 1

Basic clinical data of 9 patients with central neurocytoma.

Patient no.	Age (years)	Sex	Initial symptom	Surgical approach	Tumor location
1	22	F	Raised ICP	Transfrontal transcortical	Bilateral LV, 3V
2	43	F	Raised ICP, hemiparesis	Stereotactic biospy	Left LV, thalamus
3	17	M	Raised ICP	Transfrontal transcortical	Left LV, 3V
4	32	F	Raised ICP, seizure, blurred vision	Interhemispheric transcallosal	Right LV, 3V
5	24	F	Raised ICP, seizure	Interhemispheric transcallosal	Right LV
6	18	M	Raised ICP	Transfrontal transcortical	Right LV
7*	43	F	Raised ICP, seizure	Transfrontal transcortical	Left LV, 3V
8*	16	F	Raised ICP	Interhemispheric transcallosal	Right LV, 3V
9	28	F	Raised ICP	Transfrontal transcortical	Left LV

Abbreviation: F: female; M: male; ICP: intracranial pressure; LV: lateral ventricle; 3V: third ventricle; CT: computerized tomography; MRI: magnetic resonance image; +: performed; −: not performed.

*recurrent at age of 45 for patient 7 and at 25 for patient 8; The surgical approach used for the second operation is the same as the first.

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Table 2

Treatment and outcome of 9 patients with central neurocytoma.

Patient No.	Surgical resection	Radiotherapy (cGy)	Outcome	Follow-up period (month)
1	Total	-	Good recovery	190
2	Stereotactic biopsy	5000	Death	6
3	Subtotal	-	Death	1
4	Total	-	Moderate deficits	123
5	Total	-	Good recovery	118
6	Total	-	Good recovery	98
7	Total (total)	- (5000)	Recurrence (mild deficits)	58
8	Total (total)	- (5500)	Recurrence (mild deficits)	56
9	Total	-	Good recovery	52

Patients with good recovery or moderate neurological deficit showed no tumor recurrence. Data in parentheses in Patients 7 and 8 were treatment and outcome of tumor recurrence. No more recurrence was found after the second-time treatments. -, not performed.

Table 3

Histological features in 9 patients with central neurocytoma.

Patient No.	NSE	SP	GFAP	CG	Proliferative activity MIB1-LI (%)	Vascular proliferation
1	+	NT	−	NT	<0.1	−
2	+	+	−	−	3.6–5.4	+
3	+	+	−	−	1.3–2.2	+
4	+	+	−	−	<0.1–0.5	−
5	+	+	−	−	0.6–1.5	−
6	+	+	−	−	1.6–2.4	−
7	+ (+)	− (+)	− (+)	− (−)	2.0–6.8 (2.7–3.4)	− (+)
8	+ (+)	+ (+)	− (−)	− (−)	1.4–2.0 (0.1–0.5)	− (+)
9	+	−	−	−	1.3–1.8	−

Immunohistochemistry was used to detect the presence (+) or absence (−) of neuron-specific enolase (NSE), synaptophysin (SP), glial fibrillary acidic protein (GFAP), chromogranin (CG) and Ki-67 (MIB1). MIB1 labeling index (MIB1-LI) was calculated as percent of Ki-67-positive cells for evaluating tumor proliferation. Data in parentheses in Patients 7 and 8 were obtained from the recurrent tumors. NT: not tested.