Choroid Plexus Tumors: Experience of 10 Cases with Special References to Adult Cases

Manoj BOHARA,1 Masashi HIRABARU,1 Shingo FUJIO,1 Michiyo HIGASHI,2 Hajime YONEZAWA,1 Prasanna KARKI,1 Ryosuke HANAYA,1 Hirofumi HIRANO,1 Hiroshi TOKIMURA,1 and Kazunori ARITA1

Departments of 1Neurosurgery and 2Human Pathology, Graduate School of Medical and Dental Sciences, Kagoshima, Kagoshima

Abstract

Choroid plexus tumors (CPTs) are rare intraventricular neoplasms accounting for about 0.3–0.6% of all intracranial tumors. This retrospective study on CPTs presents clinico-pathological features and management strategies based on a 20-year single-institutional experience. This series included 10 consecutive patients with pathologically proven CPTs; 5 choroid plexus papillomas (CPPs), 3 atypical CPPs (ACPPs), and 2 choroid plexus carcinomas (CPCs). Their clinical, radiological, and histopathological features as well as management including follow-up studies were reviewed. The patients included five males and five females, aging from 0 years to 61 years with median of 28 years. The affected site was lateral ventricle in two adults and fourth ventricle in eight patients; four children and four adults. The most common symptoms were gait disturbance and memory disturbance. All the patients underwent craniotomy with total, subtotal, and partial removals achieved in 50%, 40%, and 10% of the patients, respectively. The occurrence of the high grade subtypes was 50% in both the adult and pediatric groups. The Ki-67/MIB-1 index increased across the three histological subtypes, from CPP to ACPP and then to CPC. Adjuvant therapy was administered in three patients. The two patients (one adult and one child) with CPC died of whole central nervous system dissemination. At a median of 62-month follow-up, the other eight patients were alive, with only one patient having recurrence and reoperation. The results demonstrate that gross total resection is usually curative for CPP and ACPP, and adjuvant chemoradiotherapy would be required for CPC and incompletely resected ACPP.

Key words: choroid plexus papilloma, atypical choroid plexus papilloma, choroid plexus carcinoma, magnetic resonance imaging, adult, MIB-1

Introduction

Choroid plexus tumors (CPTs) are rare intraventricular neoplasms originating from choroid plexus that account for only 0.3–0.6% of all intracranial tumors. These tumors are seen more frequently in children, especially in first two years of life, having an incidence of 1.5–4% in this age group. The incidence is very low in the non-pediatric population, 0.3/1 million population/year. They are mainly located in lateral ventricles in children and fourth ventricles in adults. Rarely, they occur in cerebellopontine angle, suprasellar region, or even in the sacral canal. The majority of the tumors are benign choroid plexus papilloma (CPP), WHO Grade I, but tumors with malignant features, choroid plexus carcinomas (CPC), and atypical choroid plexus papillomas (ACPPs) may also arise. Histological grading is recognized as an important prognostic factor and also affects the decision making concerning adjuvant radio- and chemotherapy. Herewith, we retrospectively studied 10 cases of CPTs treated in Kagoshima University Hospital during the last 20 years with special reference to adult cases.

Patients and Methods

Ten patients with CPTs who have been treated at Kagoshima University Hospital from 1995 through 2014 were retrospectively studied. The clinical, radiological, histopathological, and treatment data were obtained from patients’ records and accumulated in database under the protection of anonymity. Pre- and post-treatment neuroimages were reviewed. The pathological

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diagnoses were confirmed and additional immuno-
histochemical studies were performed as necessary. The pathological evaluation was done according to the criteria of World Health Organization (WHO) 2007 classification.10 Karnofsky Performance Status (KPS) scores were used to assess the general well-
being and daily activities of the patients after the operation.

Statistical analysis was performed using Kruskal-
Wallis test and Mann-Whitney test with Prism 5 software (GraphPad, San Diego, California, USA). Values are shown as the mean ± standard error (SE). A p value of less than 0.05 was considered statistically significant.

This retrospective study was approved by Kagoshima University Hospital’s ethical committee (Reference number 22–50) and has been performed in accordance with the ethical standards laid down in the 1964 declaration of Helsinki.

Results (Table 1)
The patients included five males and five females, aged 0–61 years with median of 28 years. The subjects comprised 0.45% (10/2,203) patients with brain tumors treated in our hospital during the same period. Six of the 10 cases were over 20 years old.

The affected site was lateral ventricle in two adult cases and fourth ventricle in eight patients; four children and four adults. The clinical features were gait disturbance (4 patients), memory disturbance (3), chronic headache (3), macrocrania (2), truncal ataxia (2), visual disturbance (1), diplopia (1), and vertigo (1). All the patients underwent preoperative computed tomography (CT) and magnetic resonance imaging (MRI) scans. On CT scans, the tumors were hypodense in six cases, iso- to hyperdense in two cases, hypo- to isodense in one case, and hyperdense in one case; calcification was seen in seven cases. On \( T_1 \)-weighted MR images, the tumors showed hypo- to isointensity in all but one case and on \( T_2 \)-weighted MR images, they were hyperintense in all the cases, with heterogenous enhancement on contrast injection. All the patients underwent craniotomy with transcortical approach in lateral ventricular tumors. Total, subtotal, and partial removals were achieved in four, four, and one cases, respectively. Two-staged operations were performed in two cases.

Histologically, five cases were diagnosed with CPP, three cases with ACPP, and two cases with CPC. The mean MIB-1 index was 2.6% (range: 1–5%) in CPP, 12.9% (range: 3–30%) in ACPP, and 50.1% (range: 30.2–70%) in CPC (p = 0.052, Kruskal-Wallis test; Fig. 1A). MIB-1 index was significantly higher in histologically higher grade tumors (ACPP and CPC combined) as compared to CPP (p = 0.04, Mann-Whitney test; Fig. 1B). MIB-1 index was significantly higher in histologically higher grade tumors (ACPP and CPC combined) as compared to CPP (p = 0.04, Mann-Whitney test; Fig. 1B). MIB-1 index was significantly higher in histologically higher grade tumors (ACPP and CPC combined) as compared to CPP (p = 0.04, Mann-Whitney test; Fig. 1B). MIB-1 index was significantly higher in histologically higher grade tumors (ACPP and CPC combined) as compared to CPP (p = 0.04, Mann-Whitney test; Fig. 1B). Mutated p53 was positive in all the cases with ACPP and CPC but only in one of five cases (20%) with CPP. INI-1 was positive in all the cases.

Postoperative KPS score increased in six patients and remained unchanged in four. One case with ACPP and CPC each had adjuvant chemoradio-
therapy. A neonate with CPC (case 1) only underwent chemotherapy. Two patients with CPC died of whole central nervous system dissemination at 10 months and 13 months postoperatively. The other eight patients were followed up for a median of 62 (range: 46–171) months after initial surgery. One of the patients (case 3) with CPP had recurrence after 7 years of initial surgery and underwent reoperation for total removal.

Illustrative Cases

I. Case 5
A 26-year-old woman presented with a 1-year history of headache and general fatigue, a 6-month history of memory disturbance, and progressive visual impairment due to papillary edema. An MRI showed a large tumor filling bilateral lateral ventricles with left sided predominance. The tumor demonstrated iso- to low-signal intensity on \( T_1 \)-WI (Fig. 2A) and iso- to high-signal intensity on \( T_2 \)-WI (Fig. 2B) due to multiple cystic components. Gadolinium contrast injection enhanced the mass heterogeneously (Fig. 2C). MR spectroscopy of the tumor demonstrated a single choline peak (Fig. 2D).

Left frontal transcortical-transventricular approach for utmost removal resulted in residual tumor due to adherence to the ventricular wall and profuse bleeding. Histologically, the resected specimen was composed of papillary proliferation of columnar and cuboidal epithelial cells with mild to moderate nuclear pleomorphism (Fig. 3A). Two or three mitoses per 10 high-power fields were found. The MIB-1 index was 5.7% (Fig. 3B), and the tumor exhibited mutated p53-positive nuclei (Fig. 3C). Immunore-
activity for INI-1 was also positive (Fig. 3D). The tumor was diagnosed as ACPP. Postoperative MRI showed residual tumor adherent to the ventricular wall. The patient was postoperatively treated with one course of ICE (ifosfamide, cisplatin, and etoposide) chemotherapeutic regimen before the second operation through the same surgical approach, which resulted in subtotal removal of the tumor (Fig. 4E). Two courses of ICE chemotherapy followed by 25 Gy radiation to whole central nervous system and 25 Gy boost radiation to whole ventricular system were performed.
<table>
<thead>
<tr>
<th>Case</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Location (ventricle)</th>
<th>CT</th>
<th>MRI</th>
<th>Surgery</th>
<th>Surgical result</th>
<th>Pathology</th>
<th>MIB-1 / Ki-67 index (%)</th>
<th>Adjuvant therapy</th>
<th>Preoperative KPS</th>
<th>Postoperative KPS</th>
<th>Outcome (GOS)</th>
<th>Follow-up months after initial surgery</th>
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<tr>
<td>1</td>
<td>0</td>
<td>M</td>
<td>Intrauterine head enlargement</td>
<td>4th</td>
<td>isoto hyperdense, ventricular dilatation</td>
<td>T1 hypo-iso, T2 hyper, heterogenous enhancement</td>
<td>Suboccipital</td>
<td>Partial</td>
<td>CPC</td>
<td>30.2</td>
<td>PE</td>
<td>20</td>
<td>20</td>
<td>Death</td>
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<td>2</td>
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<td>F</td>
<td>Macrocrania</td>
<td>4th</td>
<td>isodense, ventricular dilatation</td>
<td>T1 iso, T2 iso-hyper, heterogenous enhancement</td>
<td>Suboccipital</td>
<td>Total</td>
<td>ACPP</td>
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<td>40</td>
<td>90</td>
<td>GR</td>
<td>171</td>
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<tr>
<td>3</td>
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<td>M</td>
<td>Gait disturbance, Truncal ataxia</td>
<td>4th</td>
<td>isodense, calcification, ventricular dilatation</td>
<td>T1 iso, T2 hyper</td>
<td>1) Suboccipital 1) Total 1) Total</td>
<td>CPP</td>
<td>4</td>
<td>None</td>
<td>40</td>
<td>90</td>
<td>Reoperation for recurrence at 7 years; GR</td>
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<td>isodense, calcification, ventricular dilatation</td>
<td>T1 iso, T2 hyper, heterogenous enhancement</td>
<td>Suboccipital; 2-stage</td>
<td>Subtotal</td>
<td>CPP</td>
<td>5</td>
<td>None</td>
<td>90</td>
<td>100</td>
<td>GR</td>
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<td>26</td>
<td>F</td>
<td>Memory disturbance, Gait disturbance, Visual disturbance</td>
<td>Left lateral</td>
<td>hypo-isodense, calcification</td>
<td>T1 iso, T2 hyper heterogenous enhancement</td>
<td>Transcortical; Subtotal 2-stage</td>
<td>ACPP</td>
<td>5.7</td>
<td>Whole brain 25 Gy, Whole spine 25 Gy, Extended local 25 Gy, ICE 3 courses</td>
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<td>80</td>
<td>MD*</td>
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<td>Suboccipital</td>
<td>Total</td>
<td>ACPP</td>
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<td>100</td>
<td>GR</td>
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<td>60</td>
<td>M</td>
<td>Chronic headache</td>
<td>Right lateral</td>
<td>isodense, calcification</td>
<td>T1 iso, T2 iso-hyper heterogenous enhancement</td>
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<td>Subtotal</td>
<td>CPC</td>
<td>70</td>
<td>Whole brain 30 Gy, Local 10 Gy + Cyberknife TMZ, PE, and MTX</td>
<td>100</td>
<td>100</td>
<td>Death</td>
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<td>Memory disturbance</td>
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<td>Subtotal</td>
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<td>None</td>
<td>90</td>
<td>90</td>
<td>GR</td>
<td>49</td>
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</table>

given. At 66 months after the first surgery, there had been no recurrence but she had severe visual impairment that was sequel of the increased intracranial pressure preceding the surgery.

II. Case 8
A 46-year-old woman with no remarkable past medical history was incidentally found to have brain tumor during a medical checkup. MRI showed a lesion 30 × 30 × 30 mm in size in fourth ventricle with iso- to high-signal intensity on both T\(_1\)-WI (Fig. 4A) and T\(_2\)-WI (Fig. 4B). On contrast injection, the tumor was enhanced with cauliflower shape (Fig. 4C). MR spectroscopy of the tumor showed a single choline peak (Fig. 4D). The total resection of tumor was attempted by suboccipital craniotomy. Histologically, the resected specimen was composed of papillary proliferation. Two mitoses per 10 high-power fields were found (Fig. 5A). The MIB-1 index was 3% (Fig 5B). The tumor demonstrated mutated p53 (Fig. 5C) and INI-1 (Fig. 5D) expression. It was diagnosed as ACPP, same as case 5. Postoperative MRI revealed the complete resection of tumor (Fig. 4E). We did not perform chemotherapy and radiotherapy because of complete resection of the tumor. The latest follow-up imaging at 52 months after the initial surgery showed no recurrence of the tumor.

III. Case 9
A 60-year-old man suffering from chronic headache underwent MRI, which showed a tumor 22 × 21 × 23 mm in size in trigone of right lateral ventricle with iso- to low-signal intensity on T\(_1\)-weighted images (WIs) (Fig. 6A). The tumor showed iso- to high-signal intensity on T\(_2\)-WI (Fig. 6B) and was heterogeneously enhanced on gadolinium injection (Fig. 6C). MR spectroscopy of the tumor demonstrated a single choline peak (Fig. 6D). Positron emission tomography (PET) showed high accumulation of 11C-methionine (Fig. 6E) and moderate uptake of 18F-fluorodeoxyglucose (Fig. 6F) in the lesion. The total resection of tumor was attempted by corticotomy through the middle temporal gyrus.
The tumor was severely adhered to choroid plexus and wall of lateral ventricles. After the separation from lateral ventricular wall, the tumor was gross totally removed. Postoperative course was uneventful. Histologically, the resected tumor was composed of coarse papillary proliferation of cuboidal epithelial cells with conspicuous pleomorphism. Increased cellularity and frequent mitoses were found (Fig. 7A). The Ki-67/MIB-1 positive index was 70% (Fig. 7B). The immunoreactivity of neoplastic cells was positive for mutated p53 (Fig. 7C), INI-1 (Fig. 7D), and cytokeratin 7 (Fig. 7E). The cells were also positive for CAM 5.2 and transthyretin, and partially positive for S-100 protein but negative for cytokeratin 20 and glial fibrillary acidic protein (GFAP). These histologic features were compatible with CPC. Postoperative MRI showed gross total removal of the tumor (Fig. 6G). The patient was treated with whole brain (30 Gy/15 fractions) and extended local (10 Gy/5 fractions) radiation and concurrent temozolomide regimen (200 mg/kg/day, 5 days/month) followed by stereotactic boost radiotherapy (biologically equivalent dose to 23 Gy) using cyber-knife unit. However, 3 months after the completion of treatment, the patient had lower back pain and urinary retention. MRI showed multiple intradural extramedullary tumors in cervical, thoracic (Fig. 6H), and lumbar spine and also peri-mesencephalic dissemination. The excision of the largest tumor through Th12/L1 laminectomy provided improvement of urinary symptoms. Two cycles of PE (cisplatin and etoposide) regimen followed, which failed to control the rapid growth of metastatic lesions. Intraventricular injection of methotrexate and vincristine via Ommaya’s reservoir installed on left lateral ventricle could not control
the disseminations either. He died of respiratory failure 13 months after the first operation.

**Discussion**

CPTs are rare neoplasms comprising only 0.3–0.6% of all primary brain tumors;\(^1\) the incidence is compatible with our brain tumor series. The CPTs are well known to preferentially arise in children.\(^2,5–7\) So, it is noticeable that the age at diagnosis of our CPT patients was rather higher, median of 28 years, compared to previous reports. According to the extensive meta-analysis including 566 patients Wolff et al. performed, the median age of the patients was 3.5 years.\(^8\) It was reported to be 3 years in another meta-analysis including 857 cases by Wrede et al.\(^9\) Lateral ventricular CPTs is usually present in patients younger than 20 years whereas the age distribution shifts to older in cases with fourth ventricular CPTs; the median age was reported to be 1.5 years and 22.5 years for lateral and fourth
ventricle, respectively. Fourth ventricular CPTs have also been reported in pediatric patients; all the four pediatric patients in our study had CPTs in fourth ventricle, which is an unusual finding.

Adult cases with lateral ventricular CPTs have rarely been reported. Our series included two lateral ventricular CPTs in adults; both were high-grade CPTs, a CPC, and an ACPP. Locations of the high-grade CPTs are more frequently supratentorial than infratentorial. Of six adult patients in our study, 50% had aggressive subtypes (ACPP and CPC). The occurrence of these higher grade subtypes in adults is similar to that reported as 40% by Turkoglu et al.

As for the clinical presentation, it has been reported that children have slower onset of symptoms like headache and visual symptoms whereas adults present with more rapid symptoms mainly due to fourth ventricular obstruction. This is mainly attributed to the preferential location of the tumors, which are more common in lateral ventricles in children whereas fourth ventricle in adults. However, in our study all the pediatric patients had fourth ventricular tumors, so presented with rapid symptoms of obstruction.

CPCs are malignant CPTs with poor prognosis, designated as WHO Grade III, showing frankly aggressive histologic features; frequent mitosis, increased cellular density, nuclear pleomorphism, disarrangement of papillary pattern, and necrotic area. ACPP is an intermediate grade subtype (Grade II) of CPT which lies between CPP (I) and CPC (III), which is an intermediate grade subtype (Grade II) of CPT. The nature of the intraventricular tumors with mutated INI-1 gene is, however, still controversial, in view of possibility of misdiagnosed intraventricular atypical teratoid/rhabdoid tumor.

In order to institute proper treatment strategy for malignant CPTs before surgery, preoperative image diagnosis is essential. On MRI, CPT is generally depicted as a well demarcated multinodular intraventricular tumor with fine granular appearance reflecting papillary growth of the tumor. Differential diagnoses should include meningioma, ependymoma, and metastatic tumor. The former two tumors generally show homogeneous enhancement pattern rather than granular and multinodular appearances. In some metastatic tumors, patient’s history and multiplicity may support the preoperative diagnosis.

In terms of diagnosis of subtypes, ACPP and CPC may show heterogeneous enhancement pattern and multiple cysts with necrosis or intratumoral hemorrhage and invasion into surrounding brain tissue. In our series, case 5 with ACPP showed multiple cysts on preoperative MRI, which may be a clue for predicting malignant nature. But in case 9 with CPC, there was neither cyst nor invasion.

MR spectroscopy shows a single choline peak without N-acetylaspartate (NAA) peak in CPT. Malignant subtypes of CPTs are reported to show higher choline
peak than CPP, as was seen in our cases. Lactate peak was absent in our malignant subtypes, which was reported to be positive in previously reported cases. Previous report on PET studies has shown significantly higher uptake of Methionine in CPP compared to low-grade glioma but 18F-fluorodeoxyglucose (FDG) uptake did not differ between the two. In our case 9 with CPC, PET scan revealed higher uptake of both methionine and FDG. While uptake on PET scan has been reported to be correlated with malignancy, there are no specific studies differentiating subtypes of CPTs on the basis of PET. Future prospective studies should be carried out to determine the usefulness of these advanced neuroimaging modalities for the prediction of the grade of CPTs.

CPPs are usually cured by surgery, and rarely recur. Survival time of the patients with CPTs depends on the histological subtype. In our study, all the patients with CPP and ACPP were alive at the latest follow-up. One adult and one pediatric patient with CPC died 13 and 10 months after surgery, respectively. It has been reported that the 5-year survival rate was 58% after complete tumor resection in cases with CPC. According to Wrede et al.’s meta-analysis, complete surgical resection, radiation, and chemotherapy were associated with the better overall survivals in cases with CPC. However, the total removal of CPC is hindered by the large size, invasiveness, and excessive bleeding. Preoperative chemotherapy and multi-staged surgery could be the choices for large and invasive CPC. Intraoperative radiation therapy has also been used in case of pediatric CPC to minimize damage to normal tissue. In the reported series, the postoperative chemotherapeutic regimens widely varied in agents and doses. The ongoing international controlled randomized study (CPT-SIOP-2000) is set to know the effect of chemotherapy consisting of etoposide, vincristine, carboplatin, or cyclophosphamide in CPC and incompletely resected ACPP. The local fractionated radiation of 54 Gy is given to the patients older than 3 years after 2 courses of chemotherapy. Craniospinal irradiation (CSI) with 35.2 Gy and a local boost up to a total of 54 Gy is given to patients with CPC nonresponsive to the chemotherapy and those with metastasized CPC and ACPP. The 5-year overall survival probabilities were 100% in CPP, 89% (95% CI, 71–100%) in ACPP, and 36% (95% CI, 9–100%) in CPC patients according to the first analysis of the study published in 2009. In our series, the two patients with CPC had chemotherapy including ifosfamide, cisplatin, etoposide, temozolomide, and intrathecal methotrexate. One of them (case 9) also underwent adjuvant radiotherapy, with whole brain (30 Gy/15 fractions) and extended local (10 Gy/5 fractions) radiation, but it failed to prevent dissemination of the tumor. Considering the ominous prognosis of this pathology, more intense chemotherapeutic regimen combined with CSI could have been a choice. It has also been reported that patients with CPC who received CSI had better progression-free survival compared with those receiving less than CSI. For patients under the age of 3, chemotherapy is the only adjuvant therapy recommended based on the current literature considering the adverse effects of radiation on the developing brain.

The usefulness of Temozolomide for CPTs is yet to be determined. It has been reported that the combined bisulfite restriction analysis confirmed over 10% methylation of CpGs 17 and 31 in 58% of CPTs. In our study, immunohistochemical expression of MGMT was negative (< 10% of neoplastic cells) in 1 (10%) of 10 cases. The efficacy of Temozolomide for CPTs and its relation with MGMT methylation is to be elucidated.

The most important limitation of this study is the small number of patients in our series, thus it is not possible to reach definitive conclusions. And our study carries the limitations inherent to a study of retrospective nature. Furthermore, we presented a single institutional experience, making these results difficult to generalize. Nevertheless, this study demonstrates a long-term longitudinal follow-up of the patients with median follow-up period of 62 months and also shows that total resection is usually curative in CPP and ACPP offering long-term recurrence-free survival. Another inherent weakness of the study is 20-year long study period during which neurosurgical and chemoradiotherapeutic techniques have substantially progressed. However, optimal resection is the treatment of choice and the role of adjuvant therapy in CPTs still remains an important domain to be clarified.

**Conclusion**

We retrospectively studied 10 patients with CPTs treated in our institution for the last 20 years, with special reference to the adult cases. The results demonstrate that gross total resection is usually curative for CPP and ACPP, and adjuvant chemo-radiotherapy would be required for CPC and incompletely resected ACPP. Inclusion of more number of cases and further longitudinal follow-up should be required to deduce more definitive conclusions.
Conflicts of Interest Disclosure

Three pediatric cases of this series have been published as a case report in Japanese language previously. However, the illustrative cases presented here are different and those of adults, and none of the figures in this article has been previously published anywhere in any form.

The authors declare that there are no conflicts of interest regarding the publication of this article. All authors who are members of the Japan Neurosurgical Society (JNS) have registered Online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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Address reprint requests to: Hirofumi Hirano, MD, PhD, Department of Neurosurgery, Graduate School of Medical and Dental Sciences, Kagoshima University, 8-35-1 Sakuragaoka, Kagoshima 890-8520, Japan. e-mail: hirahira@m2.kufm.kagoshima-u.ac.jp