Choroid Plexus Papillomas of the Cerebellopontine Angle

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OBJECTIVE: **Choroid plexus papillomas (CPPs) of the cerebellopontine angle (CPA) are extremely rare. We present a series of 21 cases operated on in the last 7 years at our institution.**

METHODS: **During the period from January 2008 to October 2015, we encountered 102 histologically established cases of CPPs, of which 21 were located in the CPA region. Clinical profiles, radiologic features, surgical procedures, intraoperative findings, and outcomes were extracted from the patient records and neuroimaging data.**

RESULTS: **The 21 CPPs in the CPA region accounted for 20.5% of all CPPs. Two of the tumors occurred in pediatric patients. Tumor size was 2.5–4.7 cm. The rate of calcification was higher in the CPPs in the CPA region. Peritumoral cysts and cysts with small nodules were observed in our cases. A far lateral suboccipital approach was chosen for CPPs protruding inferiorly into the foramen magnum region ($n = 14$), and a suboccipital retrosigmoid approach was chosen for the other tumors. Total resection was achieved in 18 patients, and subtotal resection was achieved in 3 patients. During the follow-up period, only 1 patient experienced recurrence 32 months after the first operation. The recurrence turned out to be an atypical CPP.**

CONCLUSIONS: **CPPs in the CPA region cannot be easily differentiated from other tumors preoperatively. Cysts and calcifications appear on neuroimaging. CPPs in the CPA region usually protrude inferiorly into the foramen magnum region. As much tumor should be removed as possible to avoid recurrence and malignant transition.**

INTRODUCTION

Choroid plexus tumors (CPTs) are rare papillary neoplasms derived from choroid plexus epithelium; they account for only approximately 0.4%–0.6% of all intracranial tumors[1-3] but 10%–20% of brain tumors occurring throughout the first year of life.[3,4] The World Health Organization (WHO) classification of central nervous system tumors divided CPTs into choroid plexus papilloma (CPP; WHO I), atypical choroid plexus papilloma (APP; WHO II) and choroid plexus carcinoma (CPC; WHO III).[5] CPP is generally a histologically benign, slow-growing tumor; malignant evolution may occur in 10%–30% of cases, mainly in the lateral ventricle.[6,7]

These tumors predominantly arise in supratentorial locations, such as lateral ventricles in children and the fourth ventricle in adults.[8] When all age groups are considered, the most frequent location is the lateral ventricle (43%–67%), followed by the fourth ventricle (24%–39%) and third ventricle (9.5%–11%).[1,9,10] Only approximately 9% of all CPTs are located in the cerebellopontine angle (CPA), and these are almost exclusively found in adults.[10-13] To the best of our knowledge, <10 pediatric cases of CPP have been reported in the CPA region.[14-17] Other rare locations include the sellar or suprasellar region,[18,19] pineal region,[20] brainstem,[21,22] sacral canal,[23] cerebellum,[24] and cerebral hemisphere.[8]

Key words
- Cerebellopontine angle
- Choroid plexus papillomas
- Clinical features
- Malignant transition
- Surgery

Abbreviations and Acronyms
APP: Atypical choroid plexus papilloma
CPA: Cerebellopontine angle
CPC: Choroid plexus carcinoma
CPP: Choroid plexus papilloma
CPT: Choroid plexus tumor
CT: Computed tomography
GTR: Gross total resection
MRI: Magnetic resonance imaging
OS: Overall survival
WHO: World Health Organization

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We performed a retrospective analysis of the clinical features, neuroimages, treatment, and outcomes of 21 cases of CPPs in the CPA region that were treated during the past 7 years at the Beijing Tiantan Hospital, Capital Medical University. Two of the tumors were in pediatric patients, and 1 tumor underwent malignant evolution. To the best of our knowledge, this is the largest series of its kind in the medical literature to date.

MATERIALS AND METHODS

Patients

During the period from January 2008 to October 2015, we encountered 102 histologically established cases of intracranial CPPs, of which 21 were located in the CPA region. The clinical profiles, radiologic features, surgical procedures, intraoperative findings, and outcomes were extracted from the patient records and neuroimaging data. Informed consent was obtained from each patient. To protect patient privacy, we removed all identifiers from our records. Each patient had undergone resection, and the pathologic diagnosis was confirmed.

Clinical and Radiologic Evaluations

All patients underwent standard clinical and neurologic examinations and routine laboratory tests. All patients underwent preoperative craniocerebral magnetic resonance imaging (MRI) or computed tomography (CT). The extent of resection was determined by review of postoperative images and divided into total resection/gross total resection (GTR), subtotal resection (STR), and partial resection. In addition, patients were asked to judge the effects of surgery on their performance. Patients usually underwent follow-up clinical and MRI examination within 3 months of surgery. If the patient's condition remained stable or improved, clinical follow-up visits were performed at least annually. If patients had new neurologic deterioration, MRI was performed.

Histologic Examinations

Surgical specimens were fixed in 10% neutral buffered formalin. Specimens were paraffin embedded, and routine hematoxylin-eosin staining was carried out. Immunohistochemical staining was performed on the formalin-fixed, paraffin-embedded tissue according to the avidin–biotin peroxidase complex method. We used the following primary monoclonal antibodies: epithelial membrane antigen, glial fibrillary acidic protein, cytokeratin, synaptophysin, S-100 protein, and vimentin. Two pathologists (Dr. L.L and Dr. GL.L) performed all the pathologic examinations.

RESULTS

Clinical Characteristics

We encountered 126 cases of CPTs, including 102 CPPs, 20 APPs, and 4 CPCs. The 21 cases with intracranial CPPs in the CPA region accounted for approximately 16.7% of all intracranial CPTs and 20.5% of all CPPs observed within the same time period at our hospital. Patient data are listed in **Table 1**. The age distribution of patients was 7–65 years (average age, 41.8 years). The peak incidence was in the sixth decade of life. Two patients were children, age 7 years and age 14 years. There was a slight female predominance, with 9 male and 12 female patients. Patient symptoms included headache; dizziness; unsteady gait; vomiting; and paralysis of cranial nerves V, VII, and VIII. One pediatric patient presented with mental retardation. Headache was the most frequently reported first symptom ($n = 9$). Paralysis of cranial nerves V, VII, and VIII was found in 50% of the patients. The duration of symptoms ranged from 1 month to >10 years. The preoperative diagnosis was CPP in only 2 patients; in most patients in the series, the preoperative diagnosis was meningioma or schwannoma.

The tumor size, based on greatest diameter on neuroimaging, was 2.5–4.7 cm. The mean tumor size in the 2 pediatric patients was approximately 3.5 cm. There were 11 patients with tumors >3.5 cm in diameter and 10 patients with tumors <3.5 cm. The preoperative status of facial and cochlear nerve function was compared between the 2 groups with χ^2 test with 1 degree of freedom. Statistical comparison did not reveal any significant difference concerning preoperative facial and cochlear nerve function between the 2 groups.

Neuroradiologic Characteristics

Based on neuroimaging, 7 tumors were located on the left side and 12 were located on the right. Preoperative CT scans were available for 12 cases, and preoperative MRI scans were available for 18 cases. On CT scans, tumors were isodense ($n = 3$), hypodense ($n = 2$), hyperdense ($n = 5$), or mixed density ($n = 2$). Calcification was shown on CT scans. There were 8 patients (66.7% [8 of 12] of the patients with CT scans) with calcification, including stippled ($n = 4$), dense ($n = 1$) and patchy ($n = 3$) calcification (**Figure 1**). Enlargement of the internal auditory canal was not observed in any patients. On T1-weighted MRI images, lesions were found to be isointense ($n = 9$) or hypointense ($n = 9$) to the parenchyma. On T2-weighted images, lesions were isointense ($n = 8$) or hyperintense ($n = 10$). Characteristic contrast enhancement was evident; 15 patients had homogeneous enhancement, and 5 had heterogeneous enhancement (**Figure 2**). A cystic component (**Figure 3**) was encountered in 5 cases. In 4 cases, the cyst was peritumoral; however, in patient 3, MRI showed 1 giant cyst with a small nodule. The contrast enhancement was present in nodule and not the cyst. Hydrocephalus was seen in 7 cases.

Treatment and Follow-Up

Microsurgical excision was the treatment of choice for CPPs. All patients underwent surgery via a suboccipital retrosigmoid approach ($n = 7$) or a far lateral suboccipital approach ($n = 14$) in the lateral decubitus position. Total resection of the tumor was achieved in 18 cases, and STR was achieved in 3 cases. Grossly, the tumor was described as a reddish to gray, cauliflower-shaped mass with a smooth irregular surface. During the operation, the origin of CPP from the choroid tuft outside the foramen of Luschka was seen in 8 patients. Adhesion of tumor to the facial nerve and auditory nerve was observed in only a 5 patients, whereas tight adhesion to the lower cranial nerves was present in 16 cases. No tumor protruded to the internal auditory canal. All patients received intraoperative neurophysiologic monitoring to protect the cranial nerves. To protect neurologic function, 3 patients underwent STR. To reduce bleeding, disconnection of the major feeding artery before debulking the tumor was needed. In patient 3 with a

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