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Clinical Article

Unusual Radiologic Findings and Pathologic Growth Patterns on Choroid Plexus Papillomas

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Objective : Choroid plexus papillomas (CPPs) are generally regarded as benign tumors with typical radiologic and pathologic findings. However, they sometimes have unusual findings. We have analyzed radiologic findings and pathologic growth patterns on CPPs.

Methods : The study group included 5 male and 5 female patients (age range, 3 months to 58 years : median, 29 years). The study group included 3 pediatric and 7 adult patients. All patients underwent surgery; 9 patients had a gross total resection and 1 patient had a subtotal resection. We analyzed the radiologic findings (location, size, mottle-like appearance, enhancement, calcifications, and hydrocephalus) and pathologic growth patterns (typical papillary, papillary and solid, and papillary and tubular).

Results : The median follow-up duration was 21.3 months (range, 4-47.8 months). There were no recurrences after initial treatment. All patients had benign CPPs. Pediatric CPPs were 3.2 cm masses (range, 2.7-4 cm) with homogeneous enhancement and a mottle-like appearance, which pathologically showed the papillary growth pattern. Hydrocephalus was present in all pediatric patients. Postoperatively, subdural hygroma had occurred in two patients. In adults, CPPs were located in the fourth ventricle in 6 patients and suprasellar area in 1 patient. The size varied from 0.5-4.2 cm. Hydrocephalus and calcifications occurred in 3 and 4 patients, respectively. Three patients showed the heterogeneous enhancement without a mottle-like appearance and pathologically showed combined papillary and solid growth in 2 patients and papillary and tubular growth in one. Postoperatively, two patients with large masses had injuries of the brainstem and underwent shunt procedures for aggravation of hydrocephalus.

Conclusion : CPPs may show unusual radiologic findings, which preoperatively give the difficulty to be differentiated from other tumors. CPPs with unusual radiologic findings showed the combined pathologic growth patterns.

Key Words : Choroid plexus papilloma · Complications · Pathology · Radiology.

INTRODUCTION

Choroids plexus papillomas (CPPs) are rare tumors of neuroectodermal origin that account for less than 1% of brain tumors in adults and represent 2-4% of pediatric brain tumors[6,18]. The most common site of origin in the pediatric age group is within the lateral ventricle, whereas the fourth ventricle is the most common site in adults[18]. The most important radiologic finding is the intense contrast enhancement, which is homogeneous and shows typical mottled- and cauliflower-like appearance on the edge of tumors in most cases[1]. Pathologically, these tumors exhibit papillary structures lined by cuboidal epithelium with regular, rounded, centrally-located nuclei and moderate eosinophilic cytoplasm with no evidence of mitosis, necrosis, or anaplasia[14]. The pathologic papillary growth pattern could explain the cauliflower- and mottled-like appearance radiologically. CPPs are generally regarded as benign tumors with the aforementioned typical radiologic and pathologic findings. However, CPPs sometimes exhibit unusual radiologic findings, which could give the difficulty to diagnose preoperatively. In this study, we reviewed the clinical, radiologic and pathologic findings on our experience.

MATERIALS AND METHODS

Clinical characteristics of 10 patients

From June, 2006 to June 2011, we operated on 10 patients with

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CPPs per the World Health Organization classification[17] and retrospectively analyzed the data. The study group included 5 male and 5 female patients (age range, 3 months to 58 years : median, 29 years). The patients were divided into two groups depending on age : pediatric (<18 years of age) and adult patients (>18 years of age). There were 3 pediatric and 7 adult patients. The distribution of tumor sites was as follows : 2 lateral ventricles and 1 third ventricle for pediatric patients; 6 fourth ventricle and 1 suprasellar location for adults. Nine patients underwent a gross total resection (GTR) and one patient had a subtotal resection. The initial presentation, post-operative complications, and recurrences were investigated.

Radiologic and pathologic findings

We analyzed the radiologic findings on computed tomography (CT) and magnetic resonance images (MRI), as follows : size; location; signal; calcifications; shape; enhancement; and hydrocephalus. We focused on homogeneous contrast enhancement with a mottled-like appearance on the edge of the tumor, which characterized the typical radiologic findings. Histologically, all patients had benign CPPs, which showed papillary structures lined by cuboidal epithelium with no evidence of mitosis, necrosis, and anaplasia; the Ki-67 labeling index was less than 0.5% (Fig. 1A, B). Depending on the pathologic growth pattern, we divided tumors' group into the following three groups : typical papillary; combined papillary and solid; and combined papillary and tubular. Typical papillary growth indicates papillary structures lined by cuboidal epithelium (Fig. 1A). The papillary and solid growth occurs when the papillary structures are combined with a solid growth pattern (Fig. 1C), and papillary and tubular growth occurs when the papillary structures are combined with a tubular growth pattern like glandular structures (Fig. 1D). We also investigated the immunopositivity for cytokeratin (CK) 7 and CK20.

RESULTS

Clinical results

Three pediatric patients were enrolled in the current study (3 months, 8 months, and 2 years of age). All of them had evidence of preoperative hydrocephalus and they complained of vomiting, irritability, and stuporous mentality, respectively. Among these, the tumor on the trigon of the lateral ventricle was operated on via a transcortical approach through the middle temporal gyrus. The others in the foramen of Monro of the lateral ventricle and third ventricle were operated on via a transcallosal approach; the tumors were totally removed. Postoperatively, subdural hygroma had occurred in two patients, which were associated with preoperative severe hydrocephalus and had been shown during the follow-up period.

Seven adult patients were enrolled in the current study. The median age was 49 years (range, 24-58 years). They complained of headaches (n=4), and seizures, dizziness, and vomiting (n=1 each), respectively. Three patients showed hydrocephalus-related symptoms. The tumors were operated on via a midline suboccipital approach and completely removed with the exception of the tumor which was a hard, calcified mass and adherent to the brainstem. Postoperatively, two patients with large masses had injuries of the brainstem and underwent shunt procedures for aggravation of hydrocephalus. One patient had transient vocal cord palsy and recovered to a Karnofsky performance scale (KPS) of 90. The other patient had a permanent brainstem and cerebellar injury, and a KPS of 60.

The mean duration of follow-up was 21.3 months (range, 4-47.8 months). There was no recurrence after initial treatment.

Radiologic and pathologic findings

Three pediatric CPPs were located in the trigon of the lateral ventricle, the foramen of Monro of the lateral ventricle, and the third ventricle, respectively. On CT, all of the tumors were isodense and there was no association with calcifications. On MRI, the tumors showed iso-signal intensity on T1- and T2-weighted images and a median 3.2 cm sized mass (range, 2.7-4 cm) with homogeneous enhancement and a mottled-like appearance. All of the tumors were associated with hydrorocephalus. Pathologically, all of them showed a papillary growth pattern.

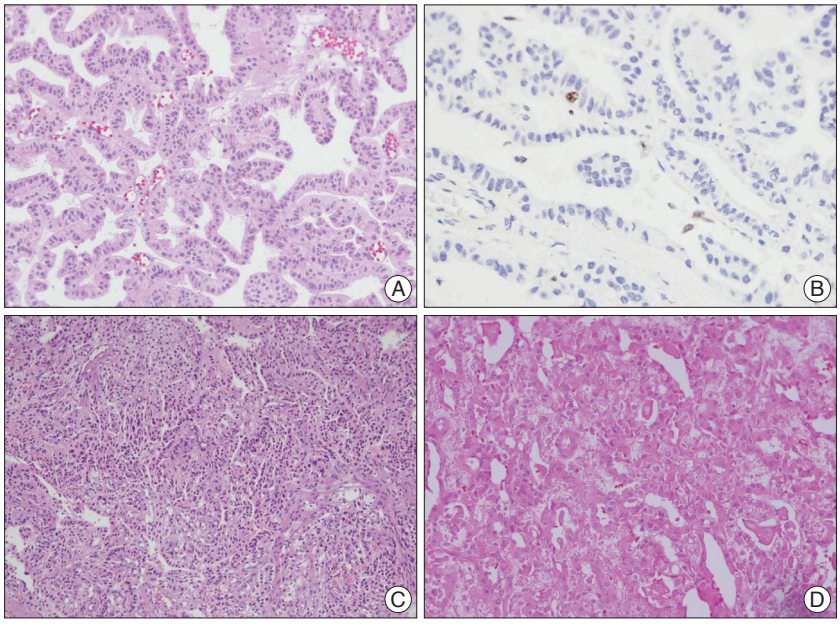

Fig. 1. The pathologic growth pattern. A : Typical papillary growth with papillary structures lined by cuboidal epithelium. There is no evidence of mitosis, necrosis, and anaplasia. B : Ki-67 labeling index is <0.5%. C : The solid growth pattern is shown. D : The tubular growth pattern is shown like glandular structure.

Seven adult CPPs were located posterior to the medulla of the fourth ventricle (Fig. 2A) in 3, posteriorly to the pons in 3 and suprasellar area one. On CT, all tumors showed isodense signal. Among these, four tumors were associated with calcifications. On MR T1-weighted images, four tumors had iso-signal intensities and three tumors had hypo-signal intensities. On T2-weighted images, the four tumors had iso-signal intensities, two tumors had hyper-signal intensities, and one tumor had a hypo-signal intensity. The signal characteristics were neither specific nor characteristic such non enhancing T1 and T2 MR images. Size varied from 0.5-4.2 cm in size. One small (1.2 cm) and two large-sized tumors (3.5 and 4.2 cm) were associated with hydrocephalus. On contrast administration, 4 lesions enhanced homogeneously and showed typical mottled like appearance on the edge of the tumors. On the contrary to these, there were three lesions in our study that showed the disappearance of mottled like appearance and heterogenous enhancement (Fig. 2B, C , D). In 4 lesions showing typical radiologic findings, their pathologic findings revealed typical papillary growth pattern. In contrast, 3 lesions showed unusual radiologic findings : combined papillary and solid growth in two and papillary and tubular growth in one. On the immunohistochemial staining, the tumors showed various CK7/CK20 staining patterns : CK7(+)/CK20(+) in one out of 10 cases), CK7(+)/CK20(-) in 5 and CK7(-)/CK20(-) in 4.

DISCUSSION

CPPs account for 0.3-0.7% of all intracranial tumors, 10-20% of brain tumors presenting during the 1st year of life, and 2-6% of tumors in childhood[12]. These tumors can develop anywhere choroid plexus tissue exists. Generally, the lateral ventricle is the most common site (50% of cases) for these tumors, followed by the fourth ventricle (40%), and the third ventricle (5-9%)[10]. The site at which CPPs develops is closely linked to patient age. CPPs in younger and adult patients tend to develop primarily in lateral and fourth ventricular lesions, respectively[11]. The mean age at the time of presentation in our series was 27.4 years (range, 3 months to 59 years). All patients of young age had supratentorial CPPs, whereas patients over 18 years of age had fourth ventricle and suprasellar CPPs. The most common presentation of CPPs is related with the signs due to increased intracranial pressure (ICP)[2,9,15]. Because these tumors produce cerebrospinal fluid (CSF) in amounts far exceeding the normal average, intraventricular pressure was increased and it causes increased ICP. Hydrocephalus is also well-accompanied because these tumors obstruct the CSF flow mechanically and disturb the CSF absorption at the subarachnoid space secondary to hemorrhage or proteinaceous material. In our cases, there were 6 patients (3/3 in pediatrics and 3/7 in adults) who complained of hydrocephalus-related symptoms.

CPPs showed iso- to hyper-dense intraventricular masses without brain invasion and intense enhancement on CT findings[12,17]. Calcifications were observed in 24 to 50% of CT images. On MR images, CPPs were iso- to hyper-intense on T1-weighted images and mixed, hypo- to hyper-intense on T2-weighted images[1,13]. The most important typical radiologic finding in CPPs is the intense contrast enhancement, which is homogeneous and shows typical mottled- and cauliflower-like appearances on

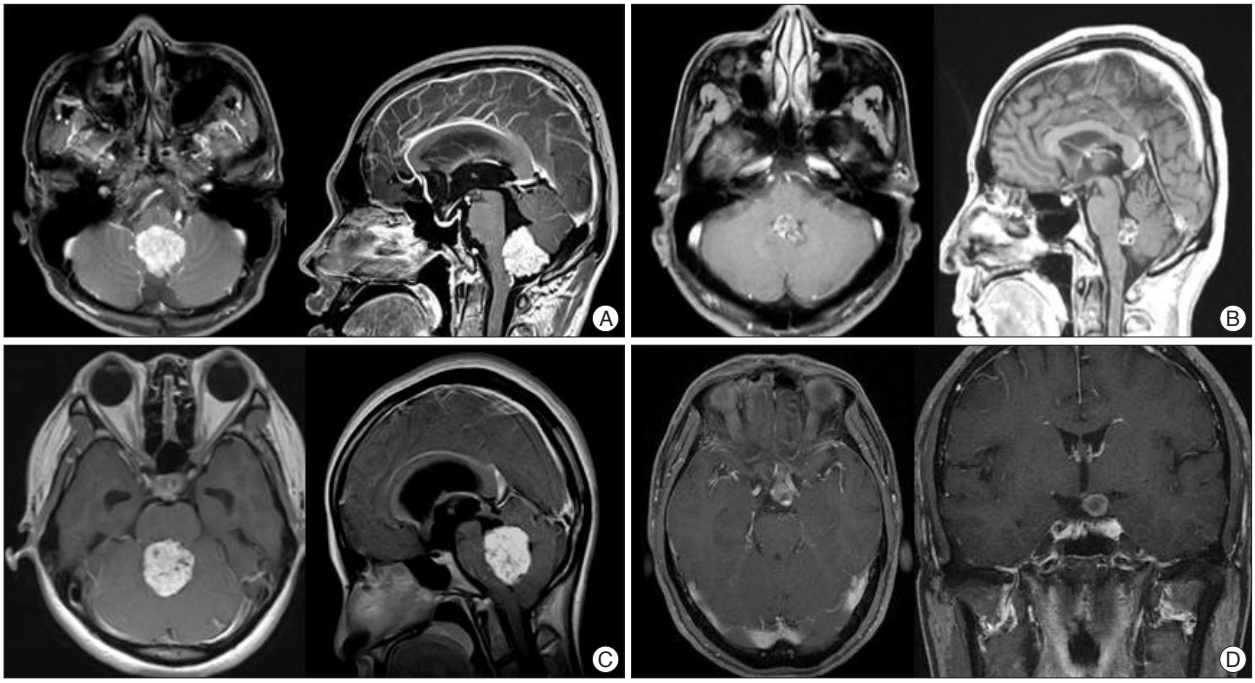

Fig. 2. Radiologic findings of CPPs. The tumors are located posteriorly to the medulla (A), which show homogenous enhancement and typical mottled like appearance on the edge. The tumors located posteriorly to the pons (B and C) and the suprasellar tumor (D) show the heterogeneous enhancement and the disappearance of mottled like appearance.

the edge of tumors. But all of CPPs do not show these finding. Some minor cases show heterogenous enhancement feature with disappearing the typical mottled like appearance. In our all 10 cases, there were 4 patients with calcified lesions on CT images. On T1- and T2-weighted MR images, the CPPs had variable signal intensities. Seven patients (3/3 in pediatrics and 4/7 in adults) showed typical MR findings which are homogeneous enhancement and mottled-like appearances. There are three patients who had the heterogeneously enhanced mass without mottled-like appearances, which gave the difficulty to diagnose preoperatively. The typical histopathologic pattern of CPPs is the papillary growth of cuboidal epithelium with regular, rounded, centrally-located nuclei and moderate eosinophilic cytoplasm with no evidence of mitosis, necrosis, or anaplasia[14,16]. On the immunohistochemical staining, the most common CK7/CK20 combination is CK7-positive and CK20-negative in 74%, but the other three combinations are also possible[5]. In our study, the tumors showed various CK7/CK20 staining patterns and the common pattern was CK7-positive and CK20-negative in 50%. CPPs can show unusual histological features; solid or tubular growth, oncocytic alterations, melanin deposition, calcification, ossification, and xanthograulomatous reaction and mitotic activity is the sole atypical features independently related with the tumor recurrence[3,7,8]. In our cases, we found three unusual histopathologic growth pattens. Two cases showed the papillary structures combined with a solid growth pattern and one case had the papillary combined with tubular growth pattern. Although these three cases showed the unusual histopathologic findings, they did not have any mitoses and displayed very low Ki-67 proliferation index. These three CPPs with unusual pathologic pattern showed the unusual radiologic findings. Even if the cases were small, we carefully suggested the unusual radiologic findings might be explained with combined growth patterns.

GTR is the first choice in the treatment of CPPs[1,4,19] and is considered the most important prognostic factor for CPPs. Because CPPs usually exist in deep brain lesion, surgical treatment targeting the gross total resection sometimes accompanies the adjacent brain parenchymal injuries and causes the complications associated with the site of origin. In our study, pediatric CPPs with preoperative severe hydrocephalus accompanied postoperative hygromas. Two cases adult CPPs with large sized tumor in the fourth ventricle had occurred accompanied brainstem injury in our study and required ventriculoperitoneal shunt procedures for hydrocephalus. Surgical complications occurred in the cases with the severe hydrocephalus or large masses of fourth ventricle.

CONCLUSION

CPPs could have unusual radiologic findings, which preoperatively give the difficulty to be differentiated from other tumors. CPPs with unusual radiologic findings showed the combined pathologic growth patterns.

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