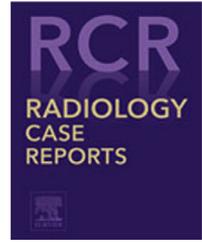


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Case Report

Posterior fossa choroid plexus papilloma with focal ependymal differentiation in an adult patient: A case report and literature review

Shu Matsushita, MD^{a,*}, Taro Shimono, MD, PhD^a, Takeo Goto, MD, PhD^b, Satoshi Doishita, MD, PhD^a, Yuko Kuwae, MD^c, Yukio Miki, MD, PhD^a

^a Department of Diagnostic and Interventional Radiology, Osaka City University Graduate School of Medicine, 1-4-3 Asahi-machi, Abeno-ku, Osaka 545-8585, Japan

^b Department of Neurosurgery, Osaka City University Graduate School of Medicine, Osaka, Japan

^c Department of Pathology, Osaka City University Graduate School of Medicine, Osaka, Japan

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ABSTRACT

Choroid plexus papillomas (CPPs) are rare neoplasms classified as World Health Organization grade I tumors. CPPs containing other tissues have occasionally been documented in the literature. However, few of these previous reports have provided clinical and radiological information. We herein report a case of a posterior fossa CPP with focal ependymal differentiation in a 42-year-old woman who presented with a 6-month history of progressive headache. Preoperative radiological images showed a hypervascular tumor protruding into the left foramen of Luschka with perilesional edema. Gross total resection of the tumor was performed. Histopathological examination revealed that the tumor was composed of papillary structures. Immunohistochemical staining of glial fibrillary acidic protein was focally positive around the capillaries, which was suggestive of “perivascular pseudorosette” formation. Our case showed similar imaging appearances as those of CPP; thus, it seems difficult to distinguish CPP with versus without ependymal differentiation by clinical and radiological features alone. The clinical significance and pathogenesis of ependymal differentiation in CPP remain unclear, and further case reports are required.

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Introduction

Choroid plexus papillomas (CPPs) are rare, indolent neoplasms classified as World Health Organization grade I

tumors [1]. To date, CPPs containing other tissues have occasionally been documented in the literature [2,3]. However, these reports have mainly focused on pathological features; few have provided clinical and radiological information. We herein report a very rare case of a posterior fossa CPP

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* Corresponding author.

E-mail address: shumatsu@med.osaka-cu.ac.jp (S. Matsushita).

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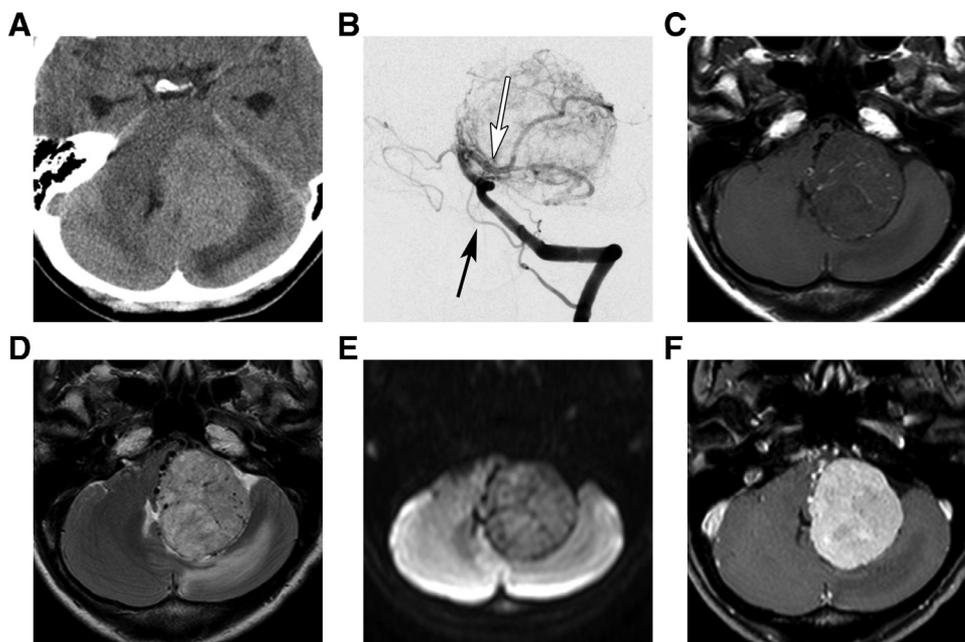


Fig. 1 – (A) Axial head computed tomography at level of the pons reveals a spontaneously dense tumor compressing the brainstem and left cerebellar hemisphere with obstructive hydrocephalus. **(B)** Left vertebral angiogram shows feeding vessels of the vascular-rich tumor from the left anterior inferior cerebellar artery (white arrow) and posterior inferior cerebellar artery (black arrow). **(C-F)** Axial brain magnetic resonance images show a tumor epicentered in the left foramen of Luschka with perilesional edema. The lesion is hypointense on **(C)** T1-weighted images and nonhomogeneously hyperintense on **(D)** T2-weighted images. Intratumoral and peritumoral flow voids are also seen. **(E)** Diffusion-weighted imaging demonstrates hypointensity (high ADC). **(F)** After administration of contrast medium, the lesion shows strong homogeneous enhancement.

with focal ependymal differentiation in an adult patient and provide detailed clinical information, including preoperative radiological images.

Case presentation

A 42-year-old woman presented with a 5-year history of gait disturbance and a 6-month history of progressive headaches and clumsiness of her left arm. She had no other significant medical or surgical history. Physical examination showed left cranial nerve VI, VII, and VIII palsy and left cerebellar ataxia. Audiometry revealed left sensorineural hearing loss.

Head computed tomography demonstrated a high-density posterior fossa mass compressing the brain stem and the left cerebellar hemisphere with hydrocephalus (Fig. 1A). Vertebral angiography findings revealed a vascular-rich tumor with visible feeding vessels from the left anterior inferior cerebellar artery and posterior inferior cerebellar artery (Fig. 1B). Magnetic resonance imaging (MRI) showed a 48-mm heterogeneous lesion protruding into the left foramen of Luschka with

perilesional edema. The lesion was slightly hypointense on T1-weighted images and hyperintense on T2-weighted images compared with the cerebellar white matter and demonstrated hypointensity on diffusion-weighted images (high apparent diffusion coefficient [ADC]) (Fig. 1C-E). Intratumoral and peritumoral flow voids were also noted. After administration of gadolinium, the lesion showed strong and homogeneous contrast enhancement (Fig. 1F). No evidence of drop metastasis was revealed in either intracranial or spinal MRI.

To prevent intraoperative massive bleeding, three-stage coil embolization of the feeding arteries was performed in advance. However, after the second embolization, the patient developed left hemiparesis and dysphagia due to a brain stem infarction. The patient underwent gross total surgical resection of the tumor via a suboccipital approach in the right semiprone park bench position. With a cortical incision of the left cerebellar hemisphere, the tumor arising from the fourth ventricle showed adhesion to the frontal wall of the fourth ventricle with involvement of left cranial nerves IX, X, and XI. Because these cranial nerves were destroyed, they were resected with the tumor.

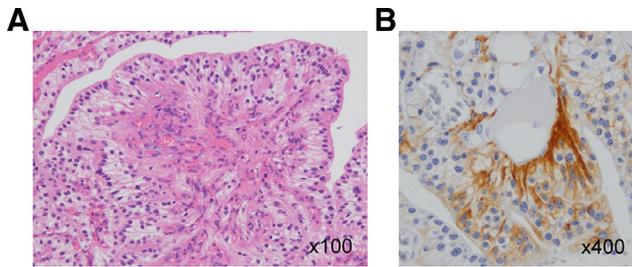


Fig. 2 – Histopathological examination of the resected specimen. (A) Hematoxylin and eosin staining shows a papillary neoplasm with cuboidal to columnar epithelial cells surrounding a fibrovascular stalk. (B) Glial fibrillary acidic protein is focally positive at the elongated fibrillated cytoplasmic processes that have formed around the capillaries, suggestive of “perivascular pseudorosette”.

Histopathological examination with hematoxylin and eosin staining revealed a papillary neoplasm with cuboidal to columnar epithelial cells surrounding a fibrovascular stalk (Fig. 2A). No mitotic activity was present. Immunohistochemical staining was diffusely positive for CAM 5.2. Glial fibrillary acidic protein (GFAP) was focally positive at the cytoplasmic processes formed around the capillaries, which was suggestive of “perivascular pseudorosette” formation (Fig. 2B). Staining for S-100 protein and epithelial membrane antigen were negative. These findings were consistent with a diagnosis of choroid plexus papilloma with focal ependymal differentiation.

Postoperatively, the patient’s left hemiparesis and dysphagia persisted. She was transferred to another hospital for rehabilitation 2 months after the operation. Her neurologic function gradually improved, and she returned home 10 months after surgery with recovery of activities of daily living.

Discussion

Focal ependymal differentiation of CPP has seldom been described in the literature. In 1981, Rubinstein and Brucher initially reported nine cases of CPPs exhibiting focal GFAP positivity in the tumor cells [4]. Since then, several reports have documented mixed tumors composed of CPP and other tissues such as ependymoma and hemangioblastoma [2,3]. Focal GFAP positive cells are often seen in CPP without ependymal differentiation [5]. Perivascular pseudorosette formation is a characteristic morphological feature of ependymoma and GFAP tends to be positive in these areas [6]. Our case, however, is unique and interesting because the ependymal differentiation was suggested not only by GFAP positivity but also by the existence of perivascular pseudorosette, a feature of ependymoma.

During development, primitive ependymal cells give rise to the ependyma lining the brain ventricles and spinal cord central canal [7]. While conventional ependymal cells develop,

these cells undergo further specialization at certain sites to form the choroid plexus and other specialized ependyma of the circumventricular organs (SECO), which comprise the pineal gland, area postrema, subfornical organ, subcommisural organ, median eminence, infundibulum, and organum vasculosum of the lamina terminalis [8]. Thus, choroid plexus cells and conventional ependymal cells have a similar embryologic background.

The reason why several tissues can be detected within a CPP is still unclear. Two hypotheses have been proposed to explain the pathogenesis of CPP containing other tissues. The first is that when pia and blood vessels invaginate into the ventricle during development of the choroid plexus, other mesenchymal components and adjacent neuroglial tissue can be included, and they may differentiate into certain types of tissues [2]. The other is that the multipotent primitive cells of the circumventricular organs, which are precursor cells of SECO, might differentiate into diverse cell lineages when they form a tumor [2].

Our case exhibited typical clinical and radiological features of CPP, which clinically occurred in the fourth ventricle, although the pathological findings were not typical. CPPs of the fourth ventricle are thought to be derived from embryonic choroidal remnants or normal choroid plexus protruding into the foramen of Luschka [9,10]. Infratentorial CPPs are evenly distributed among all age groups, while 80% of supratentorial CPPs are found in children [11]. Our patient was a 42-year-old woman. Patients with CPPs of the fourth ventricle typically present with signs and symptoms of increased intracranial pressure including headache, papilledema, nausea, vomiting, cranial nerve deficits, gait impairment, and seizures because of CSF overproduction or obstruction of CSF outflow [12]. These lesions are well vascularized, enhance with contrast, and occasionally contain cystic features on imaging. On MRI they are homogeneous or heterogeneous tumors with a cauliflower-like appearance and accompanying flow voids are common. The ADC of CPP is higher than that of malignant brain tumors [13]. A recent study showed that a lower ADC and larger tumor volume predict a poorer prognosis [14]. Our case demonstrated strong homogeneous enhancement with accompanying flow voids and hypointensity on diffusion-weighted imaging (high ADC), suggesting a benign nature.

To the best of our knowledge, only 2 reports have demonstrated CPP containing other tissues including ependymal differentiation with clinical and imaging features. The first case involved a 43-year-old man who presented with a 2-year history of postural vertigo, occipital headache, and temporary diplopia [3]. Brain MRI showed a tumor located in the fourth ventricle with strong enhancement. The tumor was grossly totally resected; however, the patient developed a coma because of substantial intraoperative bleeding. The other case involved a 5-year-old girl who presented with a 1-week history of fever and numbness of the right lower limb [2]. Brain MRI with gadolinium revealed a heterogeneously enhancing mass in the occipital horn of the left lateral ventricle with a small cystic lesion adjacent to the mass. It was totally resected, and a follow-up study showed no evidence of recurrence for 5.6 years thereafter. Both of these tumors demonstrated typical

imaging features of CPP, as in our case. Thus, the difference in the imaging appearance between CPPs with and without ependymal differentiation cannot be determined.

Radiologically, the differential diagnoses for CPP of the fourth ventricle in adults include hemangioblastoma, solitary fibrous tumor/hemangiopericytoma (SFT/HPC), and metastatic tumor. In hemangioblastoma, variably sized cysts are present in 50% to 60% of cases whereas approximately 40% of them are solid tumors [15]. Typically, the solid part shows intense enhancement. Quadery and Okamoto reported that the ADC was increased in hemangioblastomas, reflecting rich vascular spaces [16]. SFT/HPC is a mesenchymal tumor and can occur at any age and in various locations. It is usually heterogeneous with areas of low T2 signal intensity that strongly enhance after gadolinium administration. Components with a decreased ADC and elevated peak of myoinositol in magnetic resonance spectroscopy are thought to be valuable diagnostic features of SFT/HPC [17]. Intraventricular metastasis to the choroid plexus is rare and accounts for 0.9% of all brain metastases [18]. The most likely sources of these lesions are renal carcinoma and lung carcinoma [18]. The lateral ventricle is the most common site for metastatic spread, and only 0.4% of all ventricular metastases are located in the fourth ventricle [19]. Because reports of the clinical and radiological features of CPP with ependymal differentiation are lacking, the difference in the imaging appearance between CPPs with and without ependymal differentiation is unclear.

The optimal treatment of CPP with ependymal differentiation is also yet to be determined because of its rarity. Former case reports indicated no recurrence or metastasis after surgical removal of CPPs containing other tissues [2,3], and McGirr et al. reported that the presence of ependymal differentiation did not correlate with either the difficulty of resection or tendency for recurrence [20]. Although the prognosis of this tumor has not been thoroughly discussed, complete resection should be performed as for ordinary CPPs.

In summary, we have herein reported a very rare case of a posterior fossa CPP with focal ependymal differentiation in an adult patient with preoperative clinical information including detailed radiological images, which were consistent with ordinary CPP. The pathogenesis of this tumor is still unclear, but it has been implicated that divergent differentiation is expressed by primitive cells of the circumventricular organs. The clinical significance of ependymal differentiation in CPP remains to be elucidated, and further reports are expected.

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