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Case Report

Atypical Choroid Plexus Papilloma in an Adult

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We present an extremely rare case of the atypical choroid plexus papilloma in an adult which developed at the trigone of right lateral ventricle. A 62-year-old woman presented with the history of intermittent and gradually progressive headache and left side hemiparesis for 6 months. The brain magnetic resonance image showed highly enhanced and well demarcated mass at the trigone of lateral ventricle attached to the choroid plexus. Gross total resection was performed by transcortical approach via the middle temporal gyrus. The tumor was diagnosed as an atypical choroid plexus papilloma. She had no neurologic deficit after the surgery. We report a case of atypical choroid plexus papilloma in adult and introduce newly classified pathologic characteristics of this tumor.

KEY WORDS : Atypical choroid plexus papilloma · Lateral ventricle · Diffusion tensor imaging · Neuronavigation.

INTRODUCTION

The choroid plexus papillomas (CPPs) derive from the choroid plexus epithelium and mainly occur in young children[16]. It has been reported that 70% of the tumor occurs in children and at least 50% presents before the age of two[6]. The choroid plexus tumors are most commonly found in the lateral and fourth ventricles[5,13,15].

The majority of choroid plexus neoplasms are well-differentiated choroid plexus papillomas. Choroid plexus carcinoma (CPC), on the other hand, has several malignant characteristics such as brisk mitotic activity, blurring of the papillary pattern, increased cellularity, necrosis and invasion of surrounding brain parenchyma. The atypical CPP is a newly introduced entity as an intermediate grade in the 2007 World Health Organization (WHO) central nervous system (CNS) tumor classification. This tumor is mainly distinguished from the CPP by increased mitotic activity, 2 or more mitoses per 10 high power fields (HPF) while usually greater than 5 per 10 HPF in CPC[2,7].

We present a case of atypical choroid plexus papilloma which occurred in an adult with a discussion of newly described pathologic characteristics with consideration of related literatures.

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CASE REPORT

A 62-year-old woman presented with headache, dizziness and slowly progressive left side weakness over the 6 months. The patient did not have any past medical history or family history related to brain lesion. Her visual acuity was mildly decreased (0.6/0.4) but visual field was within normal limits. Brain MRI showed a well enhanced $55 \times 47 \times 44$ mm sized mass in the trigone of right lateral ventricle with mass effect compressing adjacent areas on enhanced T1-weighted images, mimicking intraventricular meningioma (Fig. 1A). The tumor had intermediate signal intensity on T2-weighted images with only slight extent of perilesional edema. There was no radiological evidence of hydrocephalus (Fig. 1B). To define the relationship between the tumor and the optic pathways and to select a proper surgical approach, the MR diffusion tensor imaging (DTI) was done and tracking of the optic tract and radiation was performed (Fig. 1C). On the basis of fusion images of tractography and MR imaging for neuronavigation, the transcortical approach was performed via the middle temporal gyrus incision at the site of the least distribution of the optic radiation fibers to minimize the risk of optic pathway injury. Grossly, the tumor was gray-colored and very friable in consistency. Massive bleeding occurred from feeding arteries of the tumor but approach vector direction was towards the arteries

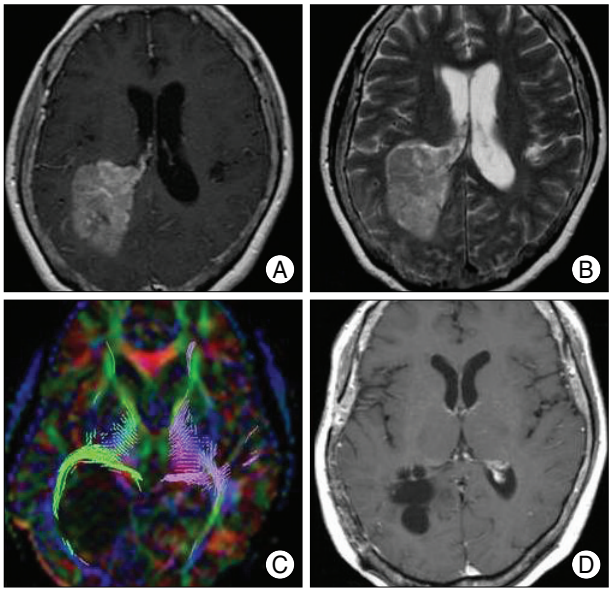

Fig. 1. Preoperative magnetic resonance (MR) image. Heterogenously and relatively well enhanced pattern of the mass is shown at the enhanced T1 weighted axial images (A). Intermediate signal intensity with inner low signal intensity is shown on T2 weighted image (B). MR diffusion tensor imaging tractography of optic radiation is displayed (C). Postoperative brain MR image at 3 months after surgery shows only postoperative change at the periventricular white matter near the trigone without residual tumor on contrast-enhanced T1-weighted axial image (D).

which aided us to control them with ease. The patient awoke from anesthesia immediately after the operation without any newly developed neurological deficit. Microscopically, the tumor revealed a portion of papillary growing pattern which consisted of cuboidal to columnar epithelial cells. Nuclear pleomorphism, increased cellularity with psammomatous calcification and microscopic foci of necrosis were noted. In areas, 2-4 mitoses per HPF were seen (Fig. 2). Based on these findings, the tumor was diagnosed as an atypical CPP. The patient's left hemiparesis was recovered, the visual acuity improved to 0.7/0.6 just after surgery and there was no visual field defects detected at 3 months post-operative ophthalmologic examination. The MR imaging which was performed at 3 months after the surgery revealed no remaining mass (Fig. 1D).

DISCUSSION

CPPs are rare neoplasms representing only 0.4 to 1% of all intracranial tumors. In children, however, they are more common, accounting for 1 to 5% of pediatric brain tumors, and 4 to 12% among patients younger than 1 year old[12]. In childhood, CPPs are usually found in the lateral ventricles and less commonly in the posterior fossa. On the contrary, in adults, the majority of these tumors are found in the fourth ventricle and its lateral recesses. The lateral ventricle

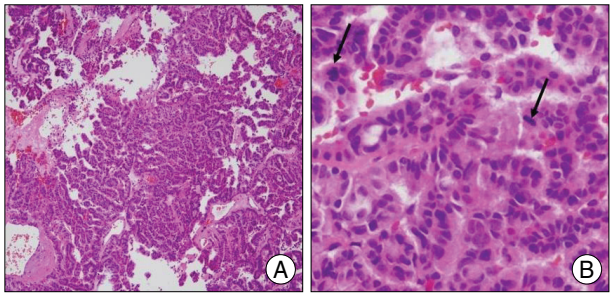

Fig. 2. Hematoxylin and eosin stained sections disclose a portion of papillary growing tumor consists of the cuboidal to columnar epithelial cells. Nuclear pleomorphism and increased cellularity with psammomatous calcification and microscopic foci of necrosis are noted. In areas, 2-4 mitoses (arrows) per HPF are seen. The tumor is diagnosed as atypical choroid plexus papilloma (A : ×100, B : ×400).

location presented in this case is relatively rare in adults[12,13].

The majority of choroid plexus tumors are well differentiated benign neoplasm. Macroscopically, CPPs appear as circumscribed cauliflower-like masses that may attach to the wall of ventricle but are usually well marginated from normal brain structures[11]. Microscopic findings of CPPs typically appear as a single layer of cuboidal epithelial cells surrounding a fibrovascular stalk, arranged in a papillary configuration with finger-like projections. The histological features of CPP characterized by signs of malignancy, including brisk mitoses, nuclear pleomorphism, raised cellular density, obscurity of the papillary growth pattern, and cell necrosis[2,10].

In 2007, WHO introduced an additional entity with intermediate characteristics, 'atypical choroid plexus papilloma', which is primarily distinguished from the CPP by raised mitotic activities. To our best knowledge, this is the first case of atypical CPP in Korean adult, under the confirmed criteria of 2007 WHO CNS tumor classification. In atypical CPP, curative operation is still possible but the recurrence rate appears to be much higher[7]. Jeibmann et al.[3] reported that the recurrence rate in atypical CPP was 6 out of 21 cases (29%). This was higher than the recurrence rate in CPP which was 6 out of 103 cases (6%). He also reported that increased mitotic activity is the only factor to predict recurrence. But, the necessity of preventive adjuvant therapy after gross total resection, such as radiotherapy and chemotherapy, for atypical CPP is still controversial[8,17].

The choice of suitable surgical approach is determined by the location of tumor, vascular supply, and the preference of the surgeons[4,8,14]. Generally, the superior parietal approach is considered for the tumors in the trigone of the lateral ventricles for the sake of safety, which enables prevention of injury to the optic radiation, the motor cortex or the Wernicke's area[1,9]. In this case, because the tumor was located

in the trigone of right lateral ventricle and it expanded laterally, the middle temporal gyrus approach was used to access the nearest route from the cortex. Also, this approach made it easy to control the feeding arteries emerging from the choroid plexus which originated from the posterior parietal artery of the right middle cerebral artery and the anterior choroidal artery. The risk of postoperative visual field defect, typically homonymous hemianopsia caused by the injury of the optic radiation adjacent to the lateral wall of the lateral ventricles following the middle temporal gyrus approach, was minimized by the neuronavigation system fused with the DTI for visualization of the optic radiation. This allowed the cortical incision to be made at the site where no optic radiation-fibers were detected.

CONCLUSION

We experienced a case of rare atypical CPP and achieved successful surgical result using MR DTI without any newly developed neurological sequalae. However, further evaluation and long term follow up are needed to evaluate characteristics, natural history and recurrence of the tumor, which are not clearly verified yet.

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