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Case Report

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J Korean Neurosurg Soc 42 : 475-477, 2007 10.3340/jkns.2007.42.6.475

Spinal Drop Metastasis from a Posterior Fossa Choroid Plexus Papilloma

Choroid plexus papillomas (CPPs) are typically considered as benign tumors, with a favorable long-term prognosis. Drop metastasis of CPP into the spinal subarachnoid space is rare. We report a 42-year-old woman who presented with headache and back pain 6 years after removal of a posterior fossa CPP. Magnetic resonance imaging revealed mass lesions in the lumbosacral subarachnoid space and recurrent intracranial tumor. The lesions were resected and histologically diagnosed was CPP. We consider that CPP can spread via cerebrospinal fluid pathways and cause spinal drop metastasis. Therefore, it is necessary to evaluate the whole spinal axis and to perform periodic follow-up examinations in patients with CPP.

KEY WORDS : Choroid plexus papilloma · Posterior fossa · Spinal · Metastasis · Cerebrospinal fluid.

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INTRODUCTION

Choroid plexus papillomas (CPPs) are typically considered as benign lesions, with a favorable long-term prognosis[5]. These are rare tumors of neuroectodermal origin that account for less than 1% of all intracranial tumors in all age groups[10] and spinal drop metastasis of CPP is even less common. We report a rare case of recurrent CPP with spinal drop metastases.

CASE REPORT

A 36-year-old female was admitted to our institution with a 2-month history of morning headache, dizziness, nausea and vomiting. Neurological examination showed right cerebellar dysfunction and right abducens nerve palsy.

Magnetic resonance imaging (MRI) scan of the brain revealed a well contrast-enhanced cauliflower-like mass in the fourth ventricle extending to the right cerebellopontine angle through the lateral recess (Fig. 1A). The posterior fossa tumor was totally removed (Fig. 1B) and the histopathological diagnosis was CPP (Fig. 2A). The patient's postoperative course was uneventful and she received neither radiotherapy nor chemotherapy.

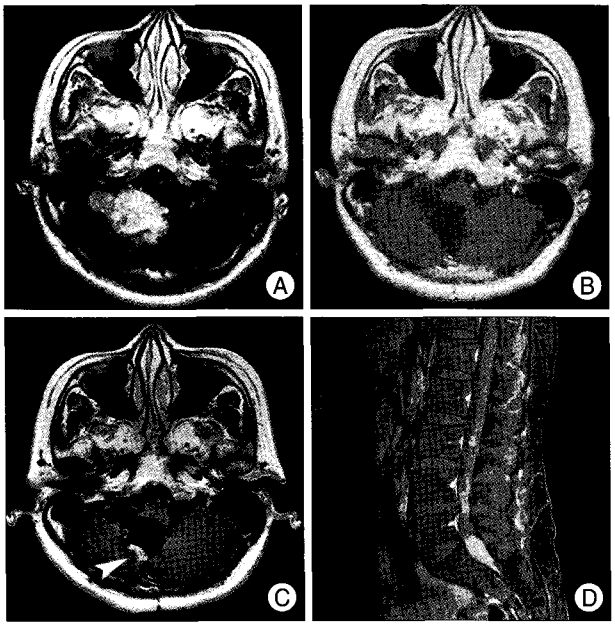

Fig. 1. Preoperative T1-weighted contrast-enhanced magnetic resonance imaging (MRI) of the posterior fossa showing a fourth ventricle mass (A) and postoperative MRI showing total removal of tumor (B). MRI taken 6 years after operation showing a recurrent tumor (C) and spinal drop metastasis at the level of the L3-S1 vertebra (D).

The patient presented 6 years later with gradually aggravated headache and low back pain radiating to the left leg. Neurological examination yielded normal findings, with the exception of numbness on the S1-S2 sensory dermatomes and a positive straight leg raising test on the left side. Brain MRI

• Received : June 1, 2007
• Accepted : November 12, 2007
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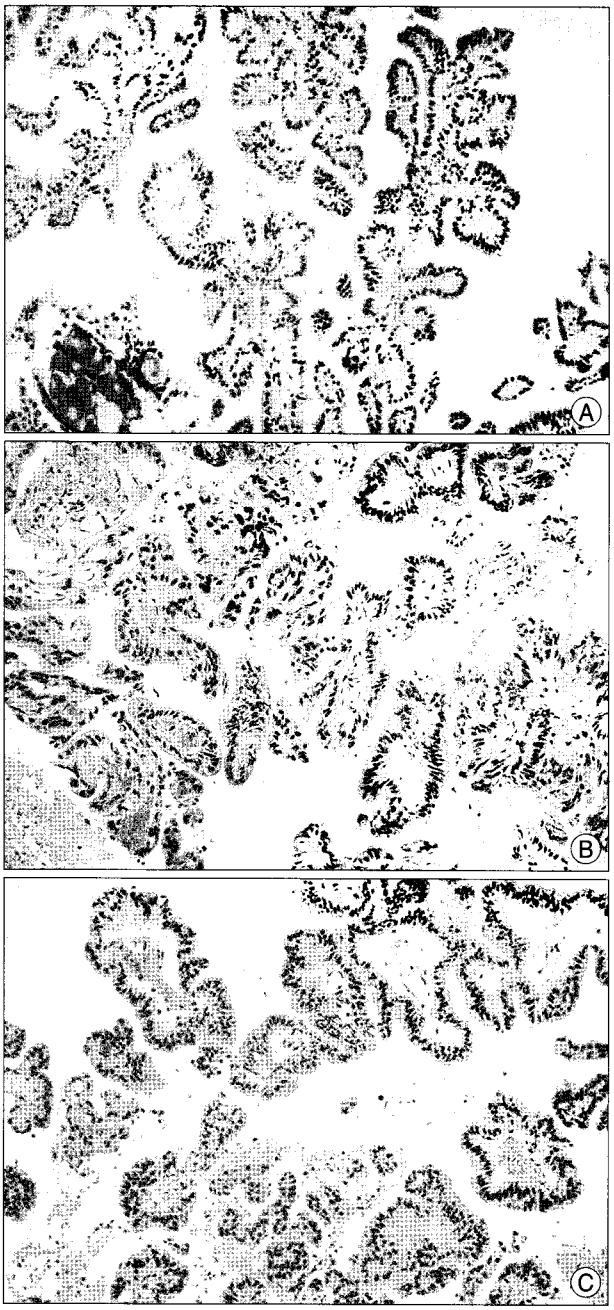

Fig. 2. Photomicrographs of a section of a biopsy specimen obtained from initial posterior fossa tumor (A) showing papillary growing fibrovascular structures with columnar epithelium. Photomicrographs of a section of a biopsy specimen obtained from recurrent posterior fossa tumor (B) and lumbar spinal tumor (C) showing similar histological findings as shown in A. There was no evidence of cellular atypism, necrosis or mitotic activity (H&Ex100).

showed a cystic mass with a small enhanced nodule in the right cerebellum, and strongly enhanced nodules in the posterior and inferior aspect of the dilated fourth ventricular margin (Fig. 1C). Tumor recurrence was suggested. Lumbar spinal MRI showed enhanced tumoral lesions in the subarachnoid space at the level of the L3-S1 vertebra (Fig. 1D). The largest one was at the level of the S1. There was no evidence of involvement in the cervical and thoracic spinal MRI. Firstly, the recurrent cerebellar tumor was totally removed and the histologic diagnosis was CPP. The tumor was composed of a branching papillary structure with a central vascular core. Small calcified nodules were observed in the surrounding tumoral tissue. There was no cellular atypia or mitosis (Fig. 2B). Three months later, L5 and S1 laminectomy was performed and the spinal tumor was subtotally removed. On histopathological examination, the tumor demonstrated papillary proliferation of the choroid plexus epithelial cells without atypism. There was no evidence of necrosis or mitotic activity. Histologic diagnosis was also CPP (Fig. 2C). The patient's postoperative recovery was uneventful and she underwent periodic chemotherapy with CCNU.

DISCUSSION

CPPs are classified as World Health Organization Grade I tumors. Because of the benign nature of these tumors, total surgical resection is typically thought to be curative[18]. The malignant form, choroid plexus carcinoma (CPC), is classified as a World Health Organization Grade III tumor and is characterized by anaplasia, mitosis, nuclear pleomorphism, necrosis and invasion[7]. CPC has an extremely poor prognosis. Although CPCs frequently metastasize, cerebrospinal metastasis in benign CPP are rare[3,13,18]. Some CPPs, termed atypical CPPs, tend to recur and cause meningeal or intracerebral metastasis[1,16]. They have only a few histologic features of malignancy such as increased mitotic activity or increase of the layers of epithelial cells[2]. Our case demonstrates drop metastasis, which originated not from malignant transformation in the primary tumor but from a well-differentiated tumor without malignant histologic features. The pathogenetic mechanism of the spinal CPP may be caused by spontaneous drop seeding of the posterior fossa CPP or by iatrogenic seeding of the initial tumor surgery.

CPPs primarily involve the fourth ventricle in adults, the lateral ventricle in children, and the third ventricle, in order of frequency[9,12,14]. Extraventricular growth of choroid plexus tumors is found in three situations : direct extension of the primary tumor, seeding along cerebrospinal fluid pathways, and development of a primary tumor from the small choroid tuft that normally project outside the foramen of Luschka[8].

The standard treatment for a CPP is complete surgical resection, typically thought to be curative. Surgery can play a role in the treatment of spinal metastatic CPP in several circumstance : First, biopsy can be useful to confirm the diagnosis of CPP metastasis. Second, laminectomies may be performed when spinal cord compression becomes significant to the point that symptoms are not tolerated by the patient.

Third, surgical resection of a metastatic lesion can be a definitive treatment or can alleviate symptoms[4]. Radiation therapy has been used in cases of CPP with subtotal resection, and stereotactic radiosurgery has been reported when a patient refuses surgery for treatment of a small CPP[6,10]. Chemotherapy in CPP is limited and there is no consensus on its impact in the management of CPP. Valencak et al.[17] reported the therapeutic efficacy of CCNU in recurrent CPP and extensive spinal seeding. There are two case reports suggesting that CCNU may be effective in CPP[11,15].

CONCLUSION

A case of recurrent CPP with spinal drop metastases is presented. This rare case serves as an important reminder to search the whole spine in patients with CPP, even if asymptomatic, and to periodically perform follow-up examinations after primary CPP resection in order to rule out drop metastasis and recurrence.

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