CASE REPORT
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Choroid Plexus Carcinoma in an Adult

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Choroid plexus carcinomas are extremely rare in adults. They can behave aggressively and their optimal management is uncertain. A 35-year-old woman was admitted with an episode of loss of consciousness. Magnetic resonance imaging showed a homogeneously enhancing mass in the trigone of the right lateral ventricle. Detailed examinations found no evidence of an extraneural primary focus. She underwent total removal of the tumor. Pathological diagnosis was confirmed as a choroid plexus carcinoma. She is doing well eight months after surgery.

KEY WORDS : Choroid plexus carcinoma · Adult · Lateral ventricle · Total removal.

Introduction

Choroid plexus tumors(CPTs) are account 0.4~0.6% of central nervous system(CNS) tumors[1,2,4,11-13]. Choroid plexus carcinomas(CPCs) are constituting 20~30% of CPTs[11,12]. Noticeably, CPCs are extremely rare in adults. Geerts et al. identified 231 cases of CPC in the world literature, of which only 16 were in the non-pediatric population[7].

The diagnosis of CPC is difficult even by histological examination, because primary CPC cannot always be differentiated from metastatic carcinoma, which may originate from extraneural tumors[9,17]. CPC is known to be highly invasive in nature with extremely poor prognosis, and its best treatment protocol is yet to be clarified[1-7,9-17].

We present a case of CPC of the lateral ventricle in a 35-year-old woman with literature review.

Case Report

A 35-year-old woman, who had a history of progressive headache and an episode of loss of consciousness for three years ago, was admitted to our hospital. General physical examination was unremarkable. Neurological examination was entirely within normal limits.

Computerized tomography scan showed a mass in the trigone of the right lateral ventricle, which was homogeneously enhanced by intravenous injection of contrast material and surrounded laterally by non enhancing multilocular cysts (Fig. 1). On magnetic resonance imaging(MRI), the mass was isosignal intensity on T1-weighted images and T2-weighted images showed a wide, diffusely hypersignal intense area. Gadolinium injection brought about homogeneous enhancement of the tumor. The laterally located surrounding cystic areas didn't enhance (Fig. 2). Selective right carotid angiography revealed a small tumor blush, which was fed by the posterior cerebral artery (Fig. 3).

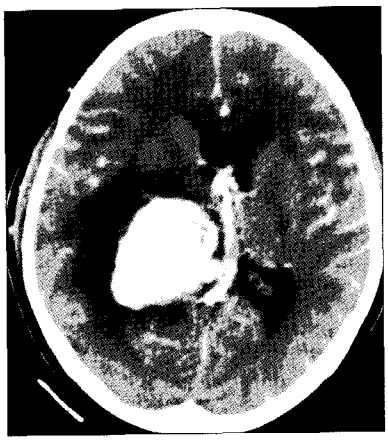

Fig. 1. Enhanced computerized tomography scan at initial presentation showing a round to ovoid, well-enhancing mass in the trigone of the right lateral ventricle, which is surrounded by multiple cysts. Mild hydrocephalus is seen.

The patient underwent an operation with neuronavigation system. A right temporoparietal craniotomy was carried out. The parietal cortex overlying the tumor was markedly atrophic. Following a right parietal corticial incision, the tumor was exposed. It was grayish purple in color, highly vascularized

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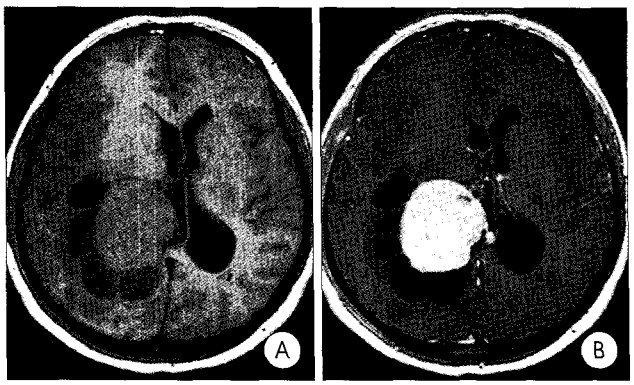

Fig. 2. Pre-operative T1 weighted axial magnetic resonance(MR) image (A) shows an iso signal intensity mass in the right ventricle, which is surrounded by multi-lobular cysts. After Gadolinium(Gd) administration, T1 weighted axial MR image (B) reveals homogenous enhancement of the mass.

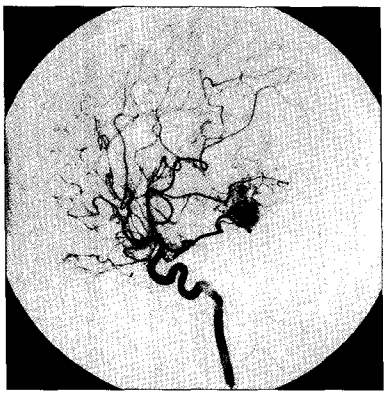

Fig. 3. Selective right internal carotid angiography demonstrates tumor blush, which is supplied by dilated posterior cerebral artery.

and adhesive to surrounding brain. As satellite lesion, multilocular cysts were present lateral to the tumor, of which content was cheesy. Gross total removal of the tumor was performed.

Histological examination revealed focal papillary architecture, evident pleomorphism, and invasion of tumor cells into the surrounding brain. Immunohistochemical examination demonstrated that tumor cells were reactive for S-100 protein, cytokeratine and vimentine, but not reactive for glial fibrillary acidic protein, alpha fetoprotein (Fig. 4). Ki-67 proliferating index was 10%. The tumor was diagnosed as a CPC.

Postoperative course was uneventful. Eight months after the operation, the patient was doing well and without any sign of recurrence on follow-up MRI (Fig. 5).

Discussion

CPTs are thought to arise from neuroectoderm and vary widely from benign papilloma to highly anaplastic carcinoma[13].

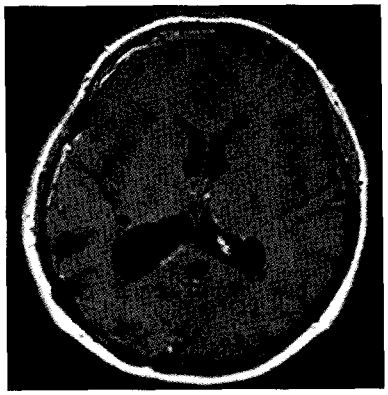

Fig. 5. Eight months after operation, Gd enhanced axial T1 weighted MR image shows that the tumor is completely removed and there is no sign of recurrence.

In the literature, most cases of primary CPC occur predominantly in the lateral ventricle in children without any sexual predilection. In the adult, the fourth ventricle seems to be the preferred location[2,3,10,13].

A review of cases of CPC show that almost all patients manifested symptoms of hydrocephalus. The hydrocephalus associated with both the benign and malignant conditions may be secondary to obstruction and hyper secretion of cerebrospinal fluid[1,2].

The differential diagnosis of CPC should include choroids plexus papilloma(CPP), atypical CPP, papillary ependymoma, and metastatic carcinoma[3]. It is difficult to differentiate CPC from CPP by clinical symptoms and signs. For the diagnosis of CPC, the histological criteria prompted by Vinken and Sloof[16] should be met. These include 1) infiltrative and destructive growth, 2) abundant cellularity, 3) pleomorphic nuclei and cell type, 4) mitoses, 5) proliferation of vascular structures, 6) necrotic foci, and 7) eradication of boundaries between stroma and parenchyma. Papillary ependymoma is generally less vascular and usually has a fibrillary neurologlial stroma and blepharoblasts in the epithelial cells, while choroidal neoplasm would not[13]. Quite a few metastatic carcinomas of the choroid plexus were misdiagnosed as primary CPC. The importance of distinguishing primary CPC from metastatic tumors in emphasized[1,2,13,17].

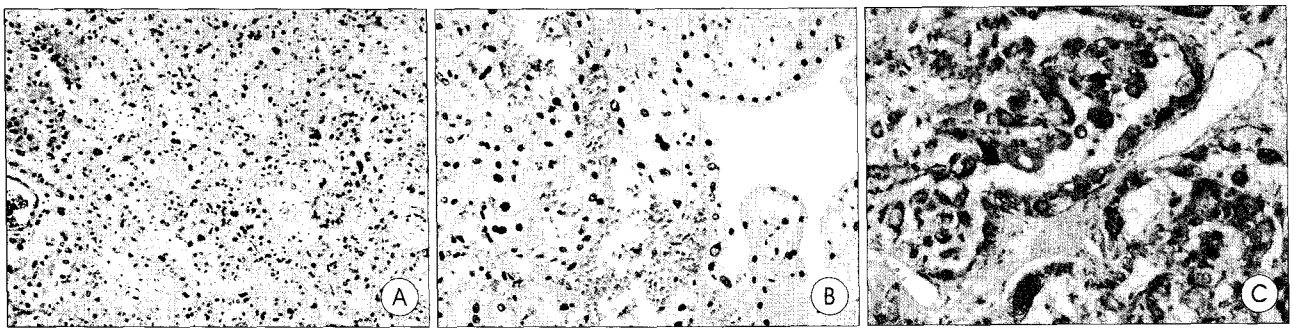

Fig. 4. Photomicrographs of choroid plexus carcinoma. Low magnification (A) shows tubulopapillary arrangement of large pleomorphic epithelial cells having abundant eosinophilic cytoplasm (H&E, x100). High magnification (B) demonstrates a transitional area from the non neoplastic choroid plexus to the carcinoma (H&E, x200). The tumor cells are strongly positive for cytokeratin immunostaining (C).

The results of immunohistochemical study of CPC vary widely. Gottschalk et al.[8] reported that positive immunoreactions ranges in CPP or CPC are 83~100% for cytokeratin, 40~94% for S-100 protein, 16~88% for vimentin, 69~71% for epithelial membrane antigen, 21~100% for neuron specific enolase, 67~100% for prealbumin, and 0~50% for carcinoembryonic antigen. Glial fibrillary acidic protein is often negative[9]. According to Felix et al.[6], S-100 protein has a higher association with CPP than with CPC and has a prognostic significance. According to Inamura et al.[10], high serum levels of CA 19-9 decreased rapidly after removal of the tumor and tumor tissue specimens obtained at surgery showed intense immunohistochemical reactivity for this antigen.

On MRI, CPCs are frequently much less homogeneous due to central necrosis, hemorrhage and cyst formation, with invasion of adjacent brain parenchyma and peritumoral vasogenic edema in the surrounding white matter. A frequent finding is the presence of curvilinear signal void representing enlarged intra tumoral blood vessels[15].

The prognosis of CPC is generally reported to be poor. In the management of CPC, gross total removal is the goal and even in cases of localized recurrence, additional surgical resections should be performed first if the patient's clinical condition permits. Carpenter et al.[2] recommended that all patients with CPC shoud have radiotherapy following surgery. In infants with a residual CPC after surgical resection and metastatic spread, as radiotherapy is associated with a certain risk of neurological morbidity, chemotherapy is preferred to radiotherapy[12,17]. As the response to chemotherapy is limited, its role is mainly controlling the tumor until radiation therapy is feasible in some cases or it is used as a measure for the reduction of vascularity and size before an attempt of total resection of a large tumor[10].

Dohrmann and Collias[3] stated that the tumor is more aggressive in children comparing the mean survival of nine months after diagnosis in children with the mean survival of three and a half years in adults. Ellenbogen et al.[5] reported all patients with subtotal removal die in spite of adjuvant therapy, while only three of nine cases with gross total removal had recurrences.

Conclusion

In adults, CPC is a very rare neoplasm. Total surgical excision is the main predictor of long-term survival, so the achievement of total excision should be the goal of treatment. But the low incidence of this tumor has prevented the establishment of the optimum treatment. We suggest to include CPC in the differential diagnosis in patients with intra ventricular tumors.

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