Extra-axial Ependymoma

—Case Report—

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Abstract

A 13-year-old boy presented with a very unusual ependymoma extending extra-axially. Computed tomography demonstrated a tumor with a cyst and calcification adjacent to the dura and extending over the right occipital and parietal lobes. The cyst wall and solid tumor were enhanced postcontrast. Magnetic resonance imaging revealed that the solid tumor was isointense on T₁-weighted images and a mixed iso- and high-intensity on T₂-weighted images. The solid tumor and tissue surrounding the cyst were enhanced markedly by gadolinium-diethylenetriaminepenta-acetic acid. Sagittal and coronal images demonstrated a multilocular tumor shadow. Cerebral angiography demonstrated a tumor fed by a posterior branch of the right middle meningeal artery but no feeders from the internal carotid and vertebral arteries. The tumor was removed en bloc. The histological diagnosis was clear cell-type ependymoma.

Key words: ependymoma, extra-axial extension, magnetic resonance imaging, computed tomography

Introduction

Ependymoma is the most common pediatric brain tumor after astrocytoma, medulloblastoma, craniopharyngioma, and germ cell tumor,^(12,13) accounting for approximately 6% of all pediatric intracranial brain tumors.³⁾ Ependymoma usually originates from ependymal cells in the cerebral ventricle and is located in the ventricular system.^(2,5,12,13) Compression of the cerebral ventricles and extension to the periventricular white matter are commonly observed.⁹⁾ However, ependymoma extending extra-axially is extremely rare. We describe a patient with ependymoma mainly localized extra-axially and discuss the tumor extension.

Case Report

A 13-year-old boy presented with early morning headache with nausea and vomiting persisting since

August, 1991. He visited a local physician on May 7, 1992 and was referred to our clinic on May 14.

Physical and neurological examinations on admission revealed no abnormalities except for slight headache. Plain x-ray films of the skull demonstrated a calcification in the right occipital area. Computed tomography (CT) demonstrated an isodense mass lesion with a calcification and a cystic low-density area. The isodense area suggested solid tumor adjacent to the right parietal bone. The solid part was highly enhanced postcontrast (Fig. 1). T₁-weighted magnetic resonance (MR) imaging demonstrated the solid part as isointense and the cystic part as slightly low-intensity areas. The calcification appeared as a lower intensity area. The solid part and the tissue surrounding the cyst were enhanced by gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA), and sagittal and coronal imaging demonstrated a multilocular tumor shadow. T₂-weighted imaging demonstrated the solid part as

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Fig. 1 *left*: Precontrast CT scan on admission, showing the isodense solid tumor abutting the right parietal bone with a calcification and a low-density area suggesting a cyst. *right*: The solid tumor was highly enhanced postcontrast.

Fig. 2 A: T₁-weighted axial MR image, showing the solid tumor as an isointense mass and the cystic lesion as a low-intensity area. B: T₂-weighted axial MR image, showing the solid tumor as a mixed iso- and high-intensity mass, and the cyst as a high-intensity area. C, D: T₁-weighted sagittal (C) and coronal (D) images, showing the solid tumor and the cyst wall enhanced markedly with Gd-DTPA, and a multilocular tumor shadow.

an irregular iso- and high-intensity area and the cystic part as a high-intensity area. The tumor was well defined by the peritumoral low-signal band (Fig.

Fig. 3 Right external carotid angiograms, arterial (*upper*) and late arterial phases (*lower*), showing the tumor shadow fed by a posterior branch of the middle meningeal artery.

2). These images suggested that the tumor was mainly localized extra-axially. Right external carotid angiography demonstrated that the tumor was fed by a posterior branch of the middle meningeal artery (Fig. 3). Right internal carotid and vertebral arteriography demonstrated compression of a parieto-occipital branch of the right posterior cerebral artery but no feeding vessels to the tumor shadow (Fig. 4). The diagnosis was cystic meningioma or meningeal sarcoma.

A parieto-occipital craniotomy was performed on June 3, 1992. The bone and the outer surface of the dura looked intact. A red-brown tumor located on the surface of the parietal lobe was well demarcated from the brain tissue and compressed the surrounding surface vessels of the brain. The tumor, containing a cyst, was fed by the middle meningeal artery and had partly adhered to the inner surface of the dura. Virtually all the tumor was removed en bloc including the calcification.

Histological examination of the surgical specimens showed the cells had round nuclei, clear cytoplasm, perivascular pseudorosettes, and immunoreactivity for glial fibrillary acidic protein (GFAP) (Fig. 5). Im-

Fig. 4 Right internal carotid (*upper*) and right vertebral angiograms (*lower*), demonstrating compression of a perieto-occipital branch of the right posterior cerebral artery but no feeding vessels to the tumor shadow.

munohistochemical staining demonstrated absence of epithelial markers, *i.e.*, epithelial membrane antigen and keratin. The diagnosis was clear cell-type ependymoma.^{8,10,11)}

The postoperative course was uneventful and he was discharged with no neurological deficits on August 25.

Discussion

Our patient harbored a tumor mainly located extra-axially and fed by the external carotid artery. The relationship with the upper ventricular system could not be sufficiently visualized by imaging. Ependymoma extending extra-axially is extremely rare, with only one reported patient with a mixed glioma composed of ependymoma and oligodendroglioma.¹⁵⁾ This tumor was associated with the cranial ventricular system and extended by dissemination into the medullary space. Zentner *et al.*¹⁶⁾ found only 10 cases of solitary intracranial extracerebral glioma, including four astrocytomas, three oligodendrogliomas, two glioblastomas, and one mixed

Fig. 5 Photomicrographs of the tumor specimen, showing oligodendroglioma-like cells with round nuclei and clear cytoplasm, forming perivascular pseudorosettes. *upper*: HE stain, $\times 100$; *middle*: HE stain, $\times 400$; *lower*: GFAP stain, $\times 100$.

glioma. Such leptomeningeal gliomas presumably originate from heterotopic nests of glial cells.^{6,7,9,16)}

There are three possible mechanisms for tumor growth in our case. First, a tumor occurred around the ventricular wall and grew gradually, penetrating the arachnoid membrane to extend extramedullarily and obtaining a blood supply from the adjacent dura. The part of the tumor around the cerebral ventricle degenerated and became necrotic, then calcified. This sequence obscured the relationship with the cerebral ventricle. The tumor may then have

Fig. 6 Possible modes of tumor extension. *left*: A bundle of subependymal neural glia entered the white matter to form cell nests separate from the cerebral ventricle. *center*: A tumor originated from the nests penetrating the arachnoid membrane to extend extramedullarily and develop a blood supply from the adjacent dura. *right*: The tumor originating from the nests degenerated and became necrotic, then calcified.

grown and become mainly localized extramedullarily. Second, a bundle of subependymal neural glia entered the white matter to form cell nests separate from the cerebral ventricle.^{1,4,12-14} A tumor originated from these nests, extended extramedullarily, and developed a blood supply from the adjacent dura, thus appearing to be mainly localized extramedullarily (Fig. 6). Third, the tumor could contain two histological components such as glioma and meningioma, but this is unlikely because all parts of the tumor specimen showed the same histology.

The present patient demonstrated a unique location for ependymoma. The possibility of glioma should be considered when treating tumors originating in the dura.

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