Medulloblastoma Associated with Cysts and Calcifications
—Case Report—

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Abstract

An 11-year-old boy presented with medulloblastoma occurring in the cerebellar vermis. Computed tomography and magnetic resonance imaging revealed numerous cysts and calcifications in the tumor. The tumor was subtotally removed and cellular synchronization radiation therapy given. He was discharged without neurological deficits. Histological examination showed the cysts represented necrotic foci. Macrophages, which appeared around the necrotic foci, were important in the development of the calcifications via proliferation of collagen fibers.

Key words: medulloblastoma, cyst, necrosis, collagen fiber, calcification, macrophage

Introduction

Medulloblastoma is thought to originate from the cells of the external granular layer which often persist in the posterior medullary velum after birth. JC papovavirus acts on the cells of the external granular layer of the cerebellar vermis to induce a primitive neuroectodermal tumor in the hamster. At present, virus-induced carcinogenesis is the most likely cause of medulloblastoma.

Radiologically detectable calcifications are rarely associated with medulloblastoma, although the occasional appearance of dystrophic calcifications after radiotherapy is well known. The reported incidence of calcifications in untreated medulloblastoma is low: three of 24 cases, one of 20 cases, and four of 30 cases. Cysts occur a little more frequently than calcifications.

We describe a patient with medulloblastoma accompanied by numerous cysts and calcifications demonstrated by computed tomography (CT) and magnetic resonance (MR) imaging, and a histological study of the mechanism of formation of the cysts and calcifications.

Case Report

An 11-year-old boy began to fall frequently from his bicycle, starting early in December, 1992. At 8:30 p.m. on December 16, he fell downstairs at home. He was admitted to a nearby hospital with a diagnosis of head injury. CT scans at the hospital demonstrated a tumor in the posterior fossa, but no intracranial hemorrhage. On December 17, he was referred to our university hospital.

On admission, signs of intracranial hypertension and truncal ataxia were apparent, although his consciousness was clear. He underwent implantation of a ventriculoperitoneal shunt on the same day. Precontrast CT revealed a relatively high-density tumor in the median region of the posterior fossa, accompanied by numerous cysts and calcifications (Fig. 1A). Postcontrast CT showed the tumor was heterogeneously enhanced. T1-weighted MR imaging showed the tumor as isointense, with hypointense cysts and calcifications (Fig. 1B). T2-weighted MR im-

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aging showed the tumor as relatively hyperintense, with hyperintense cysts and hypointense calcifications (Fig. 1C). Intravenous gadolinium-diethylene-triaminepenta-acetic acid injection heterogeneously enhanced the tumor located in the fourth ventricle (Fig. 1D). Four-vessel angiography revealed tumor stains supplied by the vermian branches of the bilateral posterior inferior cerebellar arteries. 

He underwent suboccipital craniectomy on January 6, 1993, under a diagnosis of a fourth ventricle tumor. The tumor was reddish gray, relatively soft, and bled easily. It originated from the posterior medullary velum. The tumor was most tightly adherent to the caudal part of the fourth ventricle floor. The tumor was subtotally removed, leaving this portion of the tumor unremoved.

Histological examination of the surgical specimen demonstrated that the tumor cells with poor cytoplasm and oval nuclei had proliferated and formed Homer Wright rosettes (Fig. 2A). Numerous endothelial proliferations and necrotic foci were visible, and foamy macrophages were seen around the necrotic foci (Fig. 2B). Clusters of foamy macrophages were present within the entire tumor (Fig. 2C). Calcifications were visible in areas showing remarkable proliferation of collagen fibers (Fig. 2D). Immunohistochemical examination showed that the tumor cells were negative for glial fibrillary acidic protein, S-100 protein, and neuron-specific enolase. The foamy macrophages were positive for lysozyme and alpha-1-antitrypsin. Electron microscopy showed that the tumor cells had no junctional complexes, microvilli, or cilia (Fig. 3). Based on these findings, the tumor was classified as a medulloblastoma rather than an ependymoblastoma.

Postoperatively, he received cellular synchronization radiation therapy using 1-(4-amino-2-methyl-5-pyrimidinyl)methyl-3-(2-chloroethyl)-3-nitrosourea hydrochloride and vincristine, and radiation doses of 30 Gy for the whole brain, 40 Gy for the posterior fossa, and 20 Gy for the spinal cord. After rehabilitation, he was discharged on April 3, without neurological deficits.

Discussion

Our case was characterized histologically by the presence of endothelial proliferation and numerous necrotic foci, the appearance of foamy macrophages everywhere in the tumor, the remarkable proliferation of collagen fibers, and the presence of calcifications. Large necrotic foci were detected as cysts by neuroimaging methods. Previous reports suggest that calcifications are likely to be accompanied by cysts. These facts suggest that the calcifications in the present patient developed in the following steps: 1) secretion of cytokines such as interleukin-1 and basic fibroblast growth factor from the foamy macrophages appearing around the necrotic foci, 2) proliferation of collagen fibers by fibroblasts due to stimulation by these cytokines, and 3) induction of hyaline degeneration by these collagen fibers, resulting in calcifications. Although we do not think that this mechanism can explain all calcifications which accompany medulloblastoma, we believe that macrophages were important in the development of the calcifications observed in the present case.

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Fig. 2 Light micrographs, showing the tumor cell proliferation forming Homer Wright rosettes (A); numerous endothelial proliferations and necrotic foci, and foamy macrophages around the necrotic foci (B); the tumor tissue containing numerous clusters of foamy macrophages (C); and calcifications in areas showing intense proliferation of collagen fibers (D). HE stain, ×100.

Fig. 3 Electron micrograph, showing the tumor cells with no junctional complexes, microvilli, or cilia. Bar = 2 µm.

References

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