Desmoplastic Infantile Ganglioglioma with Extraparenchymatous Cyst

—Case Report—

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Abstract

A large supratentorial tumor associated with an extraparenchymatous cyst and multiloculated intraparenchymatous cysts occurred in a 14-month-old infant. This case had all the characteristic features of desmoplastic infantile ganglioglioma both clinically and histologically. The notable difference was the extraparenchymatous cyst. The extraparenchymatous cyst was probably caused by entrapment of cerebrospinal fluid in the subarachnoid space by some check-valve mechanism because the leptomeninges were commonly involved in the tumor. A similar mechanism may explain the etiology of the intraparenchymatous, disproportionately large cyst in desmoplastic infantile gangliogliomas.

Key words: desmoplastic infantile ganglioglioma, embryonic tumor, extraparenchymatous cyst

Introduction

Embryonal cerebral tumors expressing multiple differentiation are rare, and present challenging problems of diagnosis and classification. Desmoplastic infantile ganglioglioma is a massive supratentorial neuroepithelial tumor with dense desmoplastic tissue and divergent astrocytic and ganglionic differentiation that occurs in infants.¹⁰⁾

We describe a large supratentorial tumor with an extraparenchymatous cyst and multiloculated intraparenchymatous cysts in a 14-month-old infant which presented with the histological characteristics of desmoplastic infantile ganglioglioma. This is the first report of desmoplastic infantile ganglioglioma in Japan.

Case Report

A 14-month-old boy was admitted to a local hospital with generalized convulsion. Computed tomographic (CT) scans disclosed a huge supratentorial tumor. His medical history was not significant including during pregnancy and delivery. He had

achieved normal developmental milestones both physically and mentally. His family history was not contributory.

On admission, the anterior fontanelle was not tense and his head circumference was 51.0 cm. Neurological examination revealed no abnormalities. Plain skull x-ray films showed no suture separation. CT scans and magnetic resonance (MR) images demonstrated a large mass with multiloculated cysts in the left frontoparietal area, displacing the midline structure contralaterally. The solid portion in the superior frontal convexity was isointense to the gray matter on the T₁-weighted images (Fig. 1 *upper*), and was enhanced remarkably after infusion of gadolinium-diethylenetriamine-penta-acetic acid (Gd-DTPA) (Fig. 1 *lower*). The cyst wall was not enhanced except for septum-like structures. No continuity between the cystic spaces and the ventricular system was shown on any MR image. The cyst adjacent to the dura mater was located extraparenchymatously, confirmed by cerebral angiography. Internal and external carotid angiograms showed no tumor staining.

A left frontoparietal craniotomy was performed on December 7, 1990. The dura was not tense and no abnormalities were found in its outer surface. The

Fig. 1 *upper*: T₁-weighted coronal (*left*) and sagittal (*right*) MR images, showing a large mass with multiloculated cysts in the left frontoparietal region. *lower*: T₁-weighted coronal (*left*) and sagittal (*right*) MR images with Gd-DTPA, demonstrating remarkable enhancement in the peripheral tumor. Note there is no enhancement in the cyst wall except for septum-like structures.

Fig. 2 Intraoperative photograph, showing a large cyst (*arrow*) located subdurally without attachment to the dura mater. The solid part has been dissected free from the dura mater.

solid portion of the mass was firm, yellowish in color, and attached to the dura tightly. The leptomeninges were included in the tumor and not clearly separated from the superficial cortex. A cyst contain-

ing xanthochromic fluid was located subdurally, just lateral to the tumor (Fig. 2). The cyst wall was not demarcated from the solid mass superiorly or from the surrounding arachnoid membrane inferiorly. The tumor was dissected from the more normal-appearing tissue around the lesion. In the posterior mass, no clear cleavage plane could be found. The large, deep multiloculated cyst contained xanthochromic fluid. Only the solid portion between the cyst was removed, achieving subtotal removal of approximately 80% of the mass. The laboratory analyses, including protein content, of the xanthochromic fluid obtained from both extraparenchymatous and intracerebral cysts were similar.

Histological examinations using neurohistological and immunohistochemical methods showed a marked desmoplastic appearance in the superficial tumor (Fig. 3 *upper*), in which a few neuroepithelial cells intermingled with the connective tissue. Melanocytic infiltration was also identified in this area. In a distinct, but adjacent area to, the desmoplastic areas, neoplastic neuroepithelial cells in various stages of differentiation infiltrated the surrounding brain (Fig. 3 *middle*). Immunoperoxidase staining for glial fibrillary acidic protein (GFAP) demonstrated some astrocytic differentiation (Fig. 3 *lower*). The ganglion cells were mixed with fully developed and immature cells with large, foamy, and round cytoplasm, which stained positively for neuron-specific enolase and neurofilament polypeptides (200 kD). Bielschowsky silver impregnation revealed neuritic processes. No mitotic figures or foci of necrosis were present. The specimen taken from the extraparenchymatous cyst wall also showed a dense network of connective tissue.

The postoperative course was uneventful. He received neither radiation therapy nor chemotherapy. Eighteen months after surgery, he was asymptomatic and neurological examination revealed no abnormalities.

Discussion

VandenBerg *et al.*¹⁰⁾ first proposed the name desmoplastic infantile ganglioglioma in 1987, and recently described the clinicopathological features of 16 cases of this rare entity,⁹⁾ and discussed two other cases in the literature.^{2,5)} All tumors occurred before the age of 18 months, usually within the first 4 months of life. They most often involved two hemispheric lobes, and invariably contained cysts, which often accounted for a significant fraction of the tumor volume. The solid component was remarkably firm, but associated with lack of any

Fig. 3 Photomicrographs, showing the characteristic features of desmoplastic infantile ganglioglioma. *upper*: The desmoplastic component of the tumor contains a wavy pattern of fibroblastic cells at the distinct interface with the surrounding brain. HE stain, $\times 75$. *middle*: Neoplastic neuroepithelial cells in varying stages of differentiation in the surrounding brain. Luxol fast blue and cresyl violet stain, $\times 150$. *lower*: GFAP immunohistochemical staining demonstrates typical astrocytes in the desmoplastic area. GFAP avidin-biotin-peroxidase complex immunohistochemical staining with hematoxylin counterstain, $\times 75$.

definite cleavage plane from the surrounding brain. The superficial portions had a variable attachment to the dura. None communicated with the ventricles. Tumors contained dense desmoplastic tissue superficially, with variable numbers of pleomorphic

neuroepithelial cells. Divergent astrocytic and neuronal differentiation occurred in most cases, and astrocytic differentiation in all. Successful total or near-total surgical removal was followed by a favorable postoperative course extending over many years of tumor-free survival.

Several similar cases have been reported. Taratuto *et al.*⁸⁾ described six cases of superficial cerebral astrocytoma attached to the dura. These tumors had many characteristic clinicopathological features of desmoplastic infantile ganglioglioma including occurrence in infancy, large size with cystic components, richness in reticulin fibers, and favorable prognosis. Absence of neuronal differentiation was only the difference. De Chadarevian *et al.*¹⁾ described desmoplastic cerebral astrocytoma in infancy identical to these cases with absence of neuronal elements. Ishida and Tamura³⁾ reported a large supratentorial tumor with massive calcification in a 4-year-old boy. The presence of ganglion cells, connective tissue cells and fibers, and glial cells interpreted as astrocytes was confirmed by electron microscopy and immunohistochemical techniques. They described this tumor as desmoplastic ganglioglioma. Although not identical to the desmoplastic infantile ganglioglioma of VandenBerg,^{9,10)} these may be closely related entities because variable differentiation may occur in desmoplastic infantile ganglioglioma.⁷⁾ VandenBerg *et al.*¹⁰⁾ proposed that the group of tumors reported by Taratuto *et al.*⁸⁾ represented examples of restricted astrocytic differentiation occurring in a tumor entity identical to desmoplastic infantile ganglioglioma.

Our case had all the characteristic features of desmoplastic infantile ganglioglioma, though the follow-up period was short. The notable difference was the extraparenchymatous cyst in addition to the multiloculated intraparenchymatous cysts. Although the cyst in desmoplastic infantile ganglioglioma is one of the salient gross tumor features,^{7,9,10)} no precise relationship between cyst and solid portion has been given. However, judging from the literature, most cysts associated with this tumor were not located in the solid tumor, but around the solid mass. No histological features of the cyst wall were described,^{2,5,9)} so it is difficult to discuss the etiology of cyst formation from the limited data.

Extraparenchymatous cysts have not been reported even in cases similar to desmoplastic infantile ganglioglioma such as superficial cerebral astrocytoma⁸⁾ and related entities.^{1,3)} We found dense collagenous tissue in the outer wall of the extraparenchymatous cyst, which is the most characteristic finding of this entity. In our case, the outer wall of

the cyst was not demarcated from the arachnoid membrane at the periphery, and evacuation of the xanthochromic fluid in the cyst demonstrated no membranous structure on the cerebral cortex. Therefore, this extraparenchymatous cyst was probably caused by entrapment of cerebrospinal fluid, probably in the subarachnoid space by some check-valve mechanism. Similar mechanisms explain the etiology of the intraparenchymatous, disproportionately large cysts in desmoplastic infantile ganglioglioma, because the leptomeninges were commonly involved in the tumor.^{9,10)}

The massive component of the dense desmoplasia may influence the clinical behavior of these tumors by increasing tumor size.¹⁰⁾ Leptomeningeal extension of the growth was a salient characteristic,^{9,10)} which is documented in other forms of neuroepithelial tumor such as desmoplastic medulloblastoma⁶⁾ and pleomorphic xanthoastrocytoma.⁴⁾ The mechanism of desmoplasia is not fully understood, and will be discussed elsewhere. Clinically, appropriate surgical treatment of these tumors can achieve a favorable prognosis. Unnecessary radiation therapy or chemotherapy should be avoided.

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