Desmoplastic Infantile Ganglioglioma
—Case Report—

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Abstract

A desmoplastic infantile ganglioglioma occurred in a 5-month-old boy, manifesting as delayed psychomotor development. Computed tomography showed a large cystic mass involving the left occipitoparietal lobes. The tumor was totally removed. He has survived for 13 years without tumor recurrence. Histological examination disclosed marked desmoplasia intermixed with mature-looking ganglion cells, neoplastic glial cells, and small, immature round cells. These clinical and histological features enabled the diagnosis of desmoplastic infantile ganglioglioma.

Key words: desmoplasia, ganglioglioma, brain tumor, infant

Introduction

Desmoplastic infantile ganglioglioma is a tumor characterized by voluminous size, intense desmoplasia, and the frequent presence of astrocytic and ganglionic differentiation occurring in infant patients aged less than 18 months, and mostly less than 4 months.16,18 Similar infantile tumors marked by intense desmoplasia containing a mixed population of neoplastic and neuronal cells have also been reported.5,9 Previously, this tumor was grouped with cerebral neuroblastoma or given a variety of names such as composite cerebral neuroblastoma and astrocytoma,14 cerebral ganglioglio-neuroblastoma,13 and complex cerebral tumor.8 Desmoplastic infantile ganglioglioma is differentiated from classical ganglioglioma5 by clinical presentation in infancy, the more frequent inclusion of immature cells, and the invariable presence of dense desmoplasia. In contrast to the ominous histopathology of desmoplastic infantile ganglioglioma, the clinical course appears to be favorable with successful surgical removal. Therefore, distinction of desmoplastic infantile ganglioglioma from other infantile neuroepithelial tumors is important.

Here, we report a patient with desmoplastic infantile ganglioglioma and discuss the characteristic histological features which led to the correct diagnosis.

Case Report

A 5-month-old boy was referred to our hospital for evaluation because of delayed psychomotor development. His head measured 43 cm and the anterior fontanel was 3 x 3.5 cm. His neck was not fixed and he could not change his sleeping position. Neurological examination revealed bilateral optic atrophy and right hemiparesis with increased deep tendon reflexes. Computed tomographic (CT) scans showed a large cystic mass involving the left occipitoparietal lobes with enhanced lesions in the cyst and dilated lateral ventricles postcontrast (Fig. 1). Left carotid and vertebral angiograms demonstrated an avascular mass in the left occipitoparietal lobes and a small tumor stain fed by the anterior choroidal artery.

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The tumor was removed via craniotomy on December 1, 1978. Opening the dura exposed a large cyst surrounded by a thin membrane. The medial aspect of the cyst opened into the lateral ventricle. A soft mass attached to the choroid plexus was found at the bottom of the cyst. The tumor was totally removed.

The surgical specimen was fixed in 10% buffered formalin and embedded in paraffin for light microscopy. Routine HE staining, Watanabe’s silver impregnation for reticulin, and immunohistochemical staining for glial fibrillary acidic protein (GFAP), neuron-specific enolase (NSE), and synaptophysin were carried out. The specimens showed a marked desmoplastic appearance in the superficial part of the tumor (Fig. 2A). In other areas, the neoplastic cells showed various lines of differentiation. Some areas of the tumor continued aggregates of mature-looking ganglion cells which stained positively for NSE (Fig. 2B) and synaptophysin.

Fig. 1 Postcontrast CT scans on admission, showing a large cyst with enhanced lesions in the left occipitoparietal lobes.

Fig. 3 Follow-up postcontrast CT scan 13 years after surgery, showing no evidence of recurrence and the shunt in the lateral ventricle.

(Fig. 2C), but not for GFAP. The most frequent cells were small, immature round cells with hyperchromatid nuclei, but without Homer-Wright rosettes (Fig. 2D). Only some cells stained positively for GFAP. Neoplastic astrocytes were prominent within the desmoplastic areas (Fig. 2E). Mitotic figures were present but never abundant.

Postoperative local radiation of 17 Gy to the tumor site was administered. One year later, a ventriculoperitoneal shunt was placed because of increasing ventricular dilatation. He is now 13 years old, and has a severe psychomotor development delay and hemiparesis on the right side. A recent CT scan revealed no residual tumor (Fig. 3).

Discussion

The histological features of our case were voluminous tumor size with a cyst and a mixed cell population consisting of neural and glial cells with immature cells, consistent with the previous observations of desmoplastic infantile ganglioglioma.\textsuperscript{16,18} Despite the similar histology, our case presented different clinical data. Desmoplastic infantile gangliogliomas described previously frequently occurred in the frontal and parietal lobes, and none communicated with the ventricles, while leptomeningeal extension was common.\textsuperscript{16,10} In our patient, the tumor communicating with the lateral ventricle but without an obvious connection with the leptomeninges. Desmoplasia with a neuroepithelial component is well known in other brain tumors such as pleomorphic xanthoastrocytoma,\textsuperscript{9} desmoplastic medulloblastoma,\textsuperscript{12} and desmoplastic cerebral astrocytoma of infancy.\textsuperscript{2} The superficial location of some tumors suggests that interaction with the leptomeninges is important for desmoplasia. Alternatively, astrocytes and other neuronal tumor cells may secrete collagens.\textsuperscript{3,15} The massive component of dense desmoplasia without leptomeningeal extension seen in our patient may result from the latter.

The particular cell type of origin is difficult to identify in embryonal central nervous system tumors. However, the divergent neuroglial and neuronal differentiation within desmoplastic infantile ganglioglioma strongly implies bipotential, supratentorial, fetal precursor cells. The distinctive desmoplastic leptomeningeal interaction suggests a cerebral external granular layer cell as the tumor origin.\textsuperscript{17} Primitive neuroectodermal tumors (PNETs) may also exhibit such differentiation along neuronal and glial cell lines. However, PNETs mostly consist of undifferentiated matrix cells, without bizarre neurons or giant ganglion-like cells.\textsuperscript{6,10,11} The prognosis for patients with PNETs is poor and surgical removal alone cannot achieve remission because of local recurrence.\textsuperscript{8} This prognosis is quite different from that for desmoplastic infantile ganglioglioma.

VandenBerg\textsuperscript{16} reported that the prognosis for desmoplastic infantile gangliogliomas was very favorable after surgical removal because of no recurrence in patients treated with surgical removal alone. We first diagnosed this patient as having PNET with neuronal differentiation, and therefore administered 17 Gy radiation. He has lived for 13 years without recurrence despite the small radiation dosage. The successful histological identification of desmoplastic infantile ganglioglioma, a surgically curable tumor, is important because it precludes unnecessary radiation and/or chemotherapy.

References


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