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Combined Fontanelle Puncture and Surgical Operation in Treatment of Desmoplastic Infantile Astrocytoma: Case Report and a Review of the Literature

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Desmoplastic infantile astrocytoma is a rare low-grade malignant brain tumor found in infants. Its pathological diagnosis can be made on the basis of its histological characteristics and immunohistochemical staining. A case of desmoplastic infantile astrocytoma, including its clinical manifestations, pathological characteristics, differential diagnosis, treatment, and prognosis, is reported. Presurgical percutaneous decompression and subsequent resection resulted in a satisfactory therapeutic outcome.

Keywords: desmoplastic infantile astrocytoma; tumor; pathology; treatment

Desmoplastic infantile astrocytoma is a rare tumor found in infantile cerebral hemispheres. It is usually a very large, slow-growing tumor, with a structure similar to that of sarcoma. The general prognosis is good, although large tumors are difficult to treat because of the high risk associated with surgery. This article describes the clinical manifestations, pathological characteristics, and differential diagnosis of desmoplastic infantile astrocytoma through a review of the literature and presentation of a case. We sought to analyze and describe how to remove safely and completely such a large tumor while preventing severe complications such as major collapse of brain tissue and leakage of cerebrospinal fluid.

Case Report

The patient was a 1-month-old boy, weighing 3.2 kg, who was born at full-term by cesarean section to a gravida I, para I woman. Two weeks previously, during a regular physical examination in a local hospital, the patient’s head circumference was found to be slightly enlarged. Five days before our evaluation, the patient’s parents had noted obvious enlargement of the patient’s head along with dysphoria and sleepiness without vomiting or seizure. The results of the examination at our hospital showed a head circumference of 48 cm and an anterior fontanelle measuring 4 × 4 cm, without sunset eyes syndrome. Brain magnetic resonance imaging (MRI) revealed a large area of signal abnormality, measuring 11.19 × 9.71 × 9.80 cm in the right cerebral hemisphere, almost completely filling it. An area of soft tissue signal abnormality measuring 5.05 × 2.0 × 2.35 cm was found in the posterolateral wall. On MRI, the signals were isointense on T1-weighted images; hyperintense, hypointense, and isointense on T2-weighted images; and hyperintense on diffusion-weighted images. The midline deviated to the left, and the left lateral ventricle was obviously enlarged. The anterior horn of the right lateral ventricle was enlarged, and the posterior horn had disappeared. The midbrain aqueduct was compressed and occluded (Figure 1). On the basis of the large lesion in the right cerebral hemisphere, obstructive hydrocephalus was suspected, and the patient was hospitalized. On the day of hospitalization, the patient was vomiting and exhibited dysphoria. Mannitol was administered to reduce intracranial pressure, but its efficacy was not apparent. On the second day, the patient was in a coma with diminished breath sounds, and the anterior and posterior fontanelles were tense. The right pupil was larger than the left one, but light reflex was present. Percutaneous decompression was then performed immediately from the right side of the posterior fontanelle, which slowly released...
60 mL of yellow fluid. At this time, the patient opened his eyes, his breath sounds improved, and the anterior fontanelle softened. After removal of the trocar tube, pressure was applied to the puncture site for 10 minutes. The child did not eat or cry, however, and had slow response and skin pallor. Because the patient had anemia, he was given a blood transfusion. On the third day, 60 mL of fluid was again released from the posterior fontanelle, and the patient’s condition was clearly improved; he ate and laughed and did not vomit. On the fourth day, brain computed tomography (CT) scanning showed that the lesion in the right cerebral hemisphere had diminished in size, the posterior horn of the right ventricle had appeared, and the midline deviation was corrected (Figure 2). On the fifth day, complete tumor resection was performed. The tumor was cystic and solid, with the solid part, measuring $6 \times 6 \times 2$ cm, located in the right posterior occipital lobe (Figure 3). After surgery, the patient was conscious and lucid, had a good appetite, and did not vomit. He could see and laugh without any indication of nervous system damage such as hemiplegia or seizure. On the seventh day after surgery, a CT scan showed that the tumor in the right cerebral hemisphere had been completely removed and the midline deviation had been corrected, but subdural effusion was found (Figure 4).

Pathological examination revealed a World Health Organization (WHO) classification grade 1 desmoplastic infantile astrocytoma, consisting of asteroid and spindle-shaped cells. Immunohistochemical studies showed that the tumor cells were positive for glial fibrillary acidic protein (Figure 5) and negative for synaptophysin (Figure 6) and neurofilament protein. Three months after surgery, the patient exhibited nearly normal growth and development, with the ability to follow an object with his eyes, laugh, and raise his head. Eighteen months after surgery, the infant’s development was completely normal. The brain CT scans showed an increased volume of the right cerebral hemisphere without space-occupying lesions and an absence of midline deviation. Both the right and left lateral ventricles were enlarged, with a better appearance than on the previous CT scan, and the right subdural effusion was almost completely absorbed (Figure 7).

**Discussion**

Desmoplastic infantile astrocytoma was defined as a new type of central nervous system tumor by the World Health Organization in 1993. This rare tumor, first reported by Taratuto et al in 1984, usually occurs in infants younger than 1 year and is frequently located in the
The tumor has a diameter of 6 to 12 cm and a solid area consisting of one or more cysts. It grows slowly and has significant pathological characteristics such as a fibrotic matrix arranged in an abundant bundle or a spoke-wheel pattern containing similar neuroepithelial components differentiated from astrocytes. In 1993, VandenBerg\textsuperscript{3} reported 9 cases of desmoplastic infantile astrocytoma in patients aged 1.5 to 14 months (mean 6.8 months; median 6 months) with a male:female ratio of 0.8:1.

The major clinical manifestations of desmoplastic infantile astrocytoma include (1) increased intracranial pressure, causing symptoms such as dysphoria, vomiting, bulging fontanelle, and sunset eyes (sunset syndrome); (2) abnormally increased head circumference; (3) seizure; and (4) other symptoms such as skull deformation, eyesight damage, and motion and sensory difficulties.\textsuperscript{4,5} Clinical presentations of this tumor are similar to those of space-occupying lesions, but the results of imaging studies such as CT and MRI show relatively specific features. Usually the tumor is located in a surface area and presents as nodular or patchy enhancements in the frontoparietal or temporal part of the meninges. The inner part of the tumor is usually cystic.

Diagnosis of desmoplastic infantile astrocytoma is made on the basis of pathological examination of resected specimens. The tumor tissue consists of asteroid and spindle cells. Spindle cell tissue usually has better differentiation, and its structure is the same as that of fiber cells or fibroblasts arranged in bundle or spoke-wheel patterns.
Asteroid cells resemble nests or bundles distributed among spindle cells. The nucleus of an asteroid cell is small without a nucleolus, and the chromatin is thin. Part of the nucleus is strongly basophilic, and the plasma is light or transparent. The tumor has abundant silver-staining fibers similar to those found in mesenchymal tumors. Histological examination reveals minor polymorphism. Immunohistochemical analysis shows that the majority of tumor cells are glial fibrillary acidic protein (GFAP) positive, an important finding for accurate diagnosis. The clinical biological behavior and morphology of desmoplastic infantile astrocytoma are similar to those of desmoplastic infantile ganglioglioma. Compared with desmoplastic infantile astrocytoma, desmoplastic infantile ganglioglioma has obvious neuronal components. The 2 tumors are similar histologically; although the World Health Organization places both in the same category, Louis et al considered them as 2 different types. The expanded basal cell layer is the reason for the slow growth of the tumors. On the basis of the cases reported, desmoplastic infantile astrocytoma is much less common than desmoplastic infantile ganglioglioma. In the present case, the tumor lacked clear neuronal components, and synaptophysin staining was negative, leading to the diagnosis of desmoplastic infantile astrocytoma. If histological examination had revealed ganglion cells and the immunohistochemical data were synaptophysin positive, however, the diagnosis would have been desmoplastic infantile ganglioglioma.

Complete resection is associated with a better prognosis than other treatments for patients with desmoplastic infantile astrocytoma. The risks of surgery in infants are significant, however. Brain tissue cannot quickly recover after complete resection of a giant tumor, and major brain tissue collapse can result, possibly leading to

Figure 5. Pathology. A, The tumor consists of paucicellular collagen-rich tissue sharply demarcated from underlying brain. B, The tumor cells were oval, which appear as fibroblasts, nodules of cells more obviously astrocytic are seen. C, Both the fibrotic and astrocytic cells were positive for glial fibrillary acidic protein.
bridging vein rupture and bleeding, massive subdural effusion, hydrocephalus, and even death. In the case reported here, the patient’s tumor was located in the cerebral hemisphere and caused hydrocephalus and brain herniation. If brain ventricle shunting was performed in the dilated ventricle to reduce intracranial pressure, brain herniation might have become severe. In addition, this patient was only 1-month-old, weighing only 3.2 kg, with coma as well as brain herniation. His vital signs were extremely weak. Immediate tumor surgery would have involved greater risk, with a high chance of death. These tumors are quite challenging in clinical practice. The best solution may be draining the tumors fluid by repeated puncture, which not only decreases intracranial pressure to relieve cerebral herniation but also facilitates gradual decompression of brain tissue. The fontanelles of an infant are ideal for applying this strategy. The posterior fontanelle was chosen as the puncture site because it was near the tumor. Using a 22-gauge (or even thinner) trocar, the right posterior fontanelle was punctured to avoid the posterior sagittal sinus. When the trocar entered the tumor, the trocar needle was withdrawn to allow drainage. The patient’s condition was closely observed while draining the tumors fluid. Fast draining is absolutely contraindicated. The draining can be stopped once there are signs of improvement, such as stable respiration and recovery of consciousness. After removal of the trocar tube, pressure was applied to the puncture site for 10 minutes to stop the bleeding. This simple method can be repeated several times and does not require anesthesia. Choosing a thin trocar is safer, as the injury to the brain tissue is minimal. After intracranial pressure was reduced, the midline deviation was corrected, the brain herniation disappeared, and the vital signs were stable, complete tumor resection was performed. Only a tiny remote hemorrhage was found during the operation. Although brain tissue collapse occurred during the tumor resection, it was not severe, and the bridging vein did not have obvious tense traction and suspension. Only subdural effusion without brain hemorrhage occurred, and the prognosis was good.

Generally, desmoplastic infantile astrocytoma is considered a low-grade tumor. Complete resection is associated with the best prognosis, with possible long-term survival. Setty et al. however, described the case of a 4-month-old boy with a large tumor localized in the suprasellar and hypothalamic region with 2 smaller similar masses in the posterior fossa and 1 in the spinal canal. The histopathological findings revealed desmoplastic infantile astrocytoma. Cerebrospinal fluid, obtained at surgery before the tumor manipulation, showed clusters of malignant cells, which were immunopositive for glial fibrillary acidic protein. Darwish et al. also reported on 2 patients with desmoplastic infantile ganglioglioma/desmoplastic infantile astrocytoma, who developed multiple cerebrospinal metastases. It appears that not all tumors with histological features of desmoplastic infantile astrocytoma/desmoplastic infantile ganglioglioma behave in a benign way. Therefore, even though desmoplastic infantile astrocytoma has been defined by the World Health Organization as a grade 1 tumor, close postsurgical follow-up is necessary.

In summary, desmoplastic infantile astrocytoma is a rare low-grade malignant brain tumor found in infants. Its
pathological diagnosis can be made on the basis of its histological and immunohistochemical staining characteristics. Although the tumor volume is usually relatively large, in general total removal can achieve good results. When faced with unstable vital signs, presurgical percutaneous decompression and subsequent resection are the appropriate choices.

References


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