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Desmoplastic infantile astrocytoma and characteristics of the accompanying cyst

Case report

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✓A desmoplastic infantile astrocytoma (DIA) is an extremely rare tumor that comprises a solid astrocytic tumor accompanied by a large cyst and involves the superficial cerebral cortex and leptomeninges in infants. The solid part of this type of tumor has been well described in various reports and books, but characteristics of the cystic portion have remained unclear. Because adequate resection is required to ensure a favorable prognosis, information about the cyst is very important for diagnostic purposes and surgical planning. The authors report on the clinical and histological features of the cyst in a case of a DIA. A 12-month-old boy presented with vomiting. Contrast-enhanced magnetic resonance imaging revealed a strongly enhancing single-lobed large cyst located in the deep white matter, under the solid part of the tumor attached to the dura mater of the left frontal lobe. Both the solid and cystic portions of the tumor were surgically removed. The border between the cyst wall and surrounding white matter was unclear. Histologically, the cyst wall was composed of gliosis representing a rough accumulation of reactive astrocytes, lymphocytes, and small capillary vessels in edematous parenchyma, but no tumor cells. The present case and previous reports suggest that the cyst does not contain tumor cells, even if strongly depicted on contrast-enhanced neuroimaging, and that a thickly enhancing cyst wall indicates gliosis with accumulation of numerous small vessels. *(DOI: 10.3171/PED/2008/1/2/148)*

Key Words • **cyst** • **desmoplastic infantile astrocytoma** • **desmoplastic infantile ganglioglioma** • **histology** • **magnetic resonance imaging** • **pediatric neurosurgery**

A DIA, originally referred to as a "superficial cerebral astrocytoma attached to dura with desmoplastic reaction" or "desmoplastic cerebral astrocytoma of infancy," has been defined as a large cystic astrocytic tumor in infants that involves the superficial cerebral cortex and leptomeninges, often attaching to the dura mater, with a generally good prognosis following adequate resection.[21,22] Both a DIA and a DIG have been integrated into the same tumor type class in the World Health Organization classification system, as both tumors display similar clinical and pathological features, excluding involvement of a variable neuronal component together with astrocytes in a DIG.[21] The solid part of the tumor is easily removed due to the location and clear demarcation from surrounding brain tissue.[20] Because surgical removal of the tumor leads to favorable prognosis, neurosurgeons play a large role in the treatment of a DIA and DIG. Although both a DIA and DIG are very rare, characteristics of the solid part of the tumor have been well described.[4,17,22] In contrast, characteristics of the cyst have remained unclear. For neurosurgeons, general information on clinical and histopathological features of the cyst is important for diagnosis and treatment. The current report describes a case of a DIA with a large cyst, and reviews clinical and histological findings of DIA cysts in the literature.

Abbreviations used in this paper: DIA = desmoplastic infantile astrocytoma; DIG = desmoplastic infantile ganglioglioma; MR = magnetic resonance.

Case Report

History and Examination. This 12-month-old boy was born at term after an uncomplicated pregnancy and delivery. His perinatal course was normal. One month before admission, he presented with vomiting. On admission, an examination revealed a head circumference of 50.3 cm (greater than the mean plus 2 standard deviations) but no neurological deficits. His medical history included no episodes suggestive of

systemic autoimmune disease, intracranial infection, or head injury, and revealed no previous surgery or radiation treatment. Physical and blood serum examinations revealed no abnormalities.

Magnetic resonance was performed, and T1- and T2-weighted imaging revealed a circumscribed solid lesion located superficially in the cortex of the left frontal lobe, and a large cyst in the deep white matter medial to the solid tumor. The T1- and T2-weighted imaging also showed isointense and very low signals, respectively. Gadolinium-enhanced T1-weighted imaging revealed homogeneous enhancement in the solid part of the tumor with the exception of the central region (Fig. 1). A large single-lobed cyst (maximum diameter 5.5 cm) accompanied the solid part of the tumor. Fluid in the cyst displayed greater signal intensity than cerebrospinal fluid. The thick cyst wall also displayed strong enhancement on gadolinium-enhanced T1-weighted imaging (Fig. 1). Computed tomography depicted no calcified foci in the solid part of the tumor or in the cyst wall. Left internal carotid angiography revealed slight intratumoral neovascularity fed from the operculum artery, whereas no intratumoral neovascularity was found on external carotid angiography.

Operation and Postoperative Course. A skin incision and craniotomy were performed at the region of presumed exposure of the tumor. Before opening the dura, xanthochromic fluid in the cyst was aspirated using ultrasonographic guidance until sufficient relaxation of the frontal lobe was achieved. Upon opening, the dura did not firmly adhere to the solid part of the tumor. The meningioma-like hard and reddish-gray solid portion was well demarcated due to preservation of arachnoid matter adjacent to the tumor, and was easily separated from surrounding cortex and edematous white matter. After removal of the solid portion, the cyst wall was noted to consist of a soft and gliotic membrane with a reddish-gray color. Although the cyst could be removed completely, separation of the cyst wall from surrounding white matter was more difficult than removal of the solid portion, because the border between the cyst wall and surrounding white matter was unclear. We dissected between the cyst and surrounding white matter using an ultrasonic surgical aspirator. Postoperative MR imaging demonstrated that both the solid and cystic portions of the tumor had been completely removed.

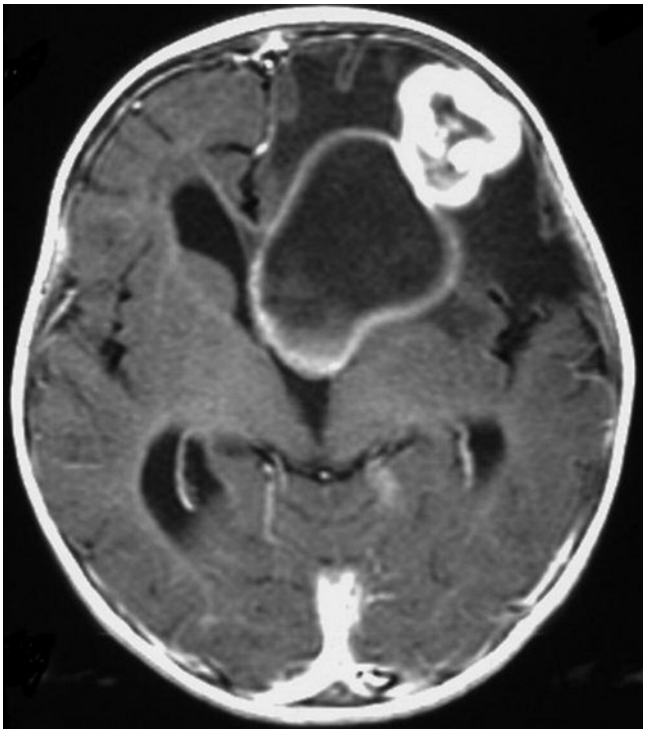

FIG. 1. Gadolinium-enhanced T1-weighted MR image reveals a heterogeneous enhancing mass attached to the dura in the left frontal convex. The mass accompanies a large and thickly enhancing cyst in the deep white matter.

Histology. The removed solid part of the tumor revealed typical histological features of proliferating spindle-shaped cells intermingling with large amounts of collagenous stroma showing storiform or whorled patterns (Fig. 2 *upper*). The collagenous stroma displayed positive results for silver staining. Spindle-shaped cells were immunohistochemically positive for glial fibrillary acidic protein. No necrosis, mitosis, calcified focus, or synaptophysin-positive ganglion cells were seen anywhere in the tumor. The histological diagnosis of the tumor was a DIA.

The cyst wall demonstrated histological features of gliosis, with numerous small round cells such as astrocytes and lymphocytes, and numerous small vessels scattered in edematous parenchyma. Reactive gliosis was observed in the surrounding white matter (Fig. 2 *lower*). In particular, small vessels had accumulated close to the outside of the cyst wall. The cyst wall and brain tissue adjacent to the tumor were thoroughly positive for glial fibrillary acidic protein. Examination of the entire cyst wall, however, revealed no tumor cells.

Discussion

The DIA and DIG tumor types are extremely rare. Only 31 cases of a DIA have been reported in the literature.[1,3,5–19,21,22,24] The tumor was accompanied by a cyst in 30 of 32 cases (93.8%), including the present case. The DIA comprised a solid tumor without a cyst in only 2 cases.[8,18] Cyst size was defined in 15 cases from 9 reports (including the present study) and varied widely between 1.5 and 12 cm (mean 9.3 cm). The cyst in a DIA is most frequently composed of a single lobule on preoperative neuroimaging as in the present case, rather than multiple lobules separated by septa. Reports have described cysts with 1, 2, and 3 lobules in 10, 3,[7,16,17] and 2 cases,[14,19] respectively. The solid part of a DIA is frequently located at the superficial cerebrum, and the cyst has been located in the deep cerebrum under the solid portion in almost all previous cases. In 19 cases for which cyst location was defined,[1,3,6,7,9,10,13–17,19,21,22,24] only 3 cases revealed the cyst to be in a location other than deep white matter medial to the solid part of the tumor.[10,19,24]

The cyst in the present case possessed a thick wall in addition to strong enhancement on gadolinium-enhanced T1-weighted imaging. Results from previous studies, however, have suggested that the cyst wall was obscure and lacked enhancement on contrast-enhanced computed tomography scans or MR images.[19,23] In the literature, 15 reports have described findings on contrast-enhanced computed tomography scans or MR images, with 11 cases demonstrating

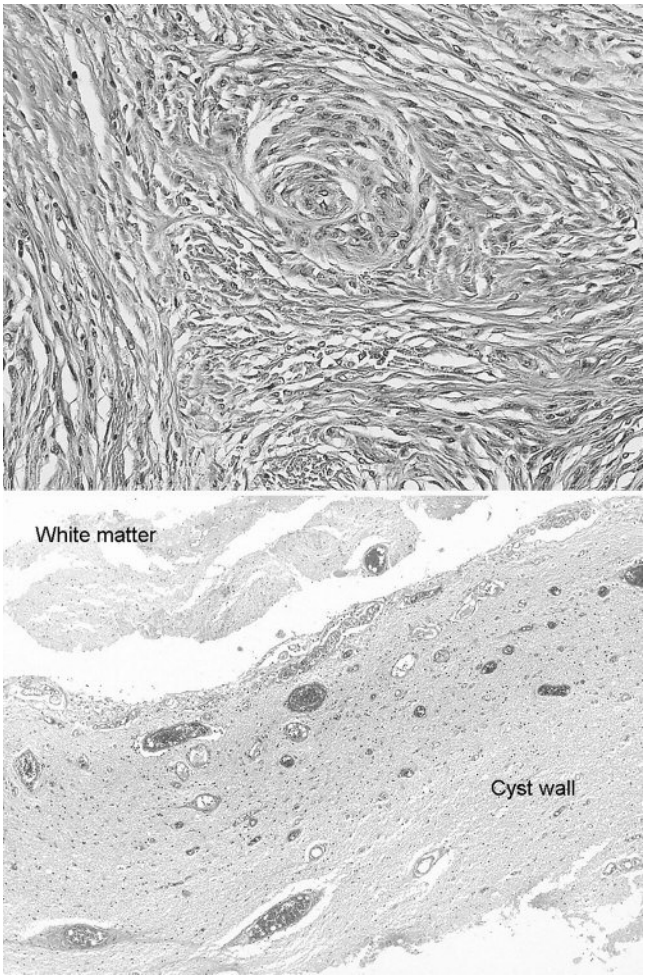

FIG. 2. Photomicrographs of the histological features of the solid *(upper)* and cystic *(lower)* portions of the tumor. *Upper:* The solid part of the tumor displays typical histological features with proliferation of spindle-shaped cells intermingling with a large amount of collagenous stroma showing storiform or whorled patterns. *Lower:* The cyst wall displays scattered small round cells such as astrocytes and lymphocytes, and small capillary vessels in edematous parenchyma. White matter adjacent to the cyst also demonstrates the same round cells. Capillary vessels are accumulated more in the outer cyst wall. H & E, original magnification × 100 *(upper)* and × 40 *(lower)*.

obscure or unenhanced cyst walls and 4 cases showing thickly enhancing cyst walls.[6,11,14,17] The cyst wall in the present case did not contain tumor cells in sections stained using H & E. Some reports have described histological features of the cyst wall. Kurose and coauthors[10] reported that a thickly enhancing cyst wall in a case of DIA did not contain tumor cells but showed scattered xanthoma cells with normal morphological features. Sugiyama and associates[20] reported 4 cases in a mixed series of DIA and DIG cases, and also noted that cyst walls were free of invading tumor cells. Kim et al.[7] observed no tumor cells within the septum of a multilobular cyst in a case involving a DIA. No reports have found tumor cells in the cyst wall of a DIA. An obscure and nonenhancing cyst wall in a DIA case showed macroscopically gliotic characteristics during surgery according to de Chadarévian and coworkers.[3] In the present case, macroscopic observation of the cyst wall revealed gliotic characteristics during surgery, and the cyst wall histologically demonstrated the features of gliosis, with numerous scattered round cells such as astrocytes and lymphocytes, and small vessels in edematous parenchyma. We speculate that enhancement of the cyst wall on neuroimaging is induced by accumulation of the small vessels within the area of gliosis, or increasing permeability of the blood–brain barrier in vessels due to gliosis. Kandalkar and colleagues[5] reported that the histology of the cyst wall in a case of a DIA showed normal but atrophic brain tissues in which the outer layer was increased due to gliosis, although results of neuroimaging were not demonstrated. They speculated that these histological changes in the cyst resulted from pressure of cyst fluid on the adjacent brain, and recommended excision of the cyst portion for seizure control after surgery. In the present case, gliosis was increased within the outer cyst wall, and was observed in adjacent white matter tissue. The border between the cyst wall and surrounding white matter was unclear. This adhesion between the cyst wall and surrounding white matter might be induced by aggressive gliosis within the outer side of the wall. The deep location, large size, and unclear border of the cyst may complicate removal of the cystic portion compared with the solid portion. Gross-total removal of a DIA or DIG leads to a favorable prognosis, but long-term survival is anticipated even with residual disease.[21,22] It needs to be emphasized that surgery for DIA only requires removal of the solid portion of the tumor when the cyst wall is nonenhanced on neuroimaging. This approach is similar to the surgical plan for a pilocytic astrocytoma, in which neurosurgeons generally assume that the cyst wall is not neoplastic or does not require resection.[2] Based on findings from the present case and the literature, we also believe that surgery for a DIA with a thickly enhancing cyst wall only requires removal of the solid portion of the tumor and that removal of the cyst wall is not necessary. If the residual cyst wall causes uncontrollable seizures after surgery, a second operation to remove the cyst wall should be considered.

Conclusions

Cyst characteristics of a DIA or DIG do not appear to have been fully explored in previous studies. Because surgery plays a large role in treatment for a DIA and DIG, characteristics of the cyst commonly located in the deep white matter under the solid part of the tumor represent important information for neurosurgeons. Clinical and histological findings in the present case and previous reports suggest that the cyst does not contain tumor cells, even if strong enhancement is seen on contrast-enhanced neuroimaging, and that a thickly enhancing cyst wall indicates gliosis with accumulation of numerous small vessels. The border between the cyst wall and surrounding white matter is unclear compared with the border between the solid portion and surrounding cortex. We therefore believe that the cyst wall does not require removal during surgery for a DIA, even when the cyst wall is thickly enhanced on neuroimaging. Further study of the cyst wall in a limited group of patients with a DIA or DIG is required to confirm our distinct impressions.

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