Desmoplastic cerebral astrocytoma of infancy (DCAI) was originally described by Taratuto et al. [23] as a distinctive astrocytic neoplasm occurring in children less than 1 year old. The prominent and distinguishing features of this rare neoplasm are its well-circumscribed dural-based cystic structure, biphasic histologic pattern consisting of neoplastic astrocytes embedded in a desmoplastic stroma and its favorable response to surgical treatment. We present another example of DCAI occurring in a 7 1/2-month-old infant. The presence of high mitotic rate, cellular pleomorphism and islands of undifferentiated small cells led us to perform ultrastructural, immunohistochemical and proliferative activity studies to further characterize the growth pattern of this rare neoplasm.

**Case report**

This 7 1/2-month-old oriental girl presented with a bulging anterior fontanelle and a head tilting to left following a 2-week history of low-grade fever, lethargy, irritability, and intermittent vomiting. She had recently recovered from an upper respiratory tract infection.

The patient had pre-existing right Erb's palsy since birth but otherwise had a normal development. Her head circumference was 50 cm, which was 4.5 cm greater than the 95th percentile for age. Mild bilateral sixth cranial nerve paresis was present.

On MRI scan a 3-cm enhancing dural-based nodule with a surrounding cyst was discovered in the right fronto-parietal region (Fig. 1). The lesion produced a midline shift with obstruction and dilatation of the left lateral ventricle. The suprasellar cistern was compressed and the brain stem was distorted with obliteration of the left paramesencephalic cistern.

Following drainage of the cyst, the tumor was removed via left frontal craniotomy. The patient has developed normally, and has no neurological deficit at follow-up examination 1 year after surgery.

**Materials and methods**

The tumor was fixed in formaldehyde-ethanol and embedded in paraffin. Sections (5 μm) were stained with H&E, Masson-Trichrome, Gordon-Sweet reticulin, and modified Bielschowsky. Immunohistochemical studies included staining for GFAP, synaptophysin (SYN), microtubule-associated protein 2 (MAP-2), neurofilament (68–200 kD; NF), desmin, vimentin, type-IV collagen, and laminin (see Table 1 for the technical data for the immunohistochemical markers). The sections were deparaffinized, rehydrated, washed in sodium citrate buffer. The endogenous peroxidase activity was terminated and nonspecific binding sites were blocked with fetal calf serum and incubated 18 h with the primer antibody. A standard streptavidin-biotin kit was used (Dako) followed by staining with diaminobenzidine and counterstaining with hematoxylin. For laminin staining, the tissue was treated with pronase.

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**Abstract.** We present a case of desmoplastic cerebral astrocytoma of infancy (DCAI) in a 7 1/2-month-old girl and include immunohistochemical, ultrastructural and proliferative activity studies. The dural-based cystic tumor showed a biphasic pattern consisting of glial fibrillary acidic protein (GFAP)-positive astrocytes embedded in a desmoplastic stroma. The astrocytic processes were lined with basal lamina at their surface contacting the collagen. Scattered islands of undifferentiated small cells were seen acquiring GFAP positivity at their peripheral zone facing the collagen. Studies with silver nucleolar organizer region and proliferating cell nuclear antigen disclosed a high proliferative activity. Flow cytometric study showed an elevated S phase and 15% hypertetraploid cell population. These findings contrast the favorable prognosis of the tumor at 26 months follow-up. Probably, extracellular-matrix-induced maturation of the undifferentiated cells with the formation of basal lamina may account for this unique disparity.

**Key words:** Dura – Cyst – Desmoplasia – Astrocytoma – Infant
The tumor was 3 cm in diameter. It was solid, bosselated and revealed a homogeneous whorled cut surface. Histologic examination revealed densely cellular tumor composed of spindle and polygonal, pleomorphic cells embedded within a dense fibroblastic stroma. The astrocytes were characterized by their rich glial fibers and branched and displaced the midline structures. Collagenous stroma. Several large islands composed of undifferentiated small cells were seen (Fig. 5). Mitotic figures were frequent, ranging three to four per high power field in cellular areas to one or two per ten high power fields in densely fibroblastic areas. The tumor contained a rich vascular network; however, there was no evidence of endothelial hyperplasia. Reticulin and Masson-Trichrome stains disclosed the rich reticulin and collagenous network entrapping the individual or groups of glial cells (Fig. 3). Bielschowsky staining did not show axonal structures within the tumor.

GFAP staining revealed both gemistocytic and fibrillary astrocytes arranged in storiform pattern (Fig. 4). The aggregates of undifferentiated cells were weakly positive for GFAP (Fig. 5). In a similar fashion to histologic counterpart, GFAP staining was consistently observed along the rim of the desmoplastic stroma either surrounding or penetrating through the islands of undifferentiated small cells (Fig. 5). Laminin and type-IV collagen staining outlined the profile of the astrocytic processes as well as delineating the vascular structures (Fig. 6). The astrocytes at the rim of the islands of undifferentiated cells were positive for laminin and type-IV collagen. Invaded islands of superficial cortex were sharply delimited from the tumor by a thin band of laminin and type-IV collagen staining. The reactive astrocytes in the normal cortex were negative with both markers (Fig. 6).

The markers to disclose the neuronal structures including NF, SYN and MAP-2 were completely negative. The small undifferentiated astrocytes were also unstained.

Electron microscope showed the biphasic pattern of the tumor composed of fibroblasts and astrocytes embedded in a rich collagenous stroma. The astrocytes were characterized by their irregular euchromatic nuclei with margined chromatin, rich endoplasmic reticulum, well-developed Golgi complexes, mitochondria, and abundant intermediate filament bundles (Fig. 7A). Most of the astrocytes, especially those with rich glial fibers were invested by basal lamina (Fig. 7B). Basal lamina formation was observed only when the glial processes were in contact with the collagenous stroma (Fig. 7B). Remarkably it was absent at the contact sites between the glial cells which often showed desmosome-like junctions. Scattered clusters of astrocytes with shorter cell processes showed no basal lamina formation. The fibroblasts had shorter processes without basal lamina formation (Fig. 7A). They were surrounded by collagen bundles and contained abundant ribosomes and granular endoplasmic reticulum. The vessels were lined by both fenestrated and nonfenestrated endothelial cells.

AgNOR counting revealed the following results: astrocytes: 6.07 per nucleus, fibroblast: 2.6 and endothelial' cells: 2.85 (Fig. 8). PCNA nuclear labeling was heterogeneous and varied from 10% to 50%, with an average 20% in most areas of the tumor (Fig. 9).

Image analysis showed a hypertetraploid (6N) cell population comprising 15% of the total cell population (318 cells) (Fig. 10A). Flow cytometry disclosed an elevated S-phase fraction (11.7%). However, the hypertetraploid cell population was not evident due to a dilution effect by the greater number of diploid cells (Fig. 10B).

Discussion

The first case similar to DCAI was reported by Bailey as "intracranial fibrosarcoma of the dura mater in childhood" [2]. The application of immunocytochemical methods have provided a precise histologic classification of neoplasms involving the pachymeninges, including primary and metastatic tumors. DCAI was described as a distinct pediatric neoplasm characterized by a GFAP-positive glial component intermixed within a dense dural stroma [23]. Further study on these cases revealed only astrocytic component without neuronal participation.