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Desmoplastic Infantile Ganglioglioma: CT and MR Features

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In 1987, VandenBerg and colleagues [1] reported 11 cases of an apparently distinct clinicopathologic entity: desmoplastic infantile ganglioglioma. The patients were all infants who presented with large tumors that showed evidence of glial and ganglionic differentiation accompanied by an extreme desmoplastic reaction. Routinely, the prognosis was excellent following either partial or complete tumor excision [2]. We recently encountered a child with a unique form of this entity, and we present here a correlational report of the CT, MR, surgical, and pathologic findings.

Case Report

A previously healthy 10-month-old girl was brought to the emergency room for treatment of a seizure. Her father had found her unresponsive in her crib making rhythmic jerking movements of her left arm and leg. Her seizures stopped after she received phenytoin

and lorazepam. The child's development had been normal and the family history was unrevealing.

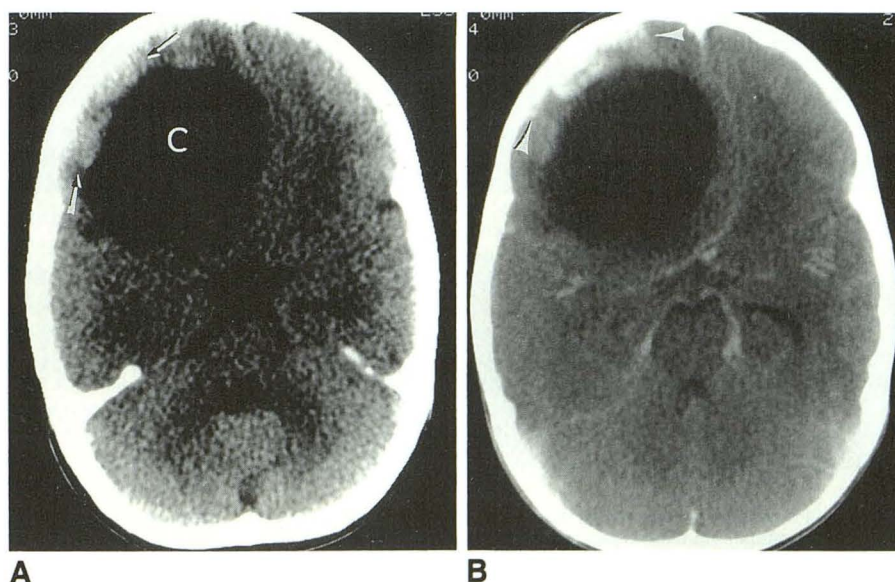
Initial physical examination showed an obtunded child who responded to stimuli by localizing with all extremities. She was afebrile. Her head circumference was 46.75 cm (90th percentile) with a soft flat anterior fontanelle. She was marginally hyperreflexic on the left with Babinski sign present ipsilaterally. The rest of her physical examination was normal.

CT (Fig. 1) and MR imaging (Fig. 2) demonstrated a large right frontal mass consisting primarily of a well-defined cystlike area as well as a smaller solid component along the cortical margin. The preoperative radiologic diagnosis was complex neoplasm, most likely primitive neuroectodermal tumor (PNET).

At surgery, a poorly vascularized tumor was identified, which was inseparable from the dura. Under the mass was a large, well-circumscribed cyst that collapsed spontaneously. After total resection of the mass, the child recovered and returned home on the fifth post-operative day. The patient received anticonvulsants but no radiation or chemotherapy and was neurologically normal at 6 months follow-up. A repeat CT scan showed no evidence of recurrence.

Fig. 1.—A, Noncontrast CT scan shows large mass in right frontal lobe that extends to meningeal surface. It contains two distinct areas of density. The larger of the two is well defined, hypodense, and cystlike (C). Along its cortical margin is a solid-appearing rim (arrows), which is slightly hyperdense relative to the analogous frontal gray matter on the left.

B, CT scan obtained after administration of contrast material shows intense enhancement of a subcalvarial segment (arrowheads) and no enhancement of cyst wall.



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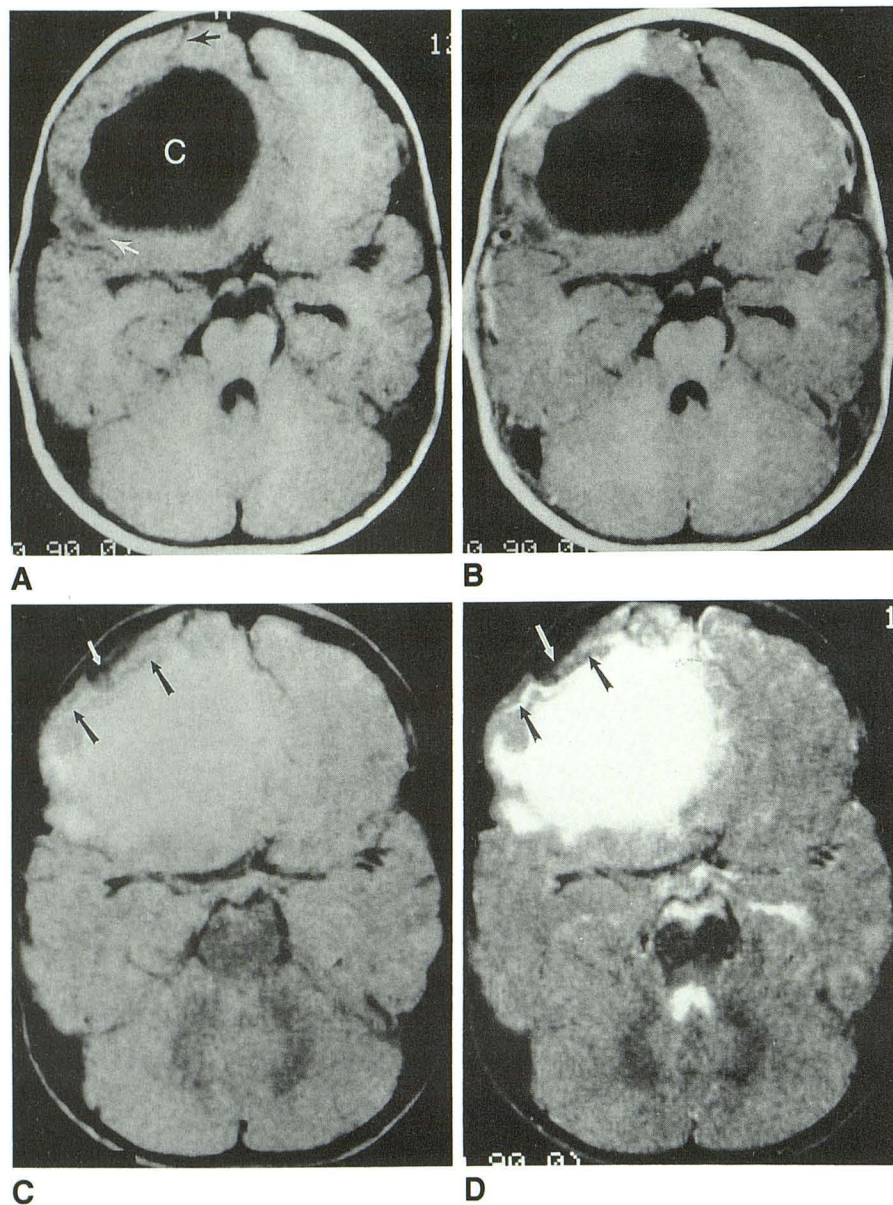


Fig. 2.—A, T1-weighted (600/50/3) axial MR image at 0.38 T shows a large hypointense area (C) with a variable but mainly isointense peripheral tumor (arrows).

B, T1-weighted (600/50/3) contrast-enhanced axial MR image shows area of intense contrast enhancement. Notice there is no enhancement of the cyst.

C and D, Balanced (2800/50/1) (C) and T2-weighted (2800/100) (D) axial MR images show T2 prolongation of the cyst and variable signal intensity of the tumor (arrows).

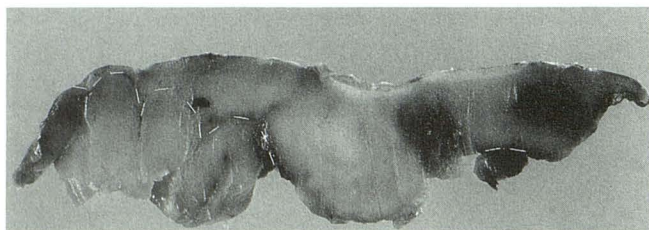


Fig. 3.—Photograph of pathologic specimen of tumor's cut surface shows fibrotic rind of leptomeningeal tumor and its relationship to underlying brain. The junction between the two (dashes) was discrete in some areas and indistinct in others.

The tissue resected from the right frontal lobe had a distinctive gross appearance (Fig. 3). The specimen measured $5.5 \times 5.5 \times 2.5$ cm and was ovoid in configuration. An outer fibrotic rind was tan-gray, measured up to 0.8 cm in depth, and had a coarse texture

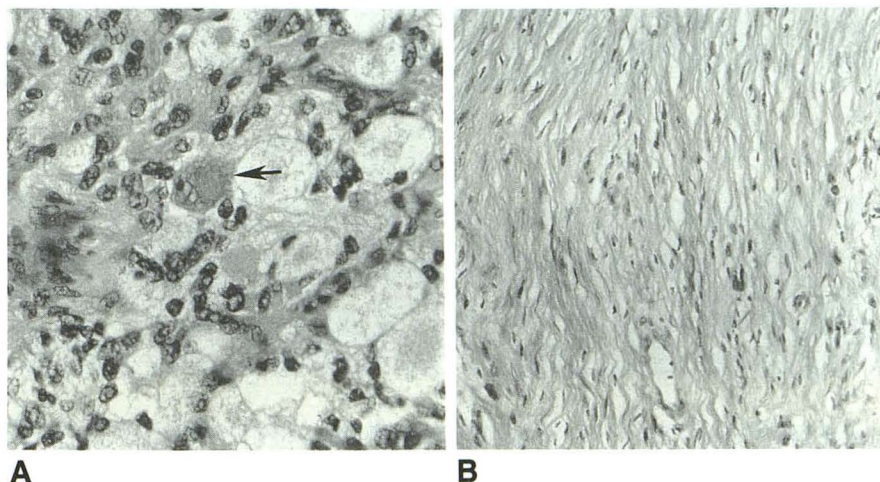
when cut. The cerebral cortical tissue deep to the rind was discolored with a yellow-gray appearance but showed evidence of gyral-sulcal patterning. A portion of a cyst measuring 2.0 cm in diameter was noted in the brain tissue on the undersurface of the specimen. Its wall was smooth and continuous with the brain tissue and showed no evidence of a lining. The cyst had no connection to the rind on the outer surface.

Microscopically, the dense rind corresponded to thickened and fibrotic leptomeninges involved with tumor (Fig. 4). Intraparenchymal involvement by tumor was limited to a few microscopic foci of tumor cells extending into the perivascular Virchow-Robin spaces of the superficial cortex. The cyst wall was composed of gliotic brain parenchyma and showed no evidence of neoplasm.

Discussion

Gangliogliomas are uncommon benign tumors [2]. Most are found in patients over 10 years old who present with

Fig. 4.—A and B, Photomicrographs show varied appearance of tumor. Cellular areas of tumor (A) contain a mixture of neuronal (arrow) and glial cells. Desmoplastic areas of tumor (B) contain spindled fibroblasts and a densely collagenized background.



seizures and headaches [3]. Desmoplastic infantile gangliogliomas (DIGs) are an uncommon variety of gangliogliomas that occur exclusively in infants. They are located in the frontal or parietal lobes. They are usually large tumors with two distinct components. There is invariably an associated cystic area that is usually an integral part of the tumor or, as in our case, a reaction to it; it often represents the majority of the visible abnormality. A solid component containing the desmoplastic reaction is always located adjacent to the meninges [1]. Although contrast enhancement is variable in gangliogliomas [2], CT scans in DIGs show intense contrast enhancement extending to the meninges [1, 4].

DIGs are a distinctive form of neonatal tumor. PNETs can occur within the frontal lobe of infants; however, they are usually deeply seated with frequent calcification and partial cyst formation. They have a poor prognosis. Supratentorial ependymomas can also occur in the frontal lobe and may have cystic areas; however, they commonly have calcification and heterogeneous contrast enhancement. Astrocytomas are frequent childhood tumors but are less common in neonates. They often contain cystic areas and may have marked contrast enhancement; but when cystic, they usually have a mural enhancing nodule. Rare cerebral astrocytomas with prominent leptomeningeal growth have been reported in infants [5]. When large enough, all these tumors may have imaging appearances identical to DIGs. Most investigators believe that the histology and clinical behavior of gangliogliomas are that of a benign neoplasm, although arguments have been raised that it might be dysplastic [6].

This case is unique because there was no significant intra-

parenchymal tumor mass. The cyst had no neoplastic component and we believe this represents a reactive or degenerative phenomenon related to the overlying mass.

The variable signal pattern of this patient's tumor may be explained by the pathologic findings. The large area of T2 prolongation represented the cyst. The area of intense contrast enhancement represented the tumor with varying amounts of intermixed desmoplasia.

The salient imaging features of the desmoplastic ganglioglioma of the neonate are (1) a bulky tumor involving the frontal and/or parietal lobe, (2) a tumor that abuts a meningeal surface, (3) a large cystic component, and (4) contrast enhancement along the meningeal side of the mass.

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