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Congenital Malignant Gliosarcoma

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Malignant gliosarcoma is an extremely rare, anaplastic tumor which to date has been found exclusively in adults. This type of sarcoma is a form of mixed glioma and sarcoma which is quite different and distinct from the type of sarcoma that arises in a glioblastoma [1]. Less than 10 cases of this entity have been reported [2–5]. We report a case of this unusual tumor in a 4-month-old girl who had lethargy and macrocephaly. Although the clinical, radiographic, and angiographic manifestations of intracranial sarcomas have been described [1–10], the computed tomographic appearance of a primary malignant gliosarcoma has not been published.

Case Report

A 4-month-old girl was noted to have a large head at birth. At age 16 weeks she had lethargy and bulging anterior fontanelle. She was the product of an uneventful 38 week gestation. Her mother (para 1, gravida 1) denied the use of drugs, ethanol, or cigarettes during the pregnancy.

The physical examination failed to reveal any focal neurologic deficits. The head circumference was 46 cm, which far exceeds the 95th percentile for her age. At birth her head measured 35.5 cm, which is at the 90th percentile level.

Unenhanced CT (fig. 1A) demonstrated a large cystic left hemispheric mass obliterating the left lateral ventricle, shifting the midline structures to the right, with moderate dilatation of the right lateral ventricle secondary to obstruction at the foramen of Monroe. The cyst fluid measured 10–12 Hounsfield units (H) above the cerebrospinal fluid values. The cyst was surrounded by focal regions of increased density secondary to calcification in tumor nodules. With contrast administration (fig. 1B), the entire cyst wall showed prominent, nonhomogeneous enhancement. The enhancing margins appeared thin and delicate medially and quite thick laterally and superiorly.

The CT was the only radiographic study required preoperatively. At surgery a large cystic mass replacing most of the left cerebral hemisphere was found. The cyst wall had a white granular appearance. The cyst contents contained viscous, brown, proteinaceous fluid.

Microscopically the tumor was highly cellular and extremely pleomorphic. In some areas it was composed of closely packed, spindle-shaped cells with small nuclei and bipolar tapering processes. Abundant reticulin fibers were present around tumor cells in these foci (fig. 1C). Other areas contained sheets of variable-size cells with prominent eosinophilic hyaline cytoplasm (fig. 1D). Many

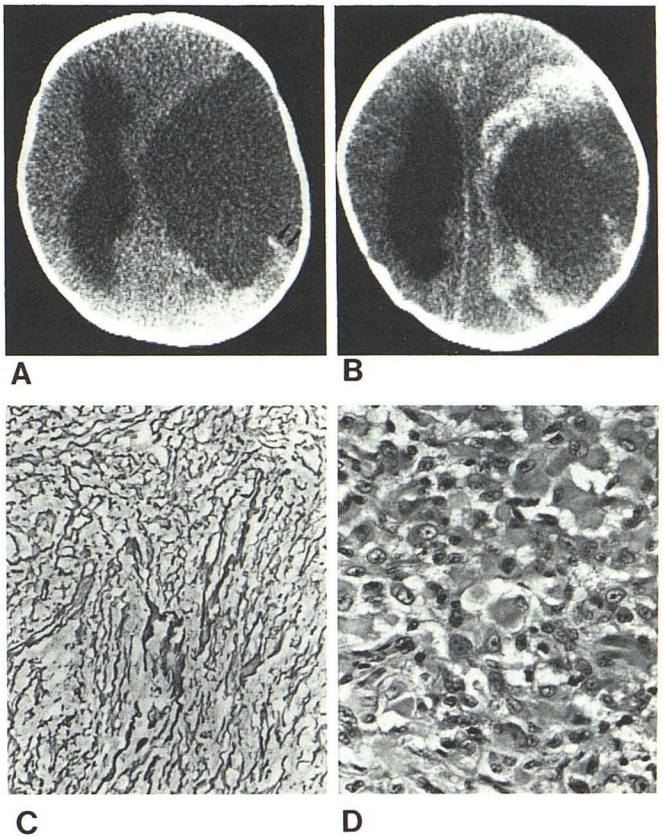

Fig. 1.—**A**, Noncontrast scan. Large cystic mass in left hemisphere effaces left lateral ventricle and causes shift of midline structures. Focal regions of calcifications within cyst wall (*arrows*). **B**, Contrast scan. Thick, nonhomogeneous rim of enhancement surrounds cyst. **C**, Abundant interconnecting reticulum fiber network surrounds individual tumor cells and several smaller blood vessels. Reticulin stain ×370. **D**, Sheets of tumor cell with variable size nuclei and prominent hyaline cytoplasm. Many of these cells contained glial fibrillary acidic protein characteristic of gliosarcoma. H and E ×600.

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of these cells contained glial fibrillary acidic protein. The neoplasm also showed abundant mitotic figures, small and large foci of necrosis, microcalcification, and a few foci of endothelial proliferation.

Discussion

Malignant gliosarcoma is a truly rare neoplasm previously reported only in adults. It usually spreads by infiltration of the contiguous brain and has a very poor prognosis. Histologically this tumor demonstrates abundant reticular fibers and multiple mitotic figures. Unlike a glioblastoma with sarcomatous elements, the tumor contains little collagen and has a paucity of blood vessels [1, 2].

The CT appearance of this sarcoma in our patient is quite striking. The tumor surrounding the cyst before contrast administration is hyperdense secondary to areas of microcalcification demonstrated histologically. The marked enhancement of the tumor after contrast administration proved to be a reliable indicator of its malignancy [11, 12].

This case is of further interest in that it almost certainly represents a true congenital intracranial neoplasm, as the patient's head was large at birth. In a review of the literature, Solitare and Krigman [13] found only 26 cases of congenital brain tumors that fulfilled their criteria of producing signs or symptoms at birth. Only one congenital cerebral sarcoma has been reported to date and this was an angiosarcoma and not a malignant gliosarcoma as in our case [14].

With the widespread use of CT, most primary intracranial neoplasms in children can be readily detected without the morbidity associated with angiography and pneumoencephalography. Although histopathologic diagnosis is not possible with CT alone, it does provide a safe, noninvasive method for preoperative diagnosis and postoperative follow-up in the neonate and very young child with an intracranial neoplasm.

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