

Giant Infantile Gliosarcoma: Magnetic Resonance Imaging Findings

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Gliosarcoma is an uncommon variant of glioblastoma multiforme, which is composed of gliomatous and sarcomatous elements. The tumor is rarely encountered in childhood. This case report presents the magnetic resonance imaging

characteristics of a giant gliosarcoma in a 3-year-old girl. Size and location of the tumor are described.

Keywords: giant infantile gliosarcoma

Gliosarcoma is a rare variant of glioblastoma multiforme, which is composed of gliomatous and sarcomatous elements.¹⁻⁵ While some authors support a sarcomatous evolution from vascular elements in glioblastoma multiforme, others proposed a monoclonal origin because of some genetically similar properties that exist in gliomatous and sarcomatous components of the tumor.^{1,2,4}

Although gliosarcomas are generally seen in men in their sixth and seventh decades, 10 cases have been described in children.⁴ The reported gliosarcoma cases in children originated in the cerebral hemispheres.¹⁻⁵

In this report, we present the case of a giant gliosarcoma in a 3-year-old girl. Magnetic resonance (MR) imaging, proton MR spectroscopy, and perfusion MR imaging findings are described.

Case Report

A 3-year-old girl presented to her pediatrician with right lateral gaze palsy, ptosis, and weakness in her left arm and leg. With the suspicion of an intracranial mass, MR imaging was performed. Images revealed a giant mass filling the prepontine and suprasellar cisterns with mass effect uplifting the floor of the third and left lateral ventricles. The lateral ventricles were dilated because of obstruction at the foramina of Monro. The tumor also abutted the left lentiform nucleus. The mass was isointense

with gray matter in T1- and T2-weighted sequences and was homogeneously enhanced after administration of paramagnetic contrast agent (Figure 1A-C). On diffusion-weighted images, the tumor was slightly hyperintense compared with the white matter, with mean (SD) apparent diffusion coefficient values from the tumoral region and from the peritumoral region of $0.77 (0.35) \cdot 10^{-3} \text{ mm}^2/\text{s}$ and $0.82 (0.23) \cdot 10^{-3} \text{ mm}^2/\text{s}$, respectively (Figure 1D). On perfusion images, the tumor showed increased perfusion compared with normal cerebral white matter (Figure 1E). The normalized regional cerebral blood volume ratio of the tumoral region was 10.5. The peritumoral regional cerebral blood volume ratio was 4.5. On MR spectroscopy, the tumoral area showed increased choline and lactate values, with decreased N-acetylaspartate and creatine values (Figure 1F).

After surgical excision of the mass, histopathologic examination showed a cellular tumor containing neoplastic cells with moderate to highly pleomorphic nuclei. It was composed of intimately intermixed glial and sarcomatous components. The sarcomatous component showed a prominent pericellular reticulin network. Reticulin was scant in the glial component. Collagen deposition in sarcomatous areas was demonstrated by a trichrome stain. In some areas, the distinction between glial and sarcomatous areas was difficult to differentiate without immunohistochemistry and reticulin stain. There was florid microvessel proliferation and necrosis with peripheral palisading. Mitotic activity was high (12 mitoses per 10 high-power fields). Although glial fibrillary acidic protein was present in the glial component, it was absent from the sarcomatous component. The Ki-67 proliferation index was approximately 10%. The final histopathologic diagnosis of the tumor was gliosarcoma (Figure 2).

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Discussion

Gliosarcoma is a mixed tumor consisting of gliomatous and sarcomatous elements.^{1,2} While it was formerly considered

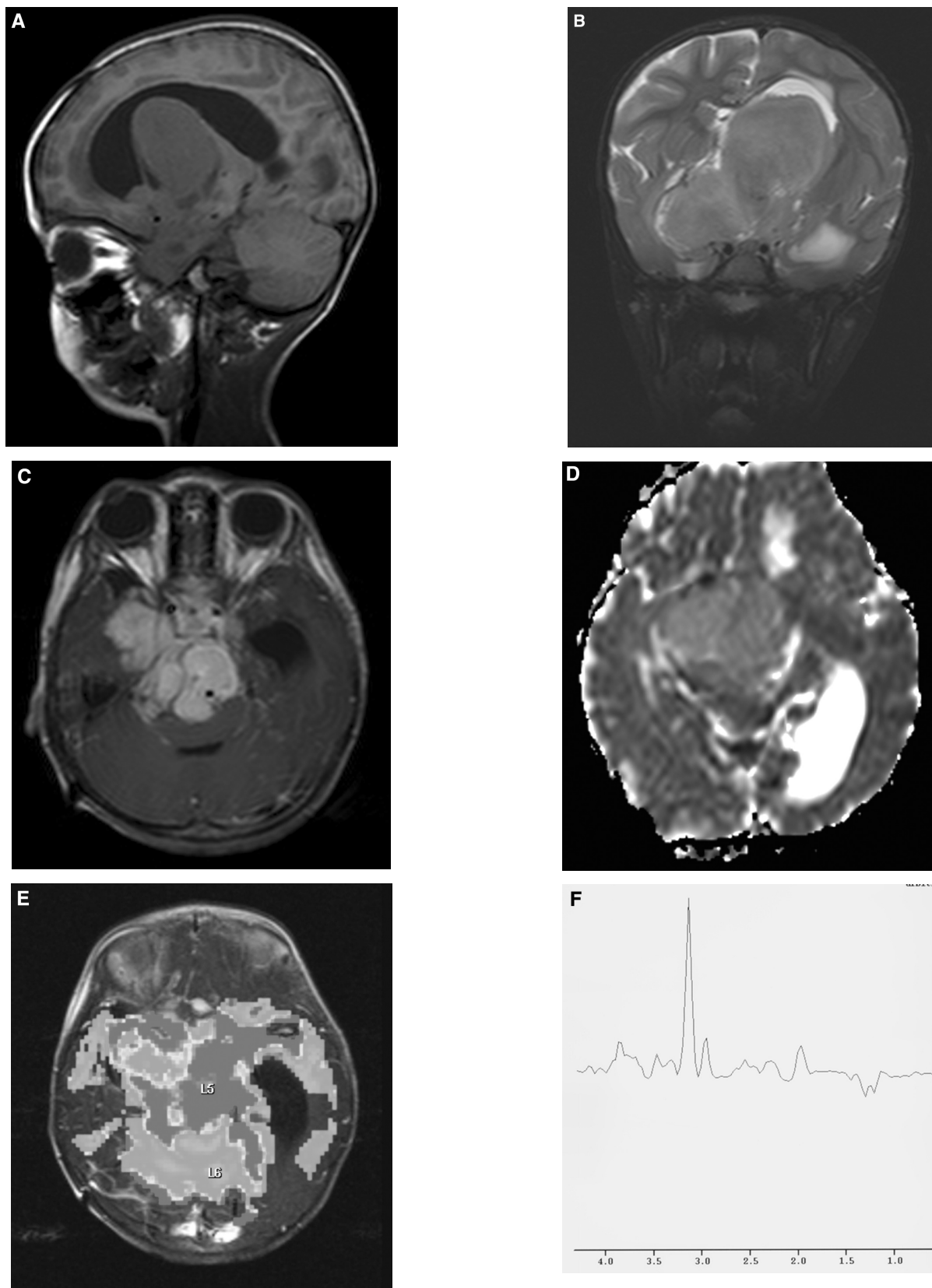


Figure 1. A 3-year-old girl with a giant mass lesion obliterating the prepontine and suprasellar cisterns. The mass is isointense with gray matter on axial T1-weighted (repetition time, 583 milliseconds; echo time, 15 milliseconds) (A) and T2-weighted (repetition time, 8000 milliseconds; echo time, 100 milliseconds) (B) spin-echo images. C, Axial postcontrast T1-weighted image shows that the tumor enhances homogeneously. D, On diffusion-weighted image, the tumor is slightly hyperintense compared with the white matter. E, Perfusion image (repetition time, 626 milliseconds; echo time, 30 milliseconds) shows increased perfusion in the tumor. F, Magnetic resonance spectroscopy spectra from the tumoral area show decreased N-acetylaspartate with increased choline and prominent lactate peaks.

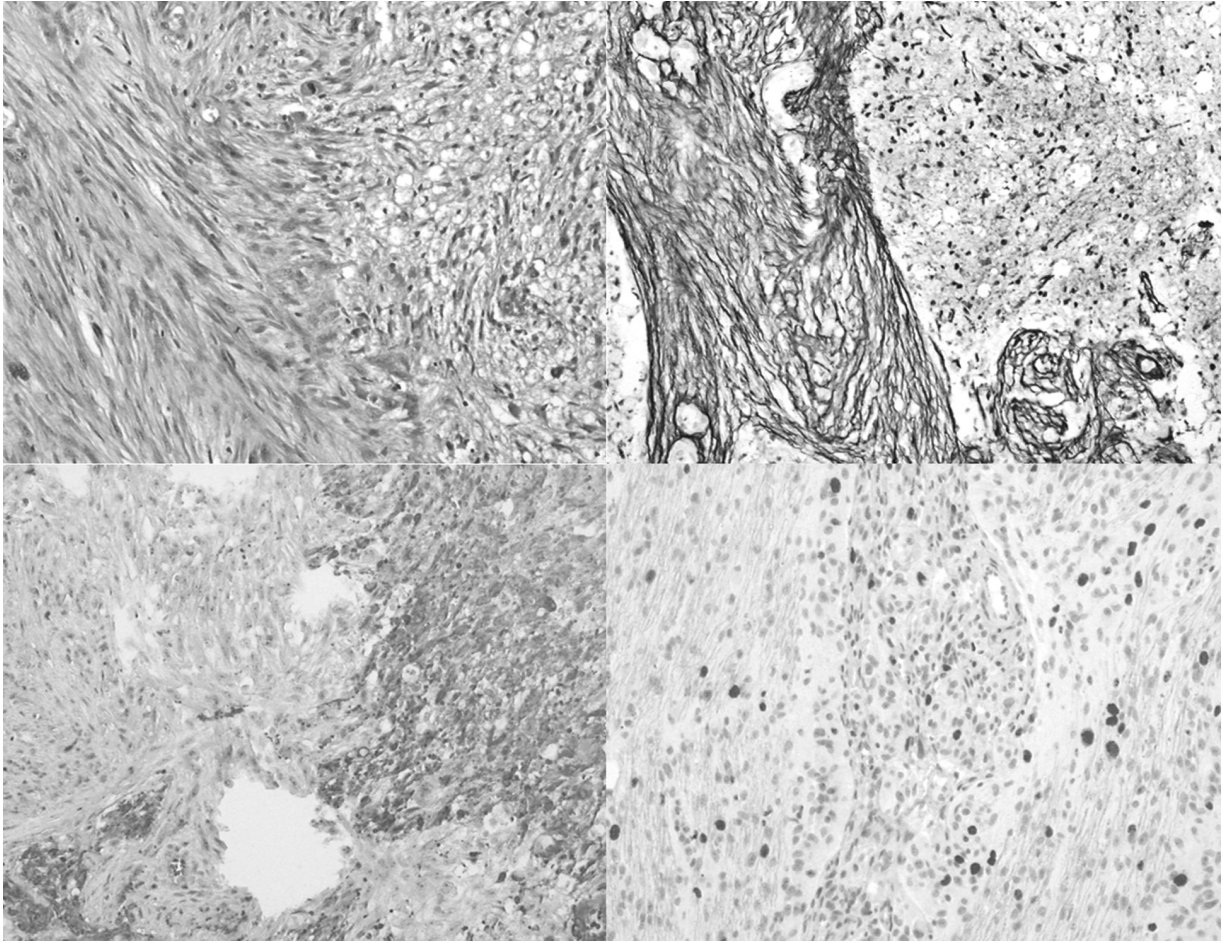


Figure 2. Intimately intermixed glial and sarcomatous components (upper left). Prominent pericellular reticulin network in the sarcomatous component and scanty reticulin in the glial component (upper right). Glial fibrillary acidic protein is present in the glial component (lower left). The Ki-67 proliferation index is approximately 10% (lower right).

glioblastoma having sarcomatous elements, with the introduction of the new World Health Organization classification,⁶ it is now referred to as gliosarcoma. Some authors have proposed its sarcomatous evolution from a vascular component in a malignant glioblastoma.¹⁻⁵ Genetic findings have indicated a monoclonal origin because of similar properties (eg, p53 mutations and p16 deletion) seen in sarcomatous and glial elements.^{1,2,4}

The incidence of gliosarcoma among all malignant gliomas is reported to be 2% to 10%.^{1,3,4} Like glioblastoma multiforme, gliosarcoma is most frequently seen in the sixth and seventh decades, with infantile cases being rare.⁴

Frequently originating in temporal lobes with a supratentorial localization, gliosarcomas tend to be in the vicinity of dura, falx, and skull.⁷ A mostly suprasellar localization, as in our patient, has not previously been reported in the literature, to our knowledge.

MR imaging characteristics of gliosarcoma in cerebral hemispheres have been reported to be variable.^{3,7}

Although they are generally isointense with gray matter in T2-weighted sequences, a heterogeneous appearance in T1- and T2-weighted sequences can be observed because of necrosis and hemorrhage. The solid mass in our patient was homogeneous and isointense with gray matter in all sequences and homogeneously enhanced after contrast agent injection.

High-grade gliomas and metastases have apparent diffusion coefficient values ranging from $0.82 \cdot 10^{-3} \text{ mm}^2/\text{s}$ to $1.4 \cdot 10^{-3} \text{ mm}^2/\text{s}$, without any major difference between their peritumoral apparent diffusion coefficient values.⁸ The mean (SD) apparent diffusion coefficient value from the tumoral region of $0.77 (0.35) \cdot 10^{-3} \text{ mm}^2/\text{s}$ in our case suggests a high-grade tumor. The tumor has decreased *N*-acetylaspartate and increased choline and lactate peaks and high regional cerebral blood volume ratio values. These features are also consistent with a malignant process. The substantial obliteration of the suprasellar cistern in our patient suggests that the mass originates in the suprasellar region,

with the same signal intensity characteristics as rhabdoid or teratoid tumors, primitive neuroectodermal tumors, and germ cell tumors.

In conclusion, although rare, gliosarcoma should be included in the differential diagnosis during early infancy if MR imaging reveals any of the following: a mass obliterating the suprasellar cisterns, an isointense mass with cerebral parenchyma in T1- and T2-weighted sequences, and a homogeneously enhancing mass with malignant findings on proton MR spectroscopy and perfusion studies.

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