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Case Report

A case of gliosarcoma in a child with neurofibromatosis type 1

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Abstract

Gliosarcoma (GS) is a rarely seen form of glioblastoma. These tumors are mostly seen in males older than 60 years of age. It is extremely rare in pediatric central nervous system (CNS) tumors. In this case report, we present a 3-year-old boy with a giant gliosarcoma. Magnetic resonance (MR) imaging and histopathologic findings are discussed. A 3 year-old boy with a clinically diagnosed NF-1 was admitted to the emergency department with a complaint of intractable vomiting. Magnetic resonance imaging (MRI) of the brain was suggestive of a large lobulated mass lesion in the left parietal lobe extending to the vertex and slightly compressing the left lateral ventricle. The final histopathologic diagnosis of the tumor was considered as gliosarcoma. To our knowledge, this case constitutes the first youngest case with neurofibromatosis type 1 reported in the literature in all pediatric cases of GS.

Keywords: Gliosarcoma; magnetic resonance imaging; neurofibromatosis 1

INTRODUCTION

Gliosarcoma (GS) is a rarely seen form of glioblastoma, containing gliomatous and sarcomatous elements (1). It is considered as a variant of glioblastoma and as a grade 4 tumor according to World Health Organisation (WHO) classification (2). Gliosarcomas are extremely rare in pediatric population. These tumors are mostly seen in males older than 60 years of age (1). Although the relative frequency of pediatric GS among glioblastoma is 1.9%, it is extremely rare in pediatric central nervous system (CNS) tumors with a relative frequency 0.5% (3).

In this case report, we present a 3-year-old boy with a giant gliosarcoma. Magnetic resonance (MR) imaging and histopathologic findings are discussed.

To our knowledge, this case constitutes the first youngest case with neurofibramatosis type 1 reported in the literature in all pediatric cases of GS.

CASE REPORT

A 3 year-old boy with a clinically diagnosed NF-1, who had been under control since he was 5 months old because of multiple café au lait lesions was admitted to the emergency department with a complaint of intractable vomiting. He had no other complaints such as weight loss or fewer. The neurologic examination of the patient was normal. There were no pathological findings in his brain MRI 1 year ago. His sister had died because of a renal tumor.

Magnetic resonance imaging (MRI) of the brain was suggestive of a large lobulated mass lesion measuring 6 × 5,5 × 5 cm in the left parietal lobe extending to the vertex and slightly compressing the left lateral ventricle. The lesion was mildly hypointense compared to gray matter in T1 weighted images and heterogeneous hyperintense in T2 weighted images (figure 1). There were T1 hyperintense, T2 hypointense focuses at the central part of the lesion suggesting hemorrhage (figure 1). Following administration of intravenous contrast there was significant enhancement of the lesion except the central portion (figure 2). On diffusion weighted images (DWI), the central part of the tumor was hypointense whereas the periphery of the tumor was hyperintense (Figure 3). On apparent diffusion coefficient (ADC) images the periphery of the lesion was hypointense indicating diffusion restriction (figure 3). The mean (SD) ADC value calculated from the periphery of the lesion was 0.84 ×10−3 mm2/s (Figure 4).

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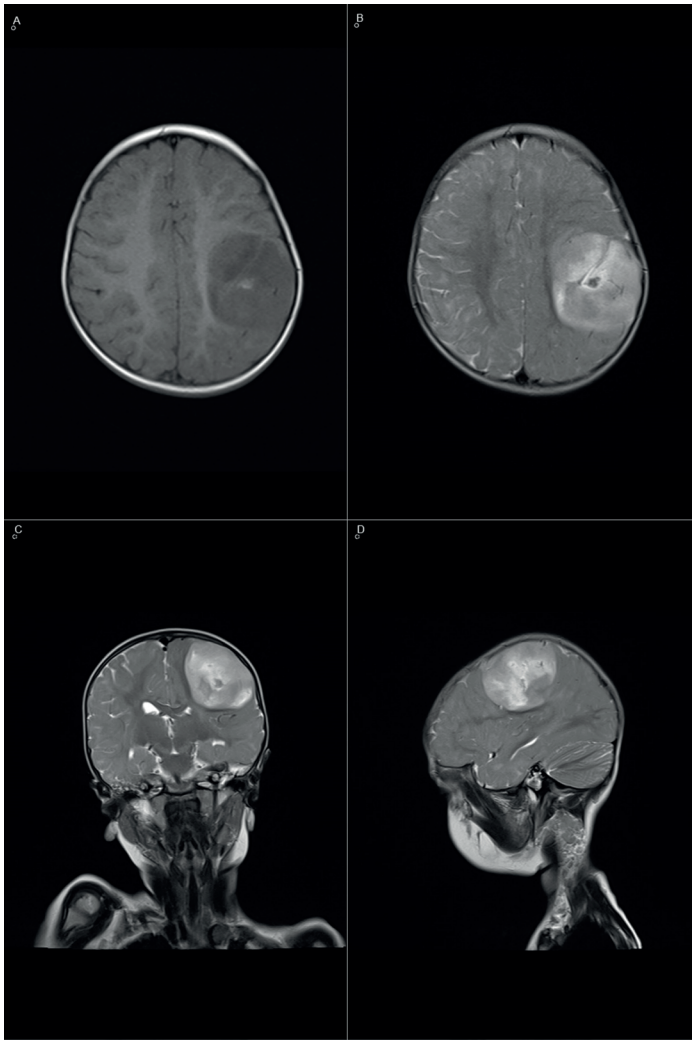

Figure 1 A. Axial T1-weighted image B. Axial T2-weighted image C. Coronal T2-weighted image D. Sagittal T2-weighted image. A 6 × 5.5 × 5 cm lesion in the left parietal lobe, mildly hypointense in T1 weighted images and heterogeneous hyperintense in T2 weighted images. T1 hyperintense, T2 hypointense focuses at the central part of lesion

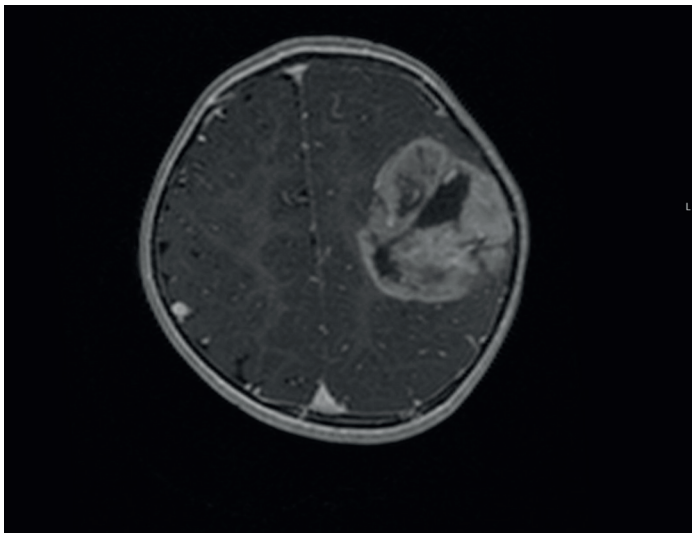

Figure 2. Axial postcontrast T1-weighted image
Significant enhancement of the lesion except the central portion

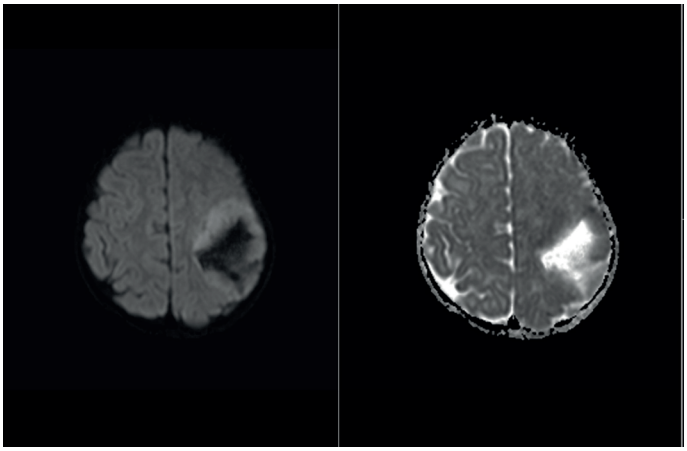

Figure 3. Diffusion-weighted image (DWI).
There is a diffusion restriction at the periphery of the lesion

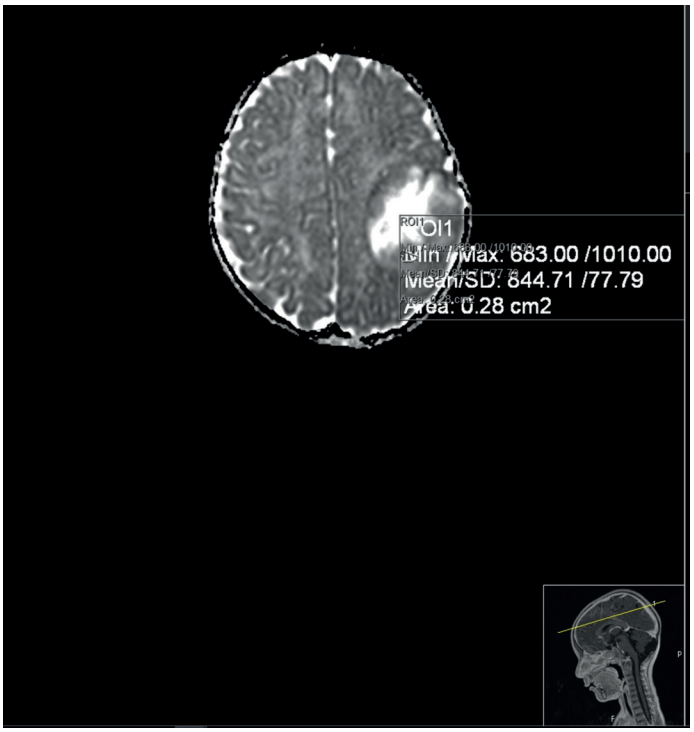

Figure 4. Apparent diffusion coefficient (ADC) images

After palliative surgical resection of the tumor, a highly cellular tumor containing neoplastic cells with neurophylic stroma, narrow cytoplasm and big vesicular hyperchromatic nuclei was seen in histopathologic examination (Figure 5a). There were cells with gemistosit nature and prominent atypical multinucleated cells in the tumor (Figure 5b). Necrotic areas and atypical mitotic figures were also seen (figure 5 c). There were tumor cells with relatively more elongated cytoplasm alining like fascicles, suggesting mesenchymal differentiation. GFAP (Figure 6a), smooth muscle actine (50%) (figure 6 b), INI-1, S-100 were stained positive and Myogeninchromogranine A, NSE, PanCK were stained negative in immunohistochemical staining. The Ki-67 proliferation index was approximately 80% (Figure 6c). The final histopathologic diagnosis of the tumor was considered as gliosarcoma.

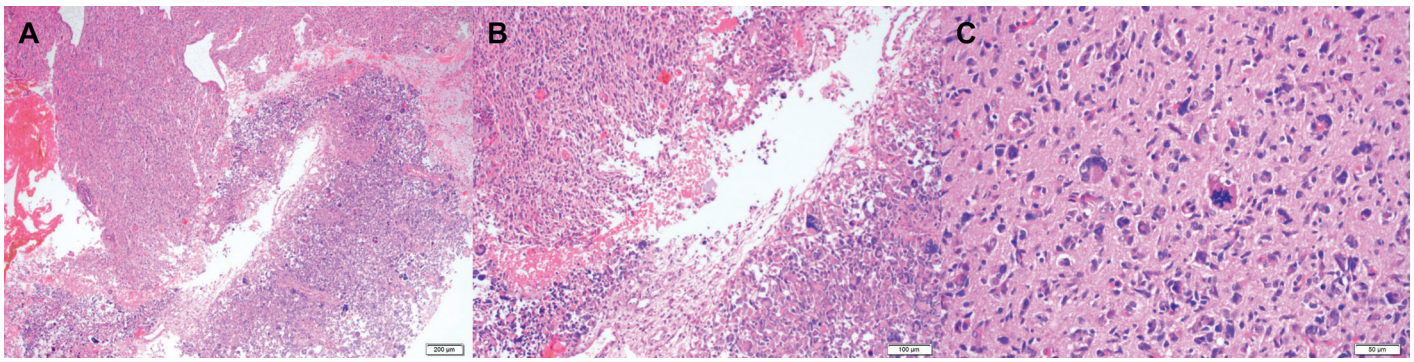

Figure 5. A. H&Ex40; Neoplastic cells with neurophylic stroma, narrow cytoplasm and big vesicular hyperchromatic nuclei, B. H&Ex100; Cells with gemistosit nature and atypical multinucleated cells, C. H&Ex200; Necrotic areas and atypical mitotic figures

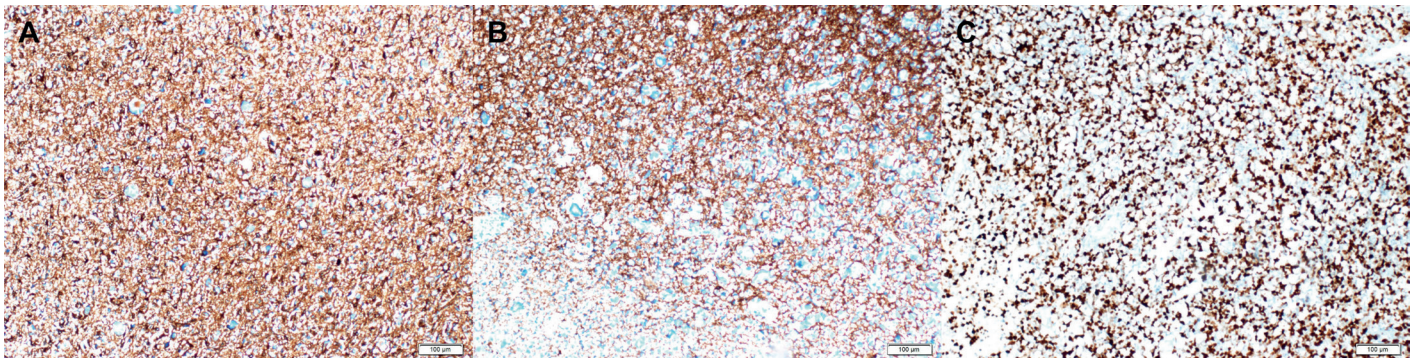

Figure 6. Immunohistochemical staining A; GFAPx100, B; SMAx100, C; Ki-67x100

Chemoradiotherapy was planned for the further treatment.

The family of the patient had given an informed consent for participation in this study.

DISCUSSION

Gliosarcoma is a mixed tumor consisting of gliomatous and sarcomatous elements, which was formerly considered sarcomatous form of glioblastoma (1). After the introduction of the new WHO classification (4) it is now classified as gliosarcoma. This tumor usually occurs in adults and older age and also very rare in children (1). When we searched the literature, we found only 25 pediatric cases and the average age of these cases was eleven (5). As a result of our research, we saw that this case constitutes one of the two youngest cases reported in all pediatric cases of GS. The former case reported was also a 3 year old child with GS in suprasellar localization (6).

On the other hand, CNS neoplasms can be seen in patients with neurofibromatosis type 1 (NF 1) which is a relatively common hereditary syndrome. The most common tumors accompanying NF 1 are the pilocytic astrocytomas (7). Gliosarcomas with NF-1 have rarely been reported. To date, 4 cases have been reported in the English written literature (8). Our patient is the youngest reported patient with NF-1 accompanying gliosarcoma in the literature.

Reported MR imaging characteristics of gliosarcoma in cerebral hemispheres are variable (9). In T2 weighted images, these tumors are generally isointense with graymatter. Because of the necrosis and the hemorrhage, in T1 and T2 weighted images heterogeneous appearance can also be observed. The tumor in our patient was heterogeneous hyperintense in T2 weighted sequences due to hemorrhage. In high-grade gliomas and metastases, the apparent diffusion coefficient values range between 0.82×10−3 mm2/s and 1.4×10−3 mm2/s (10). The calculated mean ADC value from the peripheric region of the tumor was 0.84 ×10−3 mm2/s in our case suggesting a high-grade tumor.

CONCLUSION

In conclusion, we can say that, if the brain tumors are radiologically high-grade in children with neurofibromatosis type 1, gliosarcoma should be considered.

Conflict of interest: The authors declare that they have no competing interest.

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Patient informed consent : The family of the patient had given an informed consent for participation in this study.

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