Case Report

A rare, giant, lateral intraventricular gliosarcoma ♠

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A REVIEW

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ABSTRACT

A peculiar subtype of glioblastoma multiforme is gliosarcoma. According to histological analysis, it possesses a biphasic shape that clearly distinguishes between glial and sarcomatous tissue. A gliosarcoma is often seen in the supratentorial region. Here, we describe a rare instance of giant left intraventricular gliosarcoma that manifested in a 1-year-old male. Advanced brain scan using magnetic resonance imaging identified a supratentorial tumor with radiological characteristics comparable to choroid plexus carcinoma. Histopathology determined that the tumor was a gliosarcoma. Despite its rarity, gliosarcoma should be taken into consideration when determining the differential diagnosis of intraventricular tumors in children who exhibit radiological signs of choroid plexus carcinoma.

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Introduction

The current (2021) World Health Organization (WHO) classification of central nervous system tumors still recognizes gliosarcoma as a classic variety of glioblastoma along with epithelioid and giant cell glioblastomas, despite the fact that they are not different diagnoses. They are primary intra-axial neoplasms that are extremely malignant (WHO grade 4), and they contain both glial and mesenchymal components [1]. It is noted that 2%-8% of all initial glioblastomas are gliosarcomas, a rare and uncommon kind of aggressive malignancy [2]. It is essentially a subtype of glioblastoma with glial and sarcomatous components that have differentiated separately and divergently from one another. This phenomenon was first described by Stroebel et al in 1895 [3] and later, in 1955, Feigen et al. provided a detailed description of it [4]. It is mostly found in the frontal and temporal lobes of the cerebral hemispheres. In rare cases, it can also be detected intraventricularly [2,5]. Clinically and radiologically, glioblastoma and gliosarcoma are related. It demonstrates a preference for men between the ages of 40 and 60. If the patient is not treated, it results in a poor clinical prognosis with an average survival of fewer than 6 months [2-5]. To the best of our knowledge, this article describes the exceptionally unusual case of giant lateral intraventricular gliosarcoma.

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A 1-year-old male, suffering from 1-month headache, nausea, and vomiting, was admitted to the department of neurosurgery, Children's Hospital 2. Her medical profile manifested no abnormalities. Neurological deficits were not detected during clinical evaluation. The routine laboratory tests and tumor markers were within normal ranges. Brain magnetic resonance imaging with contrast agent was immediately indicated for her. A heterogeneous mass with solid and predominantly cyst components inside the structure (139 mm × 60 mm × 110 mm), was noticed in the left lateral ventricle with surrounding vasogenic edema, on T2-weighted image (Fig. 1). The tumor triggered midline shift and mass effect. There was no hemorrhage or calcification observed inside the lesion. The mean apparent diffusion coefficient values for the solid component of mass was 0.95 × 10⁻³ mm²/s (Fig. 2). The cerebral blood flow value of solid component of mass and normal parenchyma was 45.3 and 20.3, respectively (Fig. 3). On magnetic resonance imaging spectroscopy, the choline/N-acetyl aspartate ratio of the solid part of mass was 2.17 (Fig. 4). The relative enhancement (%), peak enhancement, peak relative enhancement (%), time to peak (s), wash-in rate (s⁻¹), wash-out rate (s⁻¹), and area under the curve values of the solid mass component and the parenchyma, as measured from the axial T1-perfusion map, were 107.66 vs 2.95, 1724.78 vs 55.46, 115.10 vs 4.45, 138.58 vs 56.69, 137.14 vs 16.39, 4.17 vs 12.87, and 226,365 vs 165, respectively (Fig. 5). The first diagnosis, relied on the clinical and subclinical information, was a choroid plexus carcinoma. The patient adopted gross-total tumor resection. The histopathological evaluation of the excised tumor tissues displayed a gliosarcoma. Two weeks later, the patient was released and prompted to adopt adjuvant chemo- and radiotherapy at a distinct oncological center. This patient passed away 9 months later.

**Discussion**

Gliosarcoma was once thought to be the result of the collision of 2 distinct malignant neoplasms, one of which exhibited glial differentiation and the other a sarcomatous differentiation. Both of these neoplasms were thought to have developed from the proliferation of vascular cells. The present understanding of monoclinality has arisen as a result of recent advances in research, since it has been demonstrated that both components share the same genetic arrangement [1,5,6]. Primary gliosarcoma has been classified as a subtype of glioblastoma and given a highest grade in the 2021 WHO classification of central nervous system malignancies [1].

Although it can also affect the frontal, parietal, and occipital lobes, gliosarcoma is a unique morphological subtype of glioblastoma with a small male preponderance, age spanning from the sixth to seventh decades, and a predisposition for the temporal lobes [2,6,7-9]. Thus, gliosarcoma was very uncommon in children and outstandingly rare inside the ventricle. According to several studies, glioblastoma and gliosarcoma are difficult to identify clinically since they both occur at comparable ages, have brief clinical histories, and have short post-diagnosis survival times [1,8,10,11]. The pathophysiology of gliosarcoma is a subject of great debate. Some writers hypothesized that the hyperplastic blood arteries that are frequently present in high grade gliomas underwent neoplastic change to become the sarcomatous components [4,12]. Early descriptions of hyperplastic arteries and perivascular arrangement...
Fig. 3 – Arterial spin-labeling image of the lesion and normal-appearing parenchyma.

Fig. 4 – Magnetic resonance spectroscopy of the lesion.
of sarcomatous components in gliosarcoma by Feigin et al. provided support for this "collision tumor" idea [4,6,10]. A competing view, which has lately gained support, proposes that both gliosarcoma components have monoclonal origins, with the sarcomatous component deriving from abnormal mesenchymal differentiation of the malignant glioma [5,11–13]. The glial and mesenchymal components might be distinguished thanks to reticulin and Glial fibrillary acidic protein (GFAP). The glial component is reticulin-poor and GFAP-positive, in contrast to the sarcomatous regions, which are reticulin-rich and GFAP-negative [6]. The course of treatment is the same as for glioblastomas and includes surgical resection, depending on the patient’s clinical condition, more chemotherapy or radiation may be necessary [6,7].

**Conclusion**

Giant lateral intraventricular gliosarcoma in a child is extremely rare. It is a histologically biphasic tumor with distinct glial and sarcomatous components. It is linked to a very aggressive clinical course, fast patient deterioration, and unfavorable results. Even while receiving radiotherapy and chemotherapy in combination, the patients’ mean survival rate still falls less than 12 months.

**Author contributions**

HXT and NMD contributed equally to this article as first authorship. Each author performed all steps to complete this article.

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**Informed consent**

Informed consent has been obtained from the family members of the patient included in this study.

**Ethics approval**

This study has been approved by the hospital ethics committee (Ref: 352/NĐ2-CDT).
Data sharing statement

Not applicable

Patient consent

Informed consent for patient information to be published in this article was obtained.

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