

# A rare case of congenital glioblastoma with atypical presentation in an eleven month-old infant: Case report with review of literature

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## ABSTRACT

Glioblastoma multiforme (GBM) is the most common and malignant primary brain tumor in adults. The occurrence of this entity in infants is a rarity and portends a uniform dismal prognosis and survival in spite of all the latest available management options. The authors herein report a case of a GBM in an 11 month-old infant with tumor involving predominantly the left temporal and frontal regions who expired 10 weeks after tumor decompression. Literature and available management options have been reviewed in the context of the presented case.

**Key words:** Congenital, glioblastoma multiforme, infants, tumors

## Introduction

Congenital GBMs are a very rare entity and have seldom been reported. Their location, clinical presentation, and imageology features have distinct characteristics compared with those of adult GBMs. The case presented here is unique, with atypical clinical and unusual radiologic features. This article emphasizes the need for meticulous prenatal screening and calls for a comprehensive consensus management protocol and recognizes congenital GBMs as a typical subset of tumors.

## Case Report

This 11 month-old infant was brought by parents with gradually progressive weakness of right-sided limbs for 1 week duration. On examination, the child was stable and had right hemiparesis with bulging tense anterior fontanalle and no other abnormal findings. Contrast enhanced computed tomography (CT) showed a predominantly cystic lesion with solid component with heterogeneously enhancing wall in left temporal region

causing compression on the midbrain. Magnetic resonance imaging (MRI) revealed an intra-axial lesion measuring approximately  $5 \times 5 \text{ cm}^2$  with a predominantly hypo-intense center and a surrounding rim of iso-intense solid component on T1 weighted imaging and predominantly hyper-intense on T2 weighted imaging, showing peripheral enhancement on contrast administration in the left temporal region [Figure 1]. A left-sided fronto-parieto temporal craniotomy was done and the tumor was excised gross totally [Figure 2]. Per-operatively, the lesion was solid with cystic component containing hemorrhagic fluid, solid component being soft and suckable, mildly vascular with a relatively well-defined plane around it. The child recovered well with improvement in hemiparesis and was discharged. Histopathology of the lesion was compatible with the diagnosis of glioblastoma multiforme (GBM). Immunohistochemistry (IHC) was positive for glial fibrillary acidic protein (GFAP) and vimentin, and negative for synaptophysin [Figure 3]. The child had a persistent right-sided hemiparesis and was discharged with advice and counseling regarding chemo- and radiotherapy options. However, the parents did not consent for further management. The child succumbed 10 weeks after surgery following a episode of seizure.

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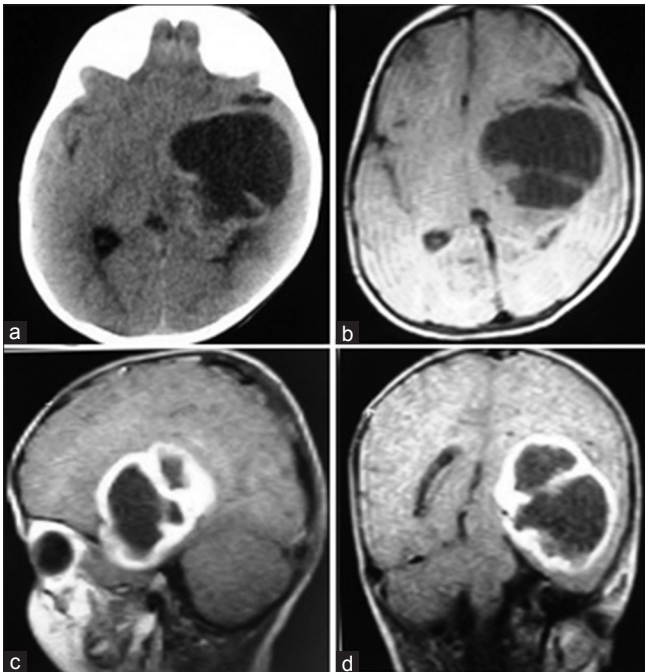
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**Discussion**

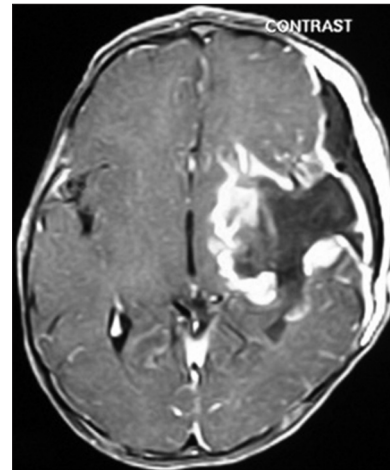
Congenital brain tumors account for 0.5-4% of all pediatric brain tumors.<sup>[1]</sup> Brain tumors are quite uncommon in infants, especially below 6 months of age and when they occur have a dismal course.<sup>[2]</sup> The sites of occurrence, histopathology, and presentation differ considerably in infants when compared with other age groups. Teratomas are the most common congenital tumors with GBM accounting for 2-9%. Solitare and Krigman divided congenital gliomas into three groups: (i) definitely congenital-where the symptoms onset is at birth, (ii) probably congenital-where the presentation is in the first week of life, and (iii) possibly congenital-where the signs or symptoms present within the first few months of life.<sup>[3]</sup> Our case can be categorized under the possibly congenital category as the presentation was during the later half of the year of birth.

Most of the pediatric tumors are located in the infratentorial compartment whereas GBM's in infants have a supratentorial predilection and this has been the same in our case. In the study by Farwell *et al.*, of the 488 children with central nervous system neoplasms, 43 (8.8%) had glioblastomas, of which 22 (51.1%) were in the cerebral hemispheres, 16 (in the brain stem, 2 in the cerebellum, and 3 in the spinal cord.<sup>[4]</sup> Although rare cases are noted intraventricularly too.<sup>[5]</sup> Mahvash *et al.*, observed that in children under 10 years GBM had predilection to infratentorial compartment when compared to those above 10 years who had a supratentorial localization.<sup>[6]</sup>

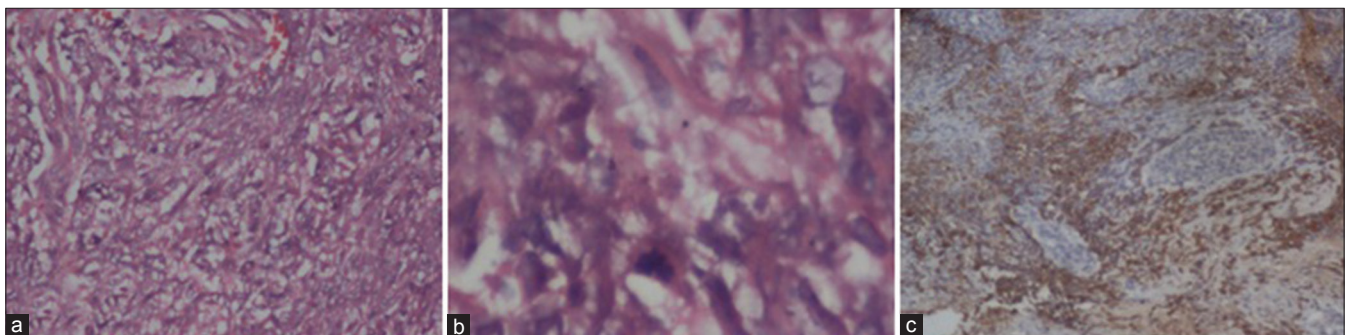
The commonest mode of presentation is an increase in head circumference, accompanied by lethargy and vomiting. The presentation of symptoms is usually delayed in infants due to the fact that the growing infant skull accommodates any encroachment of the cranial cavity due to the presence of open fontanelles which allow cerebral plasticity and thus decompensating any rise in intracranial pressure. Adding to it is the inability of infants to communicate and possible missing of subtle signs by the parents. Contrary to the usual mode of presentation our case presented with right-sided hemiparesis of 1 week duration and lack of any gross features of raised ICP which one would expect in a case of GBM.



**Figure 1:** (a) CT scan brain plain showing predominantly left temporal cystic lesion; (b) T1WI MR axial section showing cystic lesion with minimal mass effect; (c) Post contrast T1WI sagittal section showing the enhancing portion postero superiorly; (d) Post contrast T1WI coronal section



**Figure 2:** Post contrast T1WI – Post operative image showing near total excision



**Figure 3:** (a) H and E (×100) shows sheets of spindle cells with moderate pleomorphism in a fibrillary background; (b) Atypical mitotic figure; (c) IHC with GFAP (×100) shows diffuse positivity in tumor cells

Fetal MRI and prenatal ultrasonography are the latest available diagnostic tools for the detection of congenital GBM.<sup>[7]</sup> These were not done in the present case as there wasn't any antenatal history of exposure to radiation or teratogenic drugs. A conventional plain computed tomography (CT) and plain and contrast enhanced MRI were present. The presence of a predominantly cystic cum solid lesion in a typical temporal location prompted us to think of pilocytic astrocytoma as the first differential. The presence of any perilesional edema which one would expect in a GBM of this size is typically absent here. Per-operatively, the tumor had a relatively well-defined plane around it with not much significant necrosis.

The prognosis of these tumors is very poor with survivals rarely exceeding 2 months in spite of safe maximal resection with adjuvant chemo and radiotherapies. Due to the lack of a clear understanding of the disease pattern and management protocol in infants with congenital GBM's, an optimal management plan and approach with an international consensus is yet to be envisaged. Though high dose chemotherapy post surgery has been advised, the complications pertaining are not clear.<sup>[8]</sup> There is very little evidence in literature on the role of radiotherapy in management of congenital GBM's, partly due to risk of radiation exposure to developing infant brain and partly due to the absence of clear guidelines. This is another gray area where research and studies are yet to be conclusive.

According to Song *et al.*, 27 children with confirmed GBM were retrospectively reviewed for long-term outcomes following surgery and adjuvant therapies.<sup>[9]</sup> The median overall survival (OS) was 43 months and the median progression-free survival was 12 months. The OS rate was 67% at 1 year, 52% at 2 years, and 40% at 5 years, portending a rather better prognosis in children than in adults. In a study by Lang *et al.*, surgical treatment outcomes were assessed in infants younger than 6 months with brain tumors and it was concluded that tumors of different histopathology had a variable but definite impact on the quality of life with residual neurological sequelae post surgery.<sup>[2]</sup>

## Conclusion

In conclusion, though there is a clear distinction in the disease progression, management protocols and survival patterns in adult and pediatric GBM's, there is not much information available on congenital GBM's. Due to lack of significant number of cases and a consensus approach toward management, there is a need for collection of data both prospectively and retrospectively and trials on high dose chemotherapy and radiotherapy regimens with a special emphasis on antenatal detection of these cases.

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## Conflicts of interest

There are no conflicts of interest.

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