

## Gliosarcoma: A rare variant of glioblastoma multiforme in paediatric patient: Case report and review of literature

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**Author contributions:** Meena US involved in majority of work including concept, design, literature search, clinical studies, manuscript preparation, manuscript editing; Sharma S helps in data collection and manuscript preparation; Chopra S helps in manuscript editing; Jain SK helps in manuscript editing and reviewing.

**Institutional review board statement:** Not available.

**Informed consent statement:** Not available.

**Conflict-of-interest statement:** The authors declare no conflicts of interest regarding this manuscript.

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**Manuscript source:** Invited manuscript

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Telephone: +91-0141-2518801

**Received:** January 23, 2016

**Peer-review started:** January 24, 2016

**First decision:** April 18, 2016

**Revised:** May 8, 2016

**Accepted:** July 14, 2016

**Article in press:** July 16, 2016

**Published online:** September 16, 2016

### Abstract

Gliosarcoma is rare central nervous system tumour and a variant of glioblastoma multiforme with bimorphic histological pattern of glial and sarcomatous differentiation. It occurs in elderly between 5<sup>th</sup> and 6<sup>th</sup> decades of life and extremely rare in children. It is highly aggressive tumour and managed like glioblastoma multiforme. A 12-year-old female child presented with complaints of headache and vomiting from 15 d and blurring of vision from 3 d. Magnetic resonance imaging of brain shows heterogeneous mass in right parieto-occipital cortex. A right parieto-occipito-temporal craniotomy with complete excision of mass revealed a primary glioblastoma on histopathological investigation. Treatment consists of maximum surgical excision followed by adjuvant radiotherapy. The etiopathogenesis, treatment modalities and prognosis is discussed. The available literature is also reviewed.

**Key words:** Paediatric gliosarcoma; Surgery; Glioblastoma multiforme; Radiotherapy

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**Core tip:** We report a rare case of primary gliosarcoma right parieto-occipital cortex in a 12-year-old girl with review of available literature. The etiopathogenesis, treatment modalities and prognosis is discussed in view of the literature.

Meena US, Sharma S, Chopra S, Jain SK. Gliosarcoma: A rare variant of glioblastoma multiforme in paediatric patient: Case report and review of literature. *World J Clin Cases* 2016; 4(9): 302-305 Available from: URL: <http://www.wjgnet.com/2307-8960/full/v4/i9/302.htm> DOI: <http://dx.doi.org/10.12998/wjcc.v4.i9.302>

## INTRODUCTION

Gliosarcoma was first time reported by Stroebe<sup>[1]</sup>. It is a rare cerebral tumour consist of bimorphic histological pattern of glial and sarcomatous differentiation. It is considered as grade 4 tumour according to WHO classification and variant of glioblastoma multiforme<sup>[2]</sup>. Incidence of gliosarcoma is 1.8%-2.8% of glioblastoma<sup>[3]</sup>. It usually affects elderly in 5<sup>th</sup> and 6<sup>th</sup> decades of life with slight male predominance and extremely rare in paediatric population<sup>[4]</sup>. So far only few cases of paediatric gliosarcoma have been reported. We report a case of primary gliosarcoma in 12-year-old girl.

## CASE REPORT

A 12-year-old girl was admitted in our institute with history of headache and vomiting from 15 d and blurring of vision from 3 d. Her general and systemic examination was unremarkable. Neurologically Glasgow coma scale was 15/15, pupil were equal and reactive bilaterally. Left side hemiparesis was present with 4/5 power in left upper and lower limb. CECT brain was done which shows large intra axial SOL in right parieto-occipital lobe with perifocal edema, producing mass effect, causing effaced adjacent sulci gyri and ipsilateral lateral ventricle. Lesion shows solid cystic component with intense enhancement after contrast. Midline shift of 8 mm to left side. MRI brain with GD shows similar findings (Figure 1). Antiedema measures were started and early surgery was planned. She underwent right parieto-occipito-temporal craniotomy with complete excision of mass. Mass was greyish brown, avascular and firm in consistency. Post operative period was uneventful. Post operative MRI shows macroscopically complete excision of mass with reduction in mass effect and midline shift (Figure 2). Histopathological examination of the mass shows biphasic pattern comprising of alternating areas of mesenchymal and glial pattern. Mesenchymal component comprise of streaming spindle shape cells with nuclear atypia and many mitotic figures. At one focus pseudopalisading type of necrosis is also seen. In immunohistochemistry spindle shape cells show high MIB labelling index and vimentin positivity and glial cells shows GFAP positivity. Foci of reticulin-rich tumor cells also suggest sarcomatous component (Figure 3).

Patient was discharged on 7<sup>th</sup> post-operative day in good general and neurological condition with advice to attend radiotherapy clinic.

## DISCUSSION

Gliosarcoma is a rare CNS tumour with incidence of 1.8%-2.8% of glioblastomas<sup>[3]</sup>. It usually occurs in elderly between 5<sup>th</sup> and 6<sup>th</sup> decades of life and extremely rare in paediatric population. Okami *et al*<sup>[5]</sup> reported eight cases of paediatric gliosarcoma, Sarkar *et al*<sup>[6]</sup> described three cases and Salvati *et al*<sup>[7]</sup> also reported three cases. Malde *et al*<sup>[8]</sup> reported one case of radiation

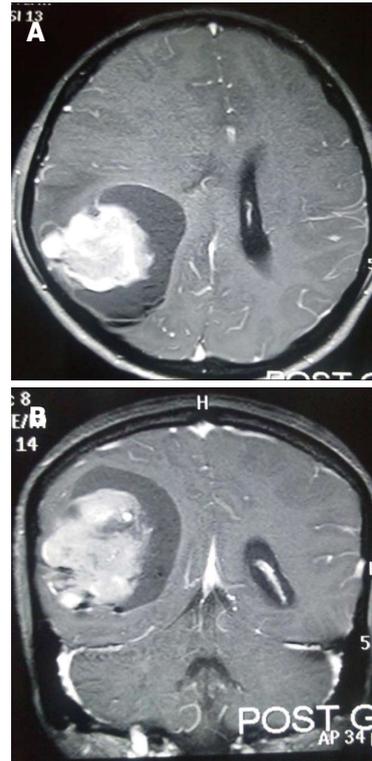


Figure 1 Post contrast magnetic resonance imaging axial (A) and coronal (B) images shows large heterogeneous lesion in right parieto-occipital region with effacement of ipsilateral lateral ventricle.

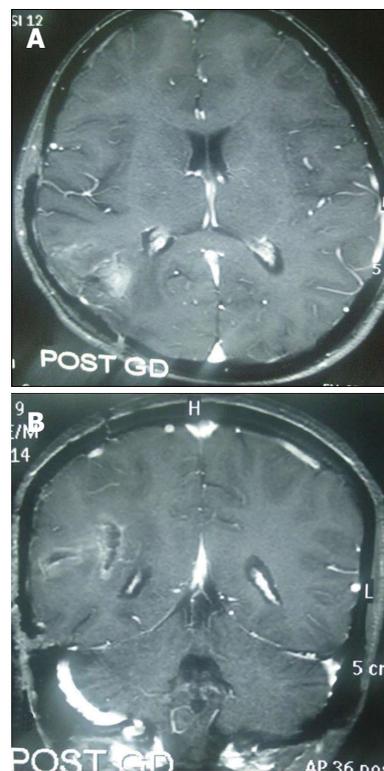
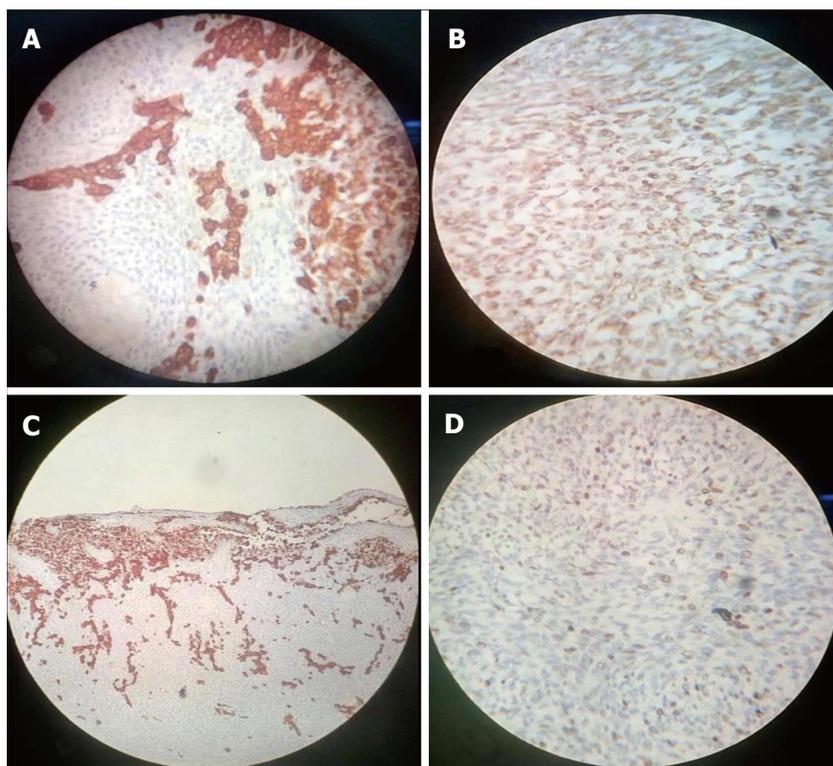


Figure 2 Post-operative contrast magnetic resonance imaging axial (A) and (B) images shows complete excision of tumor.

induced gliosarcoma in literature.

Temporal lobe is most commonly involved followed



**Figure 3** Foci of reticulin-rich tumor cells also suggest sarcomatous component. A: Spindle shape tumor cell shows high MIB labelling index (MIB, × 400); B: Spindle shape tumor cell positive for vimentin (vimentin, × 400); C: Glial cell positive for glial fibrillary acidic protein (GFAP, × 400); D: Glial cell positive for glial fibrillary acidic protein (GFAP, × 100).

by parietal and frontal lobe and can metastasize to extracranial structures like spinal cord, lung, pleura, liver, bone marrow and lymph nodes with increased frequency as compared to glioblastomas<sup>[7]</sup>. Pathogenesis of gliosarcoma is controversial topic and various studies have been proposed. The recent theory suggests monoclonal origin of gliosarcoma. The sarcomatous component originates from aberrant mesenchymal differentiation of the malignant glial cells during tumour progression<sup>[7]</sup>. Meis *et al*<sup>[9]</sup> laid down the criteria's for the diagnosis of gliosarcoma on the basis of histological features: (1) the tumor should be bimorphic, and composed of two morphologically distinct populations of malignant cells (2) one component should be glial in nature with necrotic areas and must fulfil criteria adopted for defining glioblastoma (3) the sarcomatous component should resemble a spindle-cell sarcoma; and must fill one medium power field under (10 × objective with 10 × eyepiece).

The computed tomography findings of gliosarcoma is extremely variable and generally these lesions appear hyperdense mass with heterogeneous thick ring enhancement surrounded by intense peritumoural edema. On magnetic resonance imaging gliosarcoma appear as heterogeneous mass lesion in T1 and T2 weighted sequences with irregular contrast enhancement and intense peritumoural edema. Management of gliosarcoma consist of maximum surgical excision followed by adjuvant radiotherapy. The role chemotherapy in gliosarcoma is still poorly defined although Morantz *et al*<sup>[10]</sup> reported a slight improvement in survival. The average survival in gliosarcoma after diagnosis is less than one year (6 to 14.8 mo) with few case reports of long survivors<sup>[7,11]</sup>.

Prognosis in gliosarcoma depends on histological and radiological features of mass lesion and is slightly better in lesions with predominant sarcomatous component along with meningioma like features in radiology<sup>[7]</sup>.

In conclusion, Gliosarcoma is an aggressive tumor with propensity to recur and re-grow with poor outcome. Although it occurs more commonly in the 5<sup>th</sup> and 6<sup>th</sup> decades of life with temporal lobe predilection, but can occur in any age group or any region. Further studies are needed to understand the biphasic nature of these tumours.

## COMMENTS

### Case characteristics

Patient presented with h/o of headache, vomiting and blurring of vision.

### Clinical diagnosis

She was conscious with left hemiparesis (4/5 power).

### Differential diagnosis

DD Includes malignant meningioma and central nervous system lymphoma and ruled out IHC marker studies.

### Laboratory diagnosis

All routine blood investigations (CBC, RFT, LFT and SE) were normal.

### Imaging diagnosis

Contrast-enhanced computed tomography and magnetic resonance imaging GD Brain show heterogenous mass with intense contrast enhancement surrounded by peritumoural edema.

### Pathological diagnosis

HPE shows biphasic pattern of mesenchymal(spindle cells) and glial cells.

Glial cells show glial fibrillary acidic protein positivity while spindle cells shows vimentin positivity and high MIB index.

### Treatment

Microscopic complete excision of mass done. Patient was advised to attend radiotherapy clinic for adjuvant radiotherapy.

### Related reports

Salvati M, Lenzi J, Brogna C, Frati A, Piccirilli M, Giangaspero F, Raco A. Childhood's gliosarcomas: pathological and therapeutical considerations on three cases and critical review of the literature. *Childs Nerv Syst* 2006; **22**: 1301-1306; Gajendra Singh, Kuntal K. Das, Pradeep Sharma, B. Guruprasad, Sushila Jaiswal, Anant Mehrotra, Arun K. Srivastava, Rabi N. Sahu, Awadhesh K. Jaiswal, and Sanjay Behari: Cerebral gliosarcoma: Analysis of 16 patients and review of literature. *Asian Journal Of Neurosurgery* 2015; **10**: 195-202.

### Experiences and lessons

Although it occurs more commonly in the 5<sup>th</sup> and 6<sup>th</sup> decades of life with temporal lobe predilection, but can occur in any age group or any region.

### Peer-review

It is an interesting topic with only few case reports in literature.

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**P- Reviewer:** Mocellin S, Yamagata M **S- Editor:** Qiu S  
**L- Editor:** A **E- Editor:** Lu YJ





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