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ACUTE HEMORRHAGIC PRESENTATION OF AN EXOPHYTIC PONTINE HIGH-GRADE GLIOMA MIMICKING AN EXTRA-AXIAL LESION IN A CHILD

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ABSTRACT **Background:**Brainstem gliomas constitute approximately 10–15% of pediatric central nervous system tumors [1]. According to the 2021 WHO classification, diffuse intrinsic pontine glioma (DIPG) is now categorized under diffuse midline glioma (H3K27-altered) [2]. Acute hemorrhagic presentation of brainstem Glioma is rare and may radiologically mimic extra-axial lesions, posing diagnostic and therapeutic challenges. **Case Description:** A 10-year-old female presented with acute onset of headache, vomiting ,and altered sensorium (GCS 8/15) with bilateral abducens nerve palsy. Initial CT suggested a hyperdense cerebellopontine angle lesion, raising suspicion of an extra-axial tumor. MRI revealed a hemorrhagic lesion arising from the pons with exophytic extension. The patient underwent retrosigmoid suboccipital craniotomy with evacuation of hematoma and decompression of the exophytic tumor component. Histopathology showed a high-grade glioma (WHO grade 4). Immunohistochemistry demonstrated OLIG2 positivity, retained ATRX, and absence of H3K27M mutation. Postoperatively, the patient improved neurologically and completed radiotherapy. **Conclusion:** This case highlights a rare hemorrhagic presentation of a focal/exophytic pontine high-grade glioma mimicking an extra-axial lesion. It underscores the importance of considering intra-axial pathology despite misleading imaging and supports the role of surgical decompression in life-threatening hemorrhagic presentations.

KEYWORDS : Brainstem Glioma, Pontine Glioma, Hemorrhage, Pediatric, Surgical Decompression, Diffuse Midline Glioma

INTRODUCTION

Brainstem gliomas account for 10–15% of pediatric brain tumors, most commonly involving the pons [1]. Historically termed diffuse intrinsic pontine glioma (DIPG), these tumors are now classified under the 2021 WHO classification as diffuse midline glioma, H3K27-altered, a biologically aggressive entity with poor prognosis [2].

However, brainstem gliomas represent a heterogeneous group, including focal and exophytic high-grade gliomas, which differ significantly from diffuse midline gliomas in clinical behavior, imaging characteristics, and surgical considerations[3].

Acute hemorrhagic presentation in brainstem gliomas is exceedingly rare and may mimic extra-axial lesions radiologically, leading to diagnostic uncertainty and management dilemmas[4,5].

Objective:

To report a rare case of hemorrhagic exophytic pontine high-grade Glioma presenting as an apparent extra-axial lesion and to highlight the role of surgical decompression in such emergency scenarios.

Case Report: A 10-year-old female presented with sudden onset occipital headache, projectile vomiting, and altered sensorium. There was no prior neurological deficit or systemic illness.

Clinical Examination

- GCS: E2V2M4 (8/15)
- General examination: Stable vitals, no systemic abnormality
- Neurological examination:
- Bilateral abducens nerve palsy
- No facial nerve involvement (suggesting non-nuclear involvement)
- No papilledema
- Motor and sensory exam limited due to low GCS

No history of coagulopathy or bleeding disorder was present.

Investigations

- Laboratory parameters: Within normal limits
- CSF analysis: Not contributory (lumbar puncture performed to rule out infection; details limited)

Imaging:

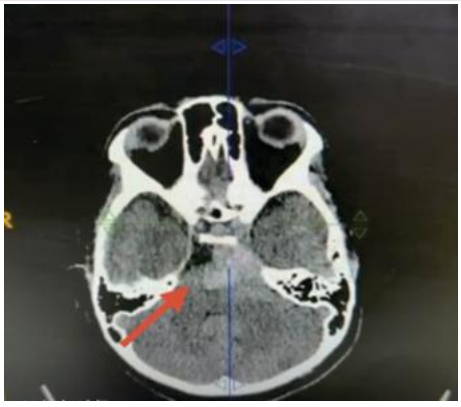

Figure 1.
Plain CT brain (axial) showing hyperdense lesion in left cerebellopontine angle/prepontine region causing mass effect on pons; initial differential diagnosis included extra-axial lesion versus hemorrhage.

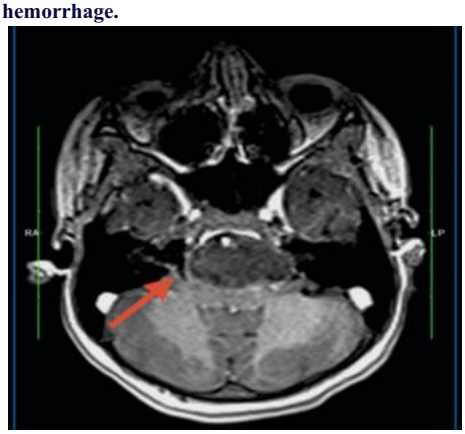

Figure 2.
MRI Brain with contrast showing

- Lobulated lesion centered in the pons with exophytic extension
- Evidence of intralesional hemorrhage (SWI blooming)
- Mild enhancement
- Compression of fourth ventricle without hydrocephalus

Preoperative imaging suggested extra-axial pathology, but intra-operative findings confirmed intra-axial origin [6]

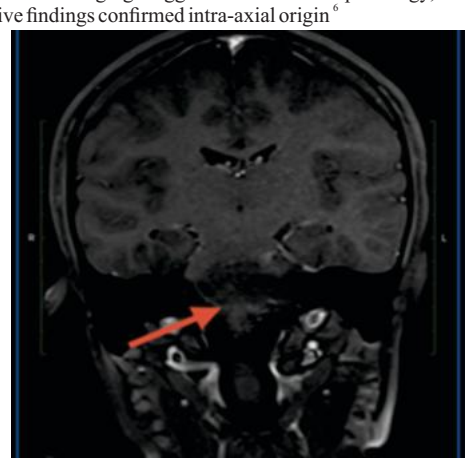

Figure 3.
MRI brain (coronal view) demonstrating lesion centered in pons with exophytic extension.

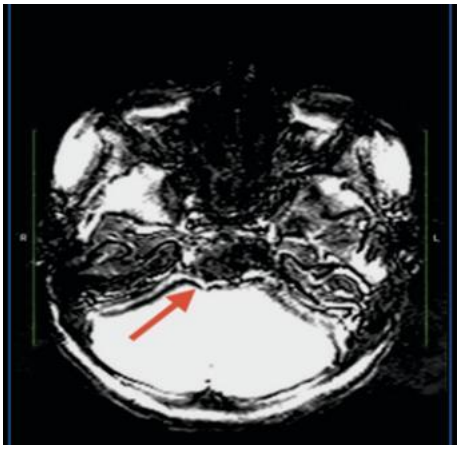

Figure 4.
MRI SWI sequence showing blooming artifact suggestive of intralesional hemorrhage.

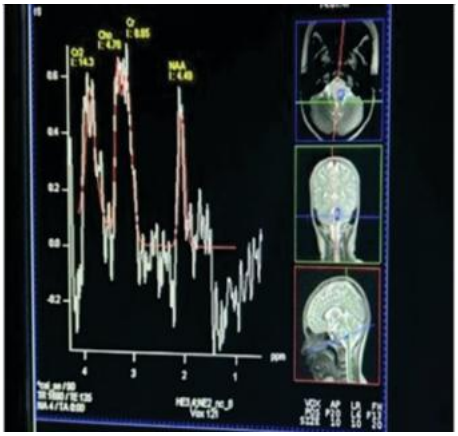

Figure 5.
MR spectroscopy demonstrating reduced Cho/Cr ratio (non-specific finding).

Management: The patient was stabilized with steroids (for cerebral edema) and taken for urgent surgery due to mass effect and neurological deterioration.

Surgical Procedure

- Left retrosigmoid suboccipital craniotomy (park bench position)
- CSF drainage from cisterna magna
- Entry via brainstem safe entry zone (confirming intra-axial nature)

Decompression of exophytic tumor component with hematoma evacuation (NOT complete excision).

The tumour was greyish, soft, moderately vascular lesion associated hematoma and was located between CN V and VII as shown in figure 6.

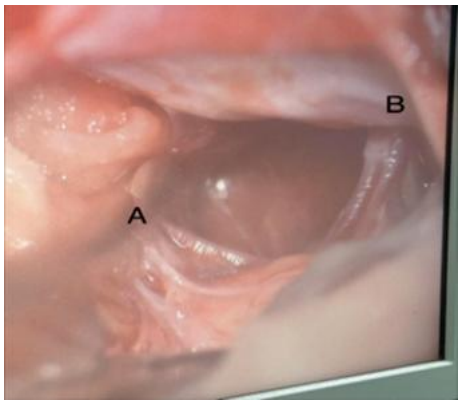

Figure 6: Intraoperative image showing decompressed lesion cavity with identification of cranial nerves V and VII.

HISTOPATHOLOGY: Features of High-grade Glioma (WHO grade 4)

- Nuclear atypia, pleomorphism
- High mitotic activity
- Necrosis

Immunohistochemistry

Table 1. Immunohistochemical Profile of the Tumour

Marker	Result
OLIG2	positive
ATRX	retained
P53	positive
H3K27M	negative
H3K27me3	retained
Ki-67	20%

Limitation: Molecular profiling (NGS/IDH) could not be done.

Postoperative Course:

- Neurological recovery from GCS 8 to 15
- Initial bulbar weakness and paraparesis improved with physiotherapy
- Patient underwent radiotherapy (30 fractions)
- At 4th week: Functional improvement with support

DISCUSSION

Brainstem gliomas are a heterogeneous group of tumors broadly classified into diffuse midline gliomas (H3K27-altered) and focal or exophytic gliomas, which differ significantly in their biological behavior, radiological features, and management strategies [2,3]. The present case does not represent a classical diffuse midline glioma; rather, it is more consistent with a focal/exophytic high-grade glioma, as evidenced by its relatively well-defined margins, exophytic growth pattern, and absence of H3K27M mutation on immunohistochemistry.

Hemorrhagic presentation in brainstem gliomas is uncommon but has been documented in the literature and may result in acute neurological deterioration [4,7]. Such presentations often obscure the underlying tumor and can lead to significant diagnostic confusion. In the present case, initial computed tomography suggested an extra-axial lesion, and subsequent magnetic resonance imaging was also misleading, failing to definitively establish the intra-axial origin. Similar radiological mimicry has been reported previously, particularly in hemorrhagic lesions, where altered imaging characteristics may resemble extra-axial pathologies such as meningioma or nerve sheath tumors [5,8].

Acute hemorrhagic presentation of brainstem gliomas remains exceedingly rare, with only a limited number of cases reported. Aghajan Y et al. described a case of high-grade glioma presenting as acute brainstem hemorrhage, highlighting both the rapid neurological decline and the diagnostic challenges associated with such cases [4]. Most available reports emphasize that hemorrhage can mask the underlying neoplasm on imaging, thereby reducing the reliability of conventional radiological assessment and necessitating histopathological confirmation.

While diffuse midline gliomas are typically not amenable to surgical

resection due to their infiltrative nature, focal or exophytic brainstem gliomas may provide a limited surgical window, particularly in emergency situations [3,9]. Surgical intervention may be considered in selected cases with life-threatening hemorrhage, significant mass effect, or diagnostic uncertainty. Previous surgical series have demonstrated that decompression of the exophytic component can result in meaningful neurological improvement without substantially increasing morbidity when performed judiciously [10]. In the present case, surgical decompression with hematoma evacuation not only relieved mass effect but also provided tissue for definitive diagnosis.

Additionally, MR spectroscopy findings must be interpreted cautiously. A reduced choline-to-creatine (Cho/Cr) ratio is non-specific and may be observed in various pathological conditions. Hematomas are more reliably characterized by the presence of lipid and lactate peaks along with reduced N-acetylaspartate (NAA), rather than isolated metabolite ratios [11].

Limitations

- No molecular profiling
- Single case
- Limited follow-up

CONCLUSION

Preoperative imaging suggested an extra-axial lesion; however, intraoperative findings and histopathology confirmed an intra-axial pontine high-grade Glioma. This case emphasizes on diagnostic pitfalls, the Importance of tissue diagnosis, and role of surgical decompression in selected hemorrhagic brainstem gliomas.

List of Abbreviations

DIPG – Diffuse Intrinsic Pontine Glioma
WHO – World Health Organization
MRI – Magnetic Resonance Imaging
CT – Computed Tomography
SWI – Susceptibility Weighted Imaging
GCS – Glasgow Coma Scale
CSF – Cerebrospinal Fluid
CN – Cranial Nerve
IHC – Immunohistochemistry
ATRX – Alpha Thalassemia/Mental Retardation Syndrome X-linked
Cho/Cr – Choline/Creatinine Ratio
NAA – N-acetylaspartate
NGS – Next Generation Sequencing

Declarations

Ethics Approval and Consent to Participate:
Ethical approval was obtained from the institutional ethics committee of Goa Medical College and Hospital, Goa. Written informed consent for participation and treatment was obtained from the patient's guardians.

Consent for Publication
Written informed consent for publication of clinical details and radiological images was obtained from the patient's parents/legal guardians.

Availability of Data and Material:
All data generated or analyzed during this study are included in this published article. Additional details are available from the corresponding author on reasonable request.

Competing Interests:
The authors declare that they have no competing interests.

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Authors' Contributions

- SD contributed to conceptualization, surgical management, manuscript drafting, final revision, and approval of the final manuscript.
- SF contributed to clinical data collection, literature review, and manuscript editing.
- JK contributed to surgical assistance, radiological interpretation, and manuscript review.
- CY contributed to supervision and critical revision of the manuscript.
- All authors read and approved the final manuscript.

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