Primary Pediatric Spinal Cord Glioblastoma: A Case-Based Insight into Diagnostic and Therapeutic Challenges

Ayari Jihen[1], Achoura Sameh[2], Balti Mehdi[3], Haddaoui Abderrazek[4]

Department of Medical Oncology, Military Hospital of Instruction, Tunisia

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ABSTRACT

Background: Primary intramedullary glioblastoma (scGBM), is exceedingly rare in children, accounting for 1.5% of pediatric spinal cord tumors. These highly aggressive neoplasms are associated with rapid neurological decline and dismal prognosis.

Case presentation: We report a 12-year-old boy with progressive tetraparesis, headaches, and cervical pain. MRI demonstrated a cervico-medullary intramedullary mass extending to C7 with heterogeneous enhancement and hemorrhagic components. Subtotal resection was achieved via suboccipital craniectomy and C1–C5 laminectomy. Histopathology confirmed glioblastoma (WHO grade IV). No adjuvant therapy was administered. The patient developed worsening respiratory dysfunction and succumbed to disease four months after diagnosis.

Conclusion: This case highlights the fulminant course of pediatric scGBM, the critical role of molecular profiling, and the therapeutic value of multimodal approaches including radiotherapy and chemotherapy. Multicenter registries and prospective pediatric trials are urgently needed to improve evidence-based management.

INTRODUCTION

Primary spinal cord glioblastoma (scGBM) in children represents a rare diagnostic and therapeutic challenge, accounting for only ~1.5% of pediatric spinal cord tumors. Prognosis is poor, with median survival typically below 14 months despite aggressive treatment. The introduction of the 2021 WHO classification and increasing emphasis on molecular profiling have reshaped the understanding of these tumors, yet management remains extrapolated from intracranial glioblastoma protocols or limited pediatric series. We present a fulminant pediatric case and discuss diagnostic, therapeutic, and prognostic considerations in the context of recent literature.

Case Presentation:

A 12-year-old boy with no prior medical history presented with two months of headaches, cervical pain, and progressive weakness of all four limbs. On admission, he exhibited tetraparesis and respiratory distress.

Spinal MRI revealed an intramedullary lesion extending from the cervicomedullary junction to C7 . The lesion appeared hypointense on T1, hyperintense on T2, with heterogeneous gadolinium enhancement and hemorrhagic components (figure1,2). Subtotal resection was performed via suboccipital craniectomy and C1–C5 laminectomy.

Histopathological analysis confirmed glioblastoma (WHO grade IV). No adjuvant radiotherapy or chemotherapy was given. Postoperatively, the patient developed progressive respiratory and autonomic dysfunction and died four months after diagnosis.

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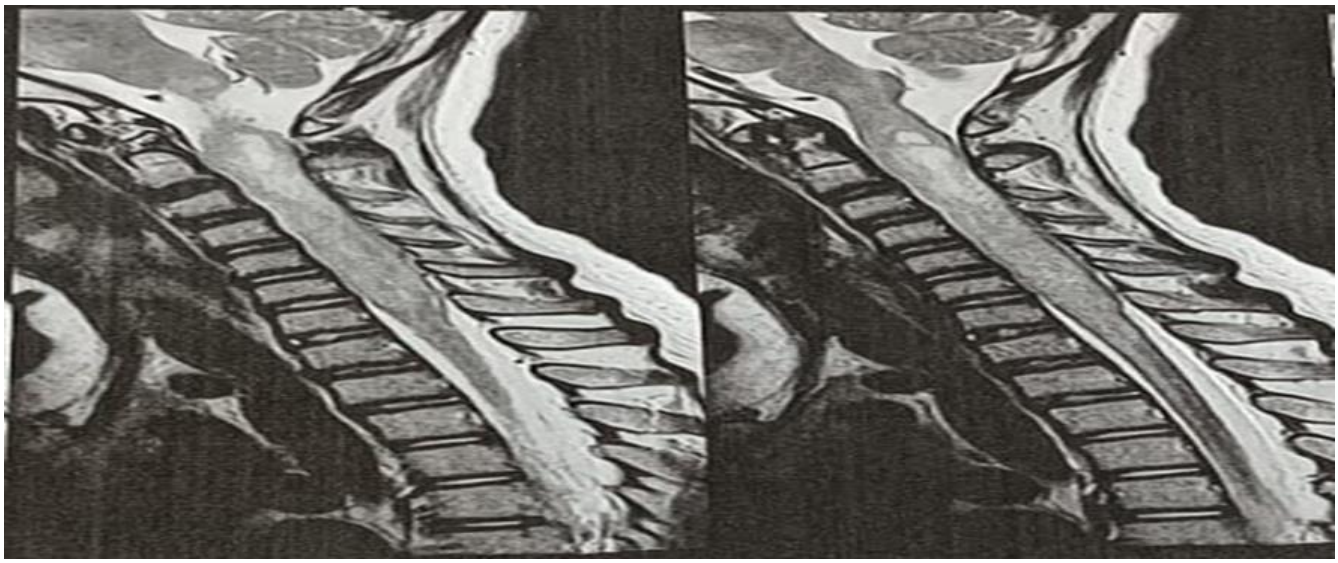

Figure 1: Cervical MRI, sagittal T2-weighted sequence showing an intramedullary hyperintense lesion expanding the spinal cord diameter between C1 and C7.

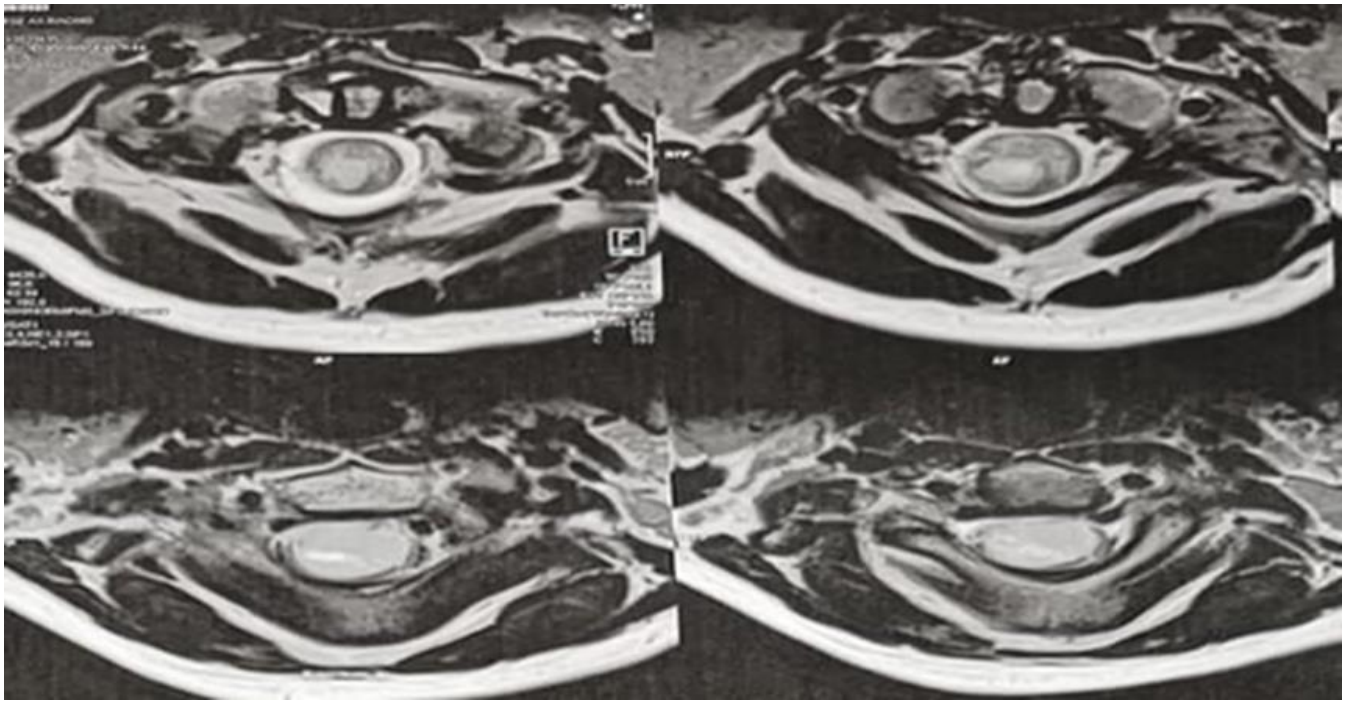

Figure 2: Cervical MRI, axial T2-weighted sequence showing an intramedullary hyperintense lesion predominantly lateralized to the right.

DISCUSSION

Epidemiology and Prognosis:

Primary pediatric spinal cord glioblastoma (scGBM) is exceedingly rare, accounting for <0.1% of pediatric CNS malignancies [1–3]. Contemporary systematic reviews consistently report median overall survival ranging between 9 and 14 months, underscoring the dismal prognosis [1–3]. Tumor location has strong prognostic implications: cervical and cervicomedullary lesions are associated with early respiratory compromise and increased mortality [2,3]. Other prognostic determinants include age, preoperative neurological status, and extent of surgical resection. Younger patients and those undergoing maximal safe resection tend to achieve superior outcomes [3,4]. These findings emphasize the need for early recognition and aggressive, multimodal treatment strategies.

Radiological and Pathological Features:

MRI with gadolinium remains the diagnostic cornerstone, typically revealing T1 hypointensity, T2 hyperintensity, and heterogeneous enhancement with necrotic or hemorrhagic components [5]. Leptomeningeal dissemination is present in up to 40% of cases, mandating neuraxis-wide MRI and, when feasible, cerebrospinal fluid (CSF) cytology [5]. Our case lacked craniospinal staging, representing a limitation.

Histomolecular profiling is increasingly recognized as indispensable. Many pediatric scGBMs harbor H3 K27 alterations, categorizing them as diffuse midline gliomas (DMG) with uniformly poor prognosis [6]. Additional recurrent alterations include TP53 mutations and PDGFRA amplification, while actionable mutations such as BRAF V600E or NTRK fusions—although rare—offer potential for targeted interventions [7,8]. The absence of molecular testing in the present case precluded definitive classification and trial eligibility, limiting therapeutic opportunities.

Therapeutic Strategies:

Maximal safe resection remains the primary therapeutic intervention, aiming to relieve mass effect and preserve function. However, true gross total resection is achieved in a minority of cases due to the infiltrative nature of scGBM and eloquent tumor location. Intraoperative neurophysiological monitoring is recommended to minimize morbidity [9,10].

Adjuvant radiotherapy represents the cornerstone of postoperative management and has been shown to significantly prolong survival compared with surgery alone. Conformal techniques, including intensity-modulated radiotherapy (IMRT) and proton beam therapy, allow delivery of adequate tumoricidal doses within spinal cord tolerance [11]. Craniospinal irradiation is considered in cases with leptomeningeal dissemination [12].

The Stupp regimen—radiotherapy with concomitant temozolomide followed by adjuvant temozolomide—remains the most widely adopted systemic therapy, though pediatric-specific evidence is limited [13]. Recent meta-analyses suggest modest benefit when combined with radiotherapy [1]. Novel therapeutic avenues under investigation include epigenetic modulators, such as panobinostat, and immune checkpoint inhibitors, particularly for H3 K27-altered tumors [7,14].

The most recent meta-analysis with Da Cunha et al[1] demonstrated that patients receiving multimodal therapy (surgery plus radiotherapy ± chemotherapy) had superior survival compared with surgery alone (median PFS 15.2 vs. 2.1 months), with a consistent overall survival advantage across series [1]. By contrast, our patient, treated without adjuvant therapy, experienced rapid decline and represents the poorest-prognosis subgroup, aligning with the literature.

In addition to disease-directed therapies, postoperative supportive management is critical—particularly for cervicomedullary lesions with high risk of respiratory failure. Evidence supports proactive strategies including close respiratory monitoring, early tracheostomy when indicated, intensive physiotherapy, nutritional support, and prevention of infectious complications [5,10]. Such measures may improve short-term survival and quality of life. The rapid respiratory deterioration observed in our case highlights the importance of early multidisciplinary supportive interventions.

Recent systematic reviews and meta-analyses [1–3] reinforce several recurring themes: the extreme rarity and aggressiveness of pediatric scGBM, survival advantage with radiotherapy and combined chemoradiation, strong prognostic weight of cervical location and leptomeningeal dissemination, and the crucial role of histomolecular profiling to refine diagnosis and guide therapy. Our case exemplifies the poor outcomes observed in patients treated with surgery alone, underscoring the pressing need for standardized multimodal protocols.

CONCLUSION

Primary pediatric spinal cord glioblastoma remains a devastating malignancy with poor prognosis. This case highlights the prognostic weight of tumor location and the limitations of surgery alone. Contemporary

management emphasizes histomolecular diagnosis and multimodal therapy, though survival benefits remain modest. Future progress depends on multicenter registries, prospective pediatric trials, and integration of targeted and immunotherapeutic approaches.

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