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Case Report Open Access

Unusual Presentation of a High Grade Spinal Cord Glioma: Case Report

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Abstract

The authors describe a rare case of a high grade spinal cord glioma (HGG) in a young patient in Angola, Africa.The article presents a brief literature review and discusses management options of this rare and controversial disease due to lack of consensus or randomizing trials .

Keywords Spinal cord tumor; Spinal cord glioblastoma; Glioma; Radiotherapy; Spine

Abbreviations AA: Anaplastic Astrocytoma; CSF: Cerebrospinal Fluid; CHT: Chemotherapy; GBM: Glioblastoma Multiforme; GTR: Gross-Total Resection; HGG: High-Grade Glioma; VAS: Visual Analog Scale of Pain; BP: Blood Pressure; HR: Heart Rate; RR: Respiratory Rate; OS: Overall Survival; PR: Partial Resection; RT: Radiotherapy

Introduction

Intramedullary spinal cord tumors are rare entities, accounting for 2% to 10% of all central nervous system tumors and for 15% of primary intradural spinal tumors in adults [1,2]. Approximately 85% of these spinal cord tumors are of astrocytic lineage, and 10 to 15% of these will have histological features consistent with a high-grade glioma (HGG) (that is, anaplastic astrocytoma (AA) or glioblastoma multiforme (GBM) [3]. The incidence of intramedullary spinalcord tumors is less than one-third of 1%, and of these only 1%-3% are high-grade spinal tumors (0.22 per 100,000) [1,4]. Tumors of the spinal cord are much less frequent than intracranial tumors. Overall prevalence is about 4 intracranial lesions for every 1 spinal tumor [5]. The most common gliomas of the spinal cord are astrocytomas and ependymomas, with ependymomas accounting for 60% to 80% of all gliomas. The anatomic distribution of intramedullary spinal cord tumors is proportional to the length of the cord, with thoracic segment having the most (50%-55%), lumbosacral the second most (25%-30%), and the cervical segment the least (15%-25%). The presenting symptoms of intramedullary spinal cord tumors generally arise slowly and progress insidiously and may include general or localized: Pain, paresthesia, weakness, spinal deformity, incontinence, and torticollis [6]. Decision-making regarding the treatment guidelines of high-grade spinal cord gliomas is yet to be established and a great challenge to Medical personal managing patients with this disease. A Systematic review showed a better outcome in patients treated with combined Surgery (gross total resection (GTR) or partial resection (PR) and adjuvant therapy: Chemoteraphy (CMT) with temozolamide and radiotherapy (RT) [7].

Case Report

A 10-years-old male patient without previous personal or family history of illness was presented at our institution with a 2 weeks history of progressive worsening of a shock like neck pain, VAS 7/10, irradiating to both arms. 1-week prior the admission developed a gait abnormality. The admission examination: BP=66/49 mm/Hg, RR=17 c/min, regular pulse, HR=106 bpm. On neurologic examination patient presented alert and conscious, paretic gait; muscular strenght spastic tetraparesis Grade IV-proximal, Grade III distal on all four limbs; Sensibility: hypoesthesia on level T1; Reflexes: presence of bilateral Babinsky and left ankle clonus.

Investigations

Haemogram, Urine examination, renal and liver function tests were normal. CSF 1 analysis showed elevated protein levels and tumor cells. Spine MRI revealed an expansive intramedullary lesion, extending from spine levels C3 to T4 (Figures 1 and 2).

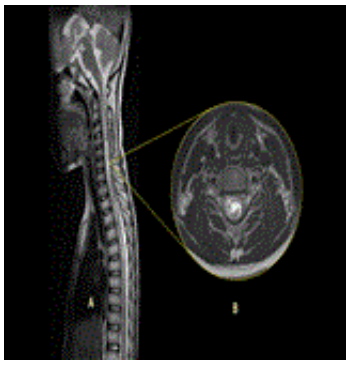

Figure 1: A. Sagital T2 Gd sequence B. Axial T2 Gd sequence: Diffuse cord expansion extending from cervical to thoracic spinal cord

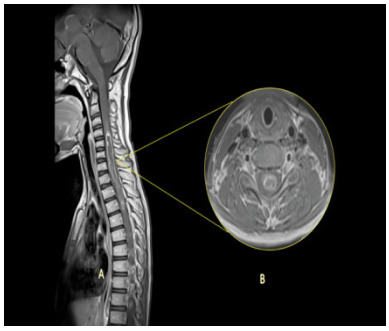

Figure 2: A. Sagital T1 Gd sequence B. Axial T1 Gd sequence : Diffuse insuflative lesion extending from cervical to thoracic spinal cord, with low absorption of contrast and areas of necrosis.

Treatment

The patient was medically managed with analgesics and steroids, submitted to a cervical C3-C4 hemi laminectomy and partial resection of the tumor. Patient didn't present worsening on neurological examination. After 2 weeks of surgery, standard RDT was performed (14 Gy/7 fr) and CMT with Etoposide and cyclophosphamide due to unavailability of Temozolamide in our institution.

Outcome and follow-up

Patient was treated with surgery and an intraoperative biopsy was obtained. After the confirmation of the diagnosis, submitted to a partial resection of the tumor with a close monitoring of the respiratory function and preservation of muscular strength as the main goals (Figure 3 and Table 1).

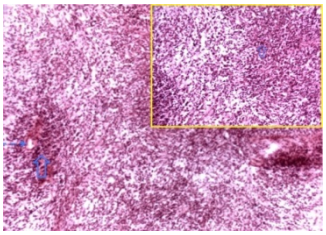

Figure 3: Pleomorphic areas with atypia (big arrow); Neovascular formations (small arrow).

Grade	Characteristics
I	Intact neurologically, normal ambulation, minimal dyesthesia
II	Mild motor or sensory deficit, functional independence
III	Moderate deficit, limitation of function, independent w/external aid
IV	Severe motor or sensory deficit, limited function, dependent
V	Paraplegia or quadriplegia, even w/flickering movement

Table 1: Modified McCormick scale.

The patient maintained the previous deficits post operatively. Pathology revealed a high-grade astrocytoma (WHO Grade IV). Patient was referred to a cancer center abroad to proceed with CMT and RT treatment. Unfortunately patient worsened his clinical performance while awaiting Transfer overseas to a cancer center with more treatment options for radiotherapy and died 5 months after surgery.

Discussion

HGG´s are extremely rare tumors and there is no standard treatment to date. The prognosis for patients with these tumors who undergo surgery and adjuvant radiation therapy and/or chemotherapy is reportedly poor (9 months approximately). According to Adams et al. and Santi et al. in their published works, the median survival of patients with spinal glioblastomas was 10 months [7,8,10]. Primary spinal glioblastomas/sarcomas mainly affect younger patients. Median age at diagnosis reported by (Beyer S et al.) was 22 years [7]. Patients with tumors located in the thoracic spinal cord have a significantly better prognosis than patients with cervical lesions because they tend to progress faster to central respiratory failure and death [9]. The question whether GTR is beneficial for the outcome of patients with spinal malignant gliomas is controversial reported that patients with initial GTR did not show longer survival compared with patients submitted to biopsy and RT [10].

The efficacy of adjuvant radiotherapy for the treatment of HGG´s has yet to be determined [11,12]. The total radiation doses ranges from 18 Gy to 75 Gy with an average value of 49.2 Gy [1,4]. Minehan et al. reported that postoperative radiotherapy significantly enhanced overall survival (OS) in patients with high-grade spinal cord gliomas, at the cost of lower limb and bladder function sacrifice [1,13].

Katoh et al. reported that the 5-year survival rate for patients with high-grade gliomas who received hypo-fractionated radiotherapy was 67%, and it was 35% for those who received conventional radiation therapy [1,14]. Patients who underwent surgery and radiation cordotomy survived longer than those who underwent surgery and conventional radiation therapy alone [14,15].

Conclusion

HGG are rare tumors, associated with a poor prognosis despite the evolution of treatment options achieved over the last decades. The characteristics and treatments of primary spinal glioblastomas are very variable and sometimes define the rate of progression of the disease. Irregular availability of CMT agents has an undesired impact on outcome of patients. Tumor subtype and histological grade are the most important prognostic factors in HGG. Prospective multicenter studies are needed to provide conclusive evidence regarding the extent of resection on patients outcome and more effective strategies to treat this disease.

Disclosures

The authors declared no potential conflicts of interest.

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