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NAVIGATING RECURRENCE IN INTRAMEDULLARY SPINAL EPENDYMOMA: THE CRITICAL ROLE OF RADIOTHERAPY

Radiotherapy

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ABSTRACT

Spinal ependymomas are rare, slow-growing tumors accounting for approximately 1.6–1.8% of all primary central nervous system tumors. Gross total resection is considered the standard treatment; however, the role of adjuvant radiotherapy following complete resection of World Health Organization (WHO) Grade II tumors remains controversial. We report a case of a 56-year-old male with a dorso-lumbar intramedullary spinal ependymoma who underwent gross total resection and subsequently developed local recurrence 10 months after surgery. As re-excision was not feasible, the patient received focal external beam radiotherapy using the Volumetric Modulated Arc Therapy technique to a total dose of 54 Gray in 30 fractions over six weeks. Treatment was well tolerated, with complete resolution of symptoms and no evidence of disease on follow-up imaging at six months. This case highlights the role of radiotherapy as an effective salvage modality and emphasizes individualized decision-making in the management of Grade II spinal ependymomas.

KEYWORDS

Ependymoma; Spinal Cord Neoplasms; Radiotherapy, Adjuvant; Radiotherapy, Conformal

INTRODUCTION

Ependymomas arise from ependymal cells lining the ventricular system and the central canal of the spinal cord. Spinal ependymomas are uncommon tumors accounting for approximately 1.6–1.8% of all primary central nervous system (CNS) neoplasms. [1–3] They are most frequently classified as World Health Organization (WHO) Grade II lesions.[4] Surgical resection remains the primary treatment modality, with gross total resection associated with favorable outcomes. However, the role of adjuvant radiotherapy following complete resection remains uncertain because of limited prospective data. We report a case of recurrent spinal ependymoma successfully managed with focal radiotherapy.

Case Report

A 56-year-old man presented with lower back pain radiating to the right leg for 6 months and progressive weakness and tingling sensation in the right leg for four months. Neurological examination revealed normal motor strength in both upper limbs (power 5/5 bilaterally). In the lower limbs, motor power was reduced on the right side with hip flexion and extension graded 2/5, knee flexion and extension 3/5, and ankle dorsiflexion and plantarflexion 3/5. The left lower limb demonstrated normal strength. Deep tendon reflexes were preserved bilaterally. Magnetic Resonance Imaging (MRI) of the spine demonstrated a heterogeneous intramedullary mass extending from the twelfth dorsal to the third lumbar vertebral level, measuring approximately 11 × 2 × 2 cm.

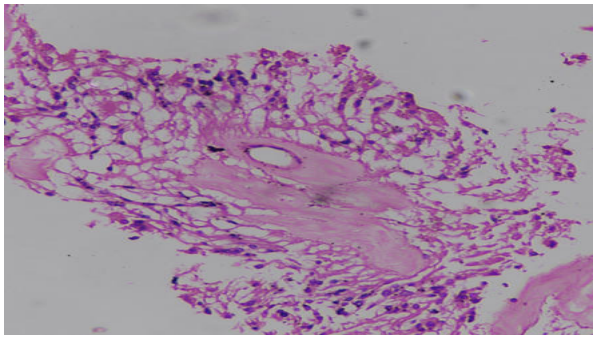

Figure 1: Perivascular pseudorosettes in ependymoma (Hematoxylin & Eosin stain). Photomicrograph (10X) showing characteristic perivascular pseudorosettes composed of tumor cells radially arranged around a central blood vessel. The neoplastic cells display oval to spindle tumour cells with irregular nuclear chromatin with fibrillar processes. These perivascular pseudorosettes are a hallmark histopathological feature of ependymoma.

The patient underwent spinal laminectomy with gross total excision of the lesion. Postoperative recovery was uneventful, and neurological examination was normal. Histopathological examination, with immunohistochemistry for glial fibrillary acidic protein (GFAP) positivity and epithelial membrane antigen (EMA) negativity, confirmed WHO Grade II cellular ependymoma (Figures 1 and 2). Adjuvant radiotherapy was advised; however, the patient declined further treatment and was kept on regular follow-up.

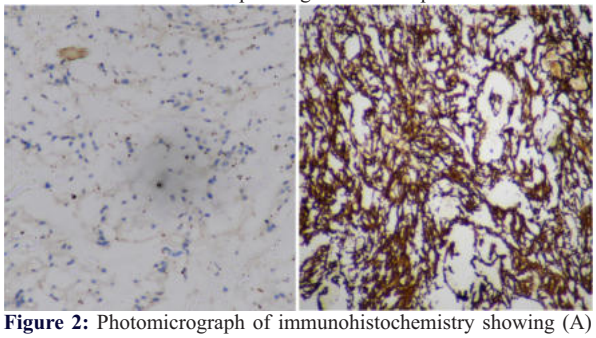

Figure 2: Photomicrograph of immunohistochemistry showing (A) GFAP positive and (B) EMA negative tumour cells

Ten months after surgery, follow-up MRI revealed a well-defined lesion inferior to the conus medullaris at the level of L1–L2, suggestive of local recurrence. Neurosurgical evaluation deemed re-excision unsuitable, and the patient was planned for focal radiotherapy.

Computed tomography (CT) simulation and treatment planning were performed. External beam radiotherapy was delivered using the Volumetric Modulated Arc Therapy (VMAT) technique. Treatment planning was performed using the MONACO® treatment planning system (version 6.1.2.0; Elekta AB, Stockholm, Sweden). Planning

Target Volume (PTV) coverage achieved 95% prescribed dose to 99.92% of the target volume while maintaining acceptable spinal cord dose constraints (Figure 3).

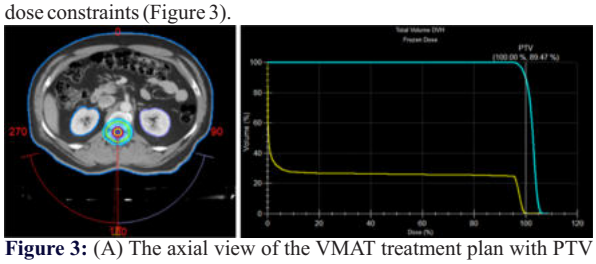

Figure 3: (A) The axial view of the VMAT treatment plan with PTV coverage of 95% dose to 99.92% of the PTV volume and (B) Dose-Volume Histogram showing dose distribution to the spinal cord (yellow) and PTV (blue); despite lying within the area of interest, only 0.076 cc (0.23%) volume of the spinal cord is receiving 100% of the dose as compared to 89.47% volume of the PTV.

A total dose of 54 Gray (Gy) in 30 fractions (1.8 Gy per fraction) was delivered over six weeks using 6 megavolt (MV) photon beams using a high-energy linear accelerator. Daily cone beam CT was used for image guidance. Treatment was well tolerated with no severe toxicities, and the patient experienced complete resolution of symptoms after radiotherapy. Follow-up MRI scans at three months and six months after treatment showed no evidence of recurrent or residual disease (Figure 4). The patient is doing well and remains on a six-monthly follow-up.

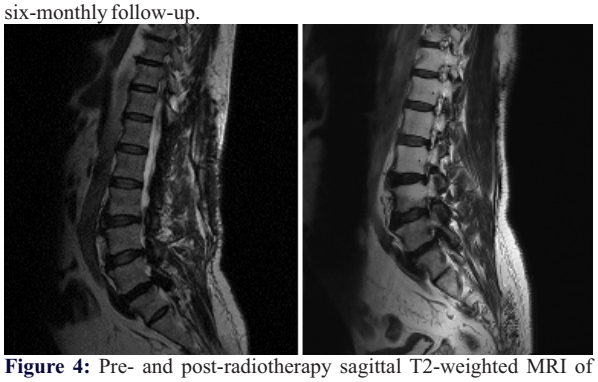

Figure 4: Pre- and post-radiotherapy sagittal T2-weighted MRI of spinal ependymoma. (A) Pre-radiotherapy sagittal T2-weighted MRI demonstrating a well-defined altered signal intensity lesion measuring approximately 40 × 9 × 7 mm, located inferior to the conus medullaris at the L1–L2 vertebral level. The lesion shows central T2 hyporintensity with a peripheral hyperintense rim, consistent with recurrent disease in a known case of spinal ependymoma. (B) Post-radiotherapy sagittal T2-weighted MRI showing no evidence of residual or recurrent lesion at the previously involved site. Only post-radiotherapy changes are noted, with resolution of the prior mass effect and no abnormal focal signal intensity suggestive of active disease.

DISCUSSION

Spinal ependymomas are slow-growing intramedullary tumors that commonly present with back pain, sensory disturbances, and motor weakness.[5] MRI remains the imaging modality of choice for diagnosis and follow-up.[6] Surgical resection aiming at gross total removal is considered the standard initial management and is associated with improved local control.[7]

Adjuvant radiotherapy is generally recommended following subtotal resection because of higher recurrence rates. Recurrence rates following subtotal resection have been reported to be as high as 70%, supporting the use of adjuvant radiotherapy to improve local control and progression-free survival.[8] However, the role of radiotherapy following gross total resection of WHO Grade II tumors remains debated. Despite complete resection in the present case, local recurrence occurred within 10 months after surgery.

Radiotherapy doses in the range of 50–54 Gy using fractionated external beam techniques have been reported to provide effective local control in spinal ependymomas.[9] Modern conformal techniques such as VMAT allow adequate target coverage while minimizing radiation exposure to the spinal cord and surrounding normal tissues.

This case demonstrates that focal radiotherapy can serve as an effective salvage treatment in recurrent disease and supports consideration of adjuvant radiotherapy in selected patients with Grade II spinal ependymomas. Larger studies are required to establish definitive treatment recommendations.

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Conflict Of Interest Statement
The authors declare that there are no conflicts of interest related to this manuscript.

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