Case Report

Primary intradural/extradural Ewing’s sarcoma of the sacral spine: A case report and literature review

Daniel B Murray¹, Jack Horan¹, Alan Beausang², Mohammed Ben Husien¹

¹National Centre for Neurosurgery, Beaumont Hospital/RCSI, Dublin, Ireland, ²Department of Neuropathology, Beaumont Hospital, Dublin, Ireland.

Case Report

A 45-year-old male presented with a 3-month history of increasing lower back pain, which had worsened acutely in conjunction with urinary retention. The magnetic resonance imaging revealed a mass extending from L5 to S2 with additional extension through the left S2-3 neural foramen. The metastatic workup was negative. At surgery, the lesion was both intradural and extradural. Following complete surgical resection, the patient was later treated with radiation and chemotherapy.

Introduction

Ewing’s sarcoma (ES) is a rare and highly malignant mesenchymal tumor, representing less than 10% of all primary bone sarcomas. It typically presents in the second decade of life.¹,²,⁷ Primary intradural/extradural ESs are even less frequently encountered as evidenced by the fact that fewer than 50 such cases are to be found in the literature – of those published cases, over 60% of tumors were located in the lumbar or sacral spine.⁷-⁹ Poor prognostic factors for overall survival include metastatic disease and the site of the primary tumor, with those involving the axial skeleton faring worse.¹,⁸ Notably, gross total resection of these lesions offers improved survival.³,⁵ Multiagent chemotherapy is then added for a treatment course of at least 10 months to further improve survival rates.³,⁵

Here, we present the case of a 45-year-old male who presented with an acute exacerbation of back pain and urinary retention, attributed to an intradural/extradural lumbosacral primary ES, which was treated with gross total surgical resection followed by adjuvant radiation and chemotherapy.

Case Report

A 45-year-old male presented with a 3-month history of increasing lower back pain, which had worsened acutely with associated urinary retention over the preceding 48 h.
examination revealed paraspinal tenderness only, with no neurological deficits. Magnetic resonance imaging (MRI) showed a large $2 \times 2 \times 6$ cm lobulated mass resulting in cauda equina compression from L5–S2, which extended through the left S2–3 neural foramen [Figures 1-3]. The mass was solid and enhancing, with hemorrhagic and cystic components. The metastatic workup was negative.

**Surgery**

Surgery consisted of wide bilateral laminectomies at L5–S2. The extradural component was pushing the thecal sac to the right side; it was excised *en bloc*. The dura was then incised and the intradural component of the tumor was also excised *en bloc* [Figure 4]. Postoperatively, the patient’s pain was markedly reduced, and there were no residual neurological deficits. The postoperative MRI revealed complete resection of the tumor [Figures 5 and 6]. The patient was referred for adjuvant chemotherapy and local radiation.

**Pathology**

The histologic examination demonstrated a rather well-circumscribed, densely cellular neoplasm, surrounded by...
fibroconnective tissue, adipose tissue, and attenuated fascicles of peripheral nerve and ganglion cells [Figure 7]. Mitoses were readily identified. The intralesional cells were small with round to oval nuclei, contained small inconspicuous nucleoli and scant cytoplasm, and were arranged mostly in patternless sheets without well-formed rosettes [Figure 8].

**Immunohistochemistry**

The cells within the tumor showed diffuse membranous immunopositivity with CD99 immunohistochemical stain [Figure 9], in addition to diffuse immunopositivity with neurofilament. Focal staining was observed with synaptophysin. Interphase FISH (using break-apart probes) detected an Ewing sarcoma breakpoint region 1 (EWSR1) gene rearrangement involving locus 22q11, confirming the diagnosis of ES, FNCLCC Grade 3.

**DISCUSSION**

Bone and soft tissue – in particular pelvis, femur, ribs, and spine – are the main sites of tumor development in ES, but less common extraosseous sites such as lung, neck and intradural extramedullary spinal ES have been reported in the literature.\(^2\,^7\,^8\) Immunoreactivity for CD99 and detection of the EWSR1 gene rearrangement are fundamental for the correct diagnosis of ES, especially for tumors found in less common locations. Spinal sarcomas are typically distinguished as sacral or nonsacral, as tumor behavior and response to therapy vary according to this division.\(^10\) Primary ES of the sacrum is rare and tumors with both intradural and extradural components even more so. Treatment of such lesions should consist of surgical excision followed by radiation and/or adjuvant chemotherapy.
CONCLUSION
Primary intradural/extradural ES of the sacrum is exceedingly rare and is best managed with gross total excision followed by adjuvant chemotherapy and radiation.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest
There are no conflicts of interest.

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