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Nancy E. Epstein, MD
Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook

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Case Report

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

Daniel B Murray[1], Jack Horan[1], Alan Beausang[2], Mohammed Ben Husien[1]

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

E-mail: *Daniel B Murray - danielmurray@rcsi.ie; Jack Horan - jackhoran@rcsi.ie; Alan Beausang - alanbeausang@beaumont.ie; Mohammed Ben Husien - mohammedbenhusien@beaumont.ie

***Corresponding author:**
Daniel B Murray,
Department of Neurosurgery,
Beaumont Hospital/RCSI,
Dublin, Ireland.

danielmurray@rcsi.ie

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ABSTRACT

Background: Ewing's sarcoma (ES) is a malignant mesenchymal tumor, most often found in the long bones, and usually affecting children and adolescents in the second decade of life. ES of the spine is a clinical rarity.

Case Description: A 45-year-old male presented with a 3-month history of lower back pain which acutely worsened 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.

Conclusion: Here, we report an adult male who acutely presented with low back pain attributable to primary intradural/extradural sacral ES.

Keywords: Ewing's sarcoma, Extradural, Intradural, Primary neuroectodermal tumor, Sacrum

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.[2,4,7] Primary intradural/extramedullary 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.[1,6] 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. The physical

examination revealed paraspinal tenderness only, with no neurological deficits. Magnetic resonance imaging (MRI) showed a large 2 × 2 × 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.

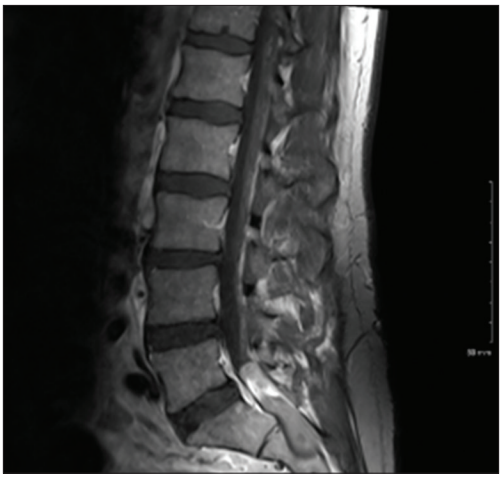

Figure 1: Preoperative T1-weighted sagittal view magnetic resonance imaging of the spine.

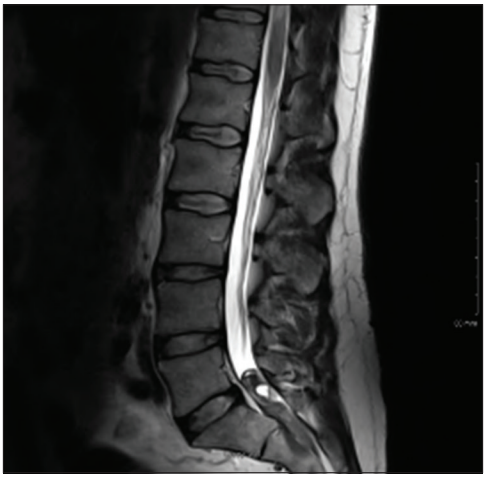

Figure 2: Preoperative T2-weighted sagittal view magnetic resonance imaging of the spine.

Pathology

The histologic examination demonstrated a rather well-circumscribed, densely cellular neoplasm, surrounded by

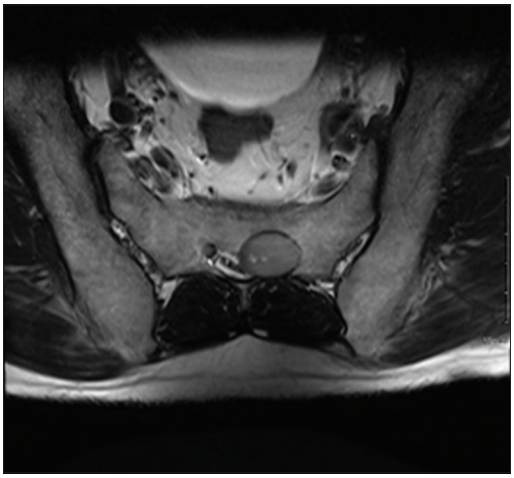

Figure 3: Preoperative T2-weighted axial view magnetic resonance imaging of the spine.

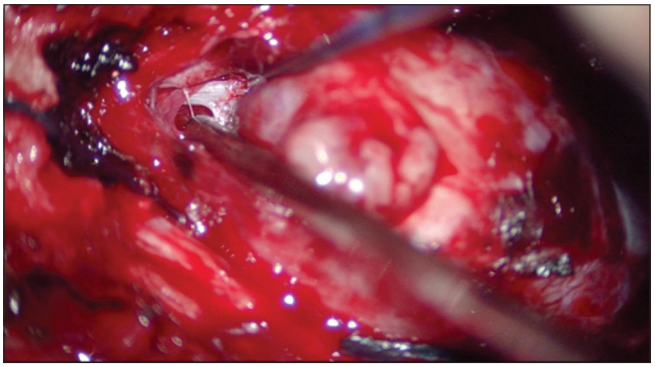

Figure 4: Intraoperative view showing the intradural component of the tumor.

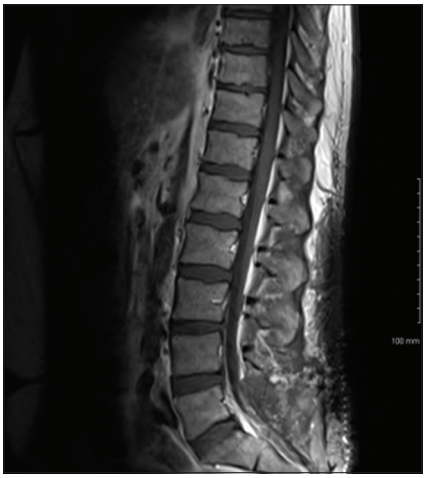

Figure 5: Postoperative T1-weighted sagittal view magnetic resonance imaging showing the resection cavity.

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.

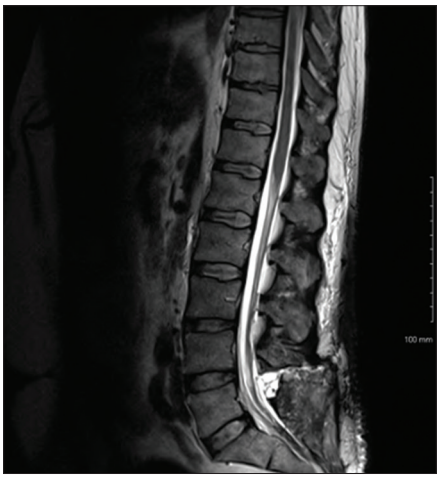

Figure 6: Postoperative T2-weighted sagittal view magnetic resonance imaging showing the resection cavity.

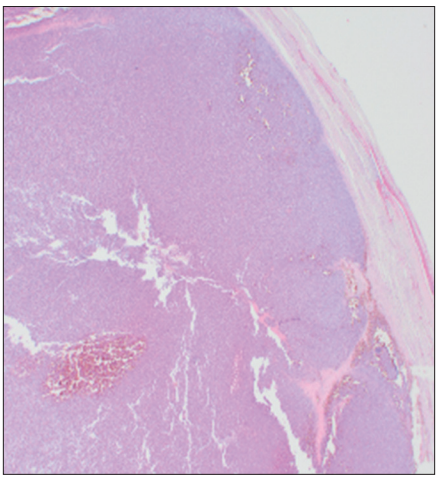

Figure 7: 2×, H and E stain.

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.

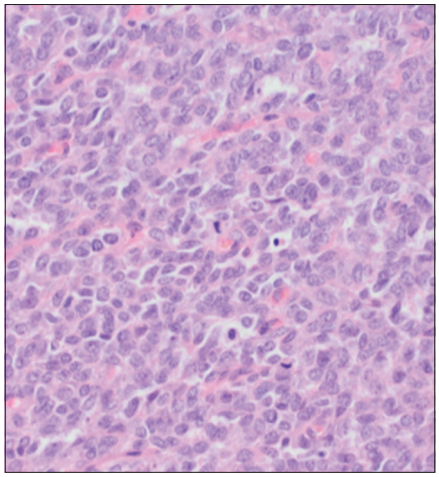

Figure 8: 40×, H and E stain.

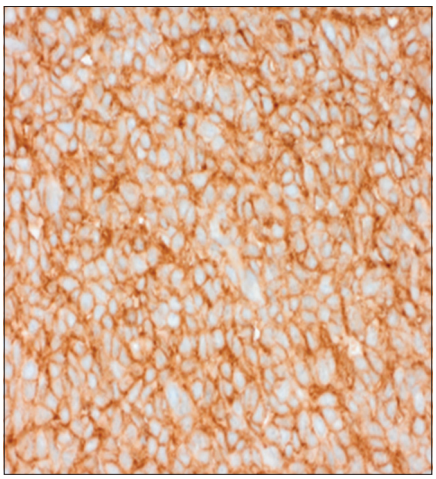

Figure 9: 40×, CD99 immunohistochemical stain.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Cash T, McIlvaine E, Krailo MD, Lessnick SL, Lawlor ER, Laack N, *et al*. Comparison of clinical features and outcomes in patients with extraskeletal versus skeletal localized Ewing sarcoma: A report from the Children's oncology group. Pediatr Blood Cancer 2016;63:1771-9.
2. Choi EY, Gardner JM, Lucas DR, McHugh JB, Patel RM. Ewing sarcoma. Semin Diagn Pathol 2014;31:39-47.
3. Dangoor A, Seddon B, Gerrand C, Grimer R, Whelan J, Judson I. UK guidelines for the management of soft tissue sarcomas. Clin Sarcoma Res 2016;6:20.
4. Esiashvili N, Goodman M, Marcus RB Jr. Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance epidemiology and end results data. J Pediatr Hematol Oncol 2008;30:425-30.
5. Gerrand C, Athanasou N, Brennan B, Grimer R, Judson I, Morland B, *et al*. UK guidelines for the management of bone sarcomas. Clin Sarcoma Res 2016;6:7.
6. Lee J, Hoang BH, Ziogas A, Zell JA. Analysis of prognostic factors in Ewing sarcoma using a population-based cancer registry. Cancer 2010;116:1964-73.
7. Lu VM, Goyal A, Alvi MA, Kerezoudis P, Haddock MG, Bydon M. Primary intradural Ewing's sarcoma of the spine: A systematic review of the literature. Clin Neurol Neurosurg 2019;177:12-19.
8. Paterakis K, Brotis A, Tasiou A, Kotoula V, Kapsalaki E, Vlychou M. Intradural extramedullary Ewing's sarcoma: A case report and review of the literature. Neurol Neurochir Pol 2017;51:106-10.
9. Paterakis KN, Brotis A, Dardiotis E, Giannis T, Tzerefos C, Fountas KN. Multimodality treatment of intradural extramedullary Ewing's sarcomas. A systematic review. Clin Neurol Neurosurg 2018;164:169-81.
10. Pilepich MV, Vietti TJ, Nesbit ME, Tefft M, Kissane J, Burgert O, *et al*. Ewing's sarcoma of the vertebral column. Int J Radiat Oncol Biol Phys 1981;7:27-31.

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