



Case Report

Extraosseous extradural ewing sarcoma of the thoracic spine: Case report and literature review

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Received : 08 August 2021

Accepted : 02 October 2021

Published : 25 October 2021

DOI

10.25259/SNI_790_2021

Quick Response Code:



ABSTRACT

Background: As part of the 'small round-cell tumor' family, Ewing's sarcoma (ES) is a malignant mesenchymal tumor classified as a primitive neuroectodermal tumor (PNET). Within the PNETs, spinal extraosseous extradural lesions are exceedingly rare.

Case Description: A 19-year-old male presented with a one-month history of back pain and paraparesis with loss of urinary control. The MRI revealed an intradural, extramedullary mass at the T3-T4 level. Intraoperatively, the tumor was solely extradural, without evidence of local invasion; it was entirely removed. Due to the high risk of metastasis, the patient was subsequently treated with chemotherapy and radiation. In addition, similar studies on PNETs were reviewed.

Conclusion: A 19-year-old male presented with a paraparesis attributed to an extraosseous extradural ES at the T3-T4 level. Following total gross resection, he was successfully managed with chemotherapy and radiation. The patient has been in remission for one month.

Keywords: Ewing's sarcoma, Extradural, Extraskelatal, Primitive neuroectodermal tumor, Spine

INTRODUCTION

Primary spinal extradural Ewing's sarcoma (ES), also known as primitive neuroectodermal tumors (PNETs), are rare. According to the World Health Organization classification, PNET is an undifferentiated round cell tumor that arises from primitive neuroepithelial cells. They constitute a heterogeneous group of malignant tumors of childhood and early adulthood. Spinal Epidural PNET/peripheral PNET have an incidence of <1% of primary spinal tumors.^[4]

Here, we evaluated a 19-year-old male with an extradural extraosseous T3-T4 ES managed with total gross excision, radiation, and chemotherapy, showing no evidence of recurrence a month later.

CASE PRESENTATION

Clinical presentation

A 19-year-old male presented with a progressive T5-level paraparesis, a T5 sensory level, and sphincter dysfunction of one month's duration [Table 1].

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Radiological evaluation

The thoracic MRI showed an intradural extramedullary lesion at the T3-T4 level; it was well-defined, predominantly right-sided with significant cord compression and lobulated, measuring 3.6 cm × 1.7 cm × 1.5 cm with “tentacles” extending superiorly. On MRI, it was isointense on T1, and heterogeneously hyperintense on T2 weighted sequences, showing partial peripheral enhancement with Gadolinium-DTPA. There was also obliteration of CSF at the level of T3/T4 maximal compression [Figures 1 and 2].

Table 1: Summary of current case.

Parameter	Current Case
Age	19 years
Gender	Male
Presenting Complaint	Back pain, paraparesis, sphincter dysfunction
Location of Tumor	Intradural, extra medullary lesion
Level of Tumor	T3-T4
Resection	Complete
Adjuvant treatment	CT + RT
CD99	Positive
t(11:22)	Positive
Outcome	NED (1 month)

NED: No evidence of disease

Surgery

The patient underwent a T3-T4 laminectomy. The lesion proved to be entirely extradural in location and was soft and vascular; the lesion was fully excised. However, there was profuse intraoperative bleeding, requiring 1 unit of RBC transfusion.

Pathology and molecular cytogenetics confirmation of the diagnosis of epidural EWS

The histological examination revealed a malignant neoplasm consistent with Epidural EWS; cells were arranged in groups, sheets, and nests. Individual tumor cells were round/oval, had uniform round nuclei and clear cytoplasm; a few cells showed a high nucleus to cytoplasm (N/C) ratio and prominent nucleoli. Atypical mitotic figures and areas of necrosis were also seen. [Figure 3].

Molecular cytogenetics further confirmed the diagnosis, along with a fluorescence *in situ* hybridization analysis to assess the EWS gene locus.

Adjuvant therapy

Following confirmation, the patient was treated with a multi-agent chemotherapy regimen as well as radiation therapy.

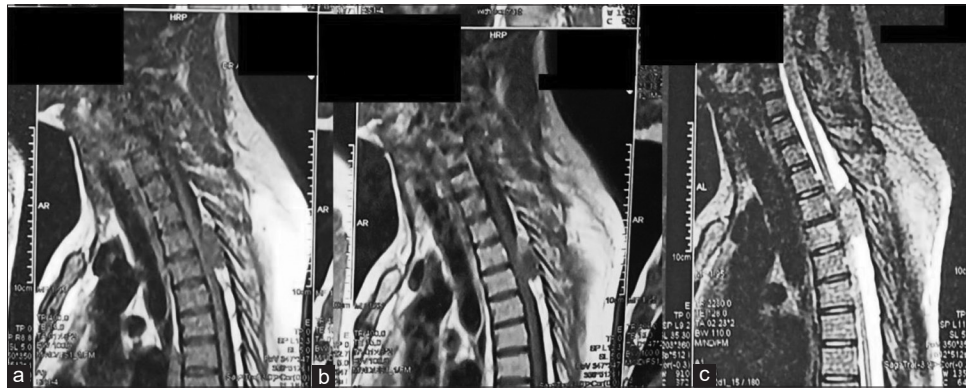


Figure 1: Preoperative sagittal view of current case showing (a) T1 (without contrast), (b) T1 (with contrast), (c) T2 (without contrast)

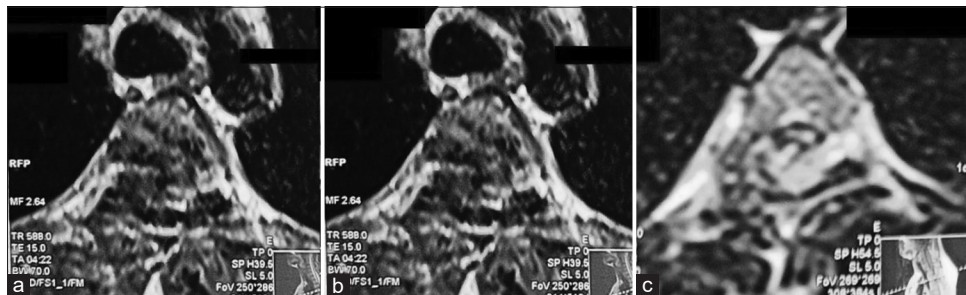


Figure 2: Preoperative axial view of current case showing (a) T1 (without contrast), (b) T1 (with contrast), (c) T2 (without contrast).

Review of literature

Forty-three studies have previously reported a total of 59 cases. The findings of these studies, including our current case, are summarised in [Table 2].

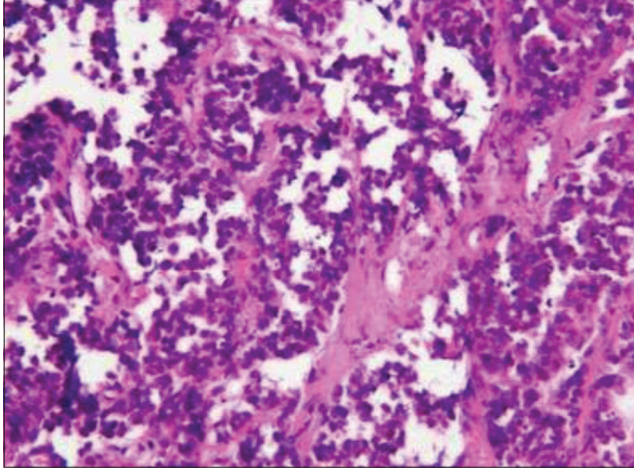


Figure 3: Immunohistochemically, the cells were positive for CD99. The complete reactivity pattern of the specimen is shown in Table 2.

Table 2: Summary of literature review.	
Feature	Frequency
Gender	M (n=35) F (n=25)
Level	Cervical (n=11) Cervicothoracic (n=6) Thoracic (n=15) Thoracolumbar (n=3) Lumbar (n=15) Lumbosacral (n=6) Sacral (n=2)
Resection	Partial (n=31) Complete (n=28) N/A (n=1)
Adjuvant treatment	RT only (n=2) CT only (n=7) RT + CT (n=47) None (n=2) N/A (n=2)
CD99	+ (n=22) N/A (n=38)
t(11;22)	+ (n=5) - (n=1) N/A (n=54)
Outcome	DOD (n=20) NED (n=33) AWD (n=3) N/A (n=4)
M: Male, F: Female, N/A: Not applicable, RT: Radiotherapy, CT: Chemotherapy, DOD: Dead of disease, NED: No evidence of disease, AWD: Alive with disease	

DISCUSSION

Till today, about 40 case reports/series of extradural spinal ED have been reported, mostly occurring in adolescents and young adults and mostly in males. The most common presenting complaint is chronic back pain, paraparesis, and sphincter dysfunction later in the clinical course.

Most extradural ES involve multiple levels and may vary in location for each case, mostly presenting in the sacral region. CT scans best document dumb-bell-shaped tumors with foraminal widening,^[1] and scalloping of bones,^[6] soft tissue mass with or without pleural and rib damage.^[5] Periodic acid-Schiff stain was positive in all reported cases. On light microscopy, poorly differentiated, small round cells are seen with mitotic figures, a high N/C ratio, and scanty cytoplasm that form nests, sheets, lobules, or occasionally rosettes. On electron microscopy, scanty cytoplasmic organelles are seen, accompanied by growth cones suggestive of glial cell differentiation.^[5] About 95% of the EES cases show non-specific CD99 (MIC2) positivity.^[3] Other tumor markers that are usually ordered include CD-34, CD-20, CD-10, Tdt, and others. The t(11;22) (q24;q12) translocation is a chromosomal abnormality seen in 90% of cases of ES, which results in the formation of a fusion transcript.^[2] Treatment regimens include partial/gross total resection, followed by radiotherapy and chemotherapy. Cases not receiving any adjuvant therapy typically died only a few months later.

CONCLUSION

Extraskeletal Ewing sarcomas are exceedingly rare. The best outcomes are achieved with gross total resection, followed by adjuvant radiation and chemotherapy.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Farooq M, Mustafa B, Sultan KA, Ashraf M, Ashraf N, Siddique A. Extraosseous extradural ewing sarcoma of the thoracic spine: Case report and literature review. *Surg Neurol Int* 2021;12:542.