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Metastatic thoracic and lumbar intramedullary and extramedullary Ewing's sarcoma: a rare case report and literature review

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Abstract

Ewing sarcoma (ES) is a highly aggressive bone and soft tissue tumor that occurs mainly in young children and adolescents and is associated with primary and metastatic disease. Intramedullary ES (either primary or secondary) is rare, and the ideal management remains inconclusive. We herein report intramedullary and extramedullary metastatic ES in a single patient. A 46-year-old woman was referred to our outpatient clinic from the oncology clinic with progressive paraparesis and paresthesia for 1 week prior to presentation. She had developed left clavicular ES 2 years earlier for which surgery and chemoradiotherapy had been performed. At the present evaluation, she was diagnosed with intramedullary thoracic and lumbar extradural masses. Thoracic surgery was performed, and a biopsy of the lesion was obtained. The diagnosis of ES was confirmed histopathologically, and she underwent adjuvant chemotherapy. Her neurological status did not improve after surgery, and she underwent rehabilitation and physical therapy. The lumbar lesion resolved with chemotherapy. Metastasis of ES to the spinal cord, especially intramedullary lesions, is extremely rare, and there is no standard management guideline. However, surgical decompression and adjuvant chemotherapy are the main treatments in these cases.

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Introduction

Ewing sarcoma is a highly aggressive bone and soft tissue tumor that occurs mainly in children, with a survival rate of 70% to 80% in localized disease and 25% to 30% in metastatic disease.[1] Ewing sarcoma is the second most common malignancy in childhood and adolescence, and treatment comprises surgical resection, radiotherapy, chemotherapy, and re-do surgeries.[2] Most patients present with locoregional disease, and a small number present with overt metastasis. Although the most common site of Ewing sarcoma is the diaphysis of the long bones, tumors can arise in any bone and soft tissue, and the symptoms vary accordingly.[2]

Although metastasis of Ewing sarcoma to the central nervous system (CNS) as well primary CNS disease have been reported previously, involvement of the spinal cord is extremely rare and requires specific management and considerations.[3] To date, only 36 cases of intramedullary Ewing sarcoma have been reported (summarized in Table 1),[4–36] among which there were 31 primary cases[4–11,13,15–17,19–32,34,36] and 5 recurrent and metastatic cases.[12,14,18,33,35] Because the number of reported cases of intramedullary Ewing sarcoma is limited, disease management and patient survival data, as well as patient outcomes, remain inconclusive. Previously, intradural extramedullary cases of Ewing sarcoma were reviewed extensively;[37] however, intradural intramedullary cases of Ewing sarcoma have not been reviewed. In the current article, we report metastatic intramedullary thoracic Ewing sarcoma in an adult following complete treatment of a previously detected Ewing sarcoma.

Case presentation

A 46-year-old woman was referred to our outpatient clinic from the oncology clinic with progressive paraparesis and paresthesia for 1 week prior to presentation (August 2021). She was diagnosed with Ewing sarcoma in December 2019, and again, with left clavicular Ewing sarcoma approximately 2 years prior to presentation. She presented with a progressively enlarging neck mass on the left side that had developed within the previous 2 months. Spiral computed tomography (CT) of the chest and mediastinum revealed a large mildly enhanced mass measuring approximately 60×80 mm in the apex of the left lung with small extension to the base of the neck on the left side, just touching the tracheal wall. No signs of bone destruction or bone erosion were seen. The patient underwent surgery, and the tumor was totally excised and histopathologically proven to be Ewing sarcoma of the clavicle. She then underwent chemotherapy with vincristine sulfate, doxorubicin hydrochloride, and cyclophosphamide, followed by ifosfamide and etoposide phosphate (VAC/IE protocol) and subsequent radiotherapy. Positron emission tomography (PET) was performed in March 2021. The findings revealed mild soft tissue thickening in the right apical hemithorax with insignificant fluordeoxyglucose (FDG) activity demonstrating post-therapy changes with no

Table 1. Summary of cases of metastatic intramedullary Ewing sarcoma in the literature.

No	Author	Year	Age	Sex	Location	Outcome	Follow-up	Adjuvant therapy
Recurrent cases								
1	Current Case	2022	46	Female	Thoracic Conus Medullaris	Alive with disease	10 months	Chemoradiotherapy
2	Fukushima et al.[12]	2021	39	Male	Cervical	Died of disease	2.5 months	Chemoradiotherapy
3	Yurtsever et al.[35]	2016	51	Male	Thoracic	Alive with disease	2.5 months	Chemoradiotherapy
4	Jia et al.[18]	2009	28	Male	Thoracic	Alive with disease	N/A	Chemoradiotherapy
5	Gorgulu et al.[14]	2005	28	Male	Cervical	Alive with disease	N/A	Chemoradiotherapy
6	Weil et al.[33]	2001	21	Male	Thoracolumbar	Disease-free survival	30 months	Chemoradiotherapy
Primary cases								
1	Yamada et al.[34]	2020	23	Male	Cervical	Alive with disease	10 months	Radiotherapy
2	Chen et al.[7]	2019	16	Male	Cervicothoracic	Died of other cause	1 month	Chemoradiotherapy
3	Khwaja et al.[20]	2019	44	Female	Cervicothoracic	Alive with disease	3 years	Chemoradiotherapy
4	Wang et al.[32]	2017	26	Male	Thoracolumbar	Disease-free survival	14 months	Chemoradiotherapy
5	Coulibaly et al.[36]	2015	16	Male	Thoracolumbar	Alive with disease	2 years	Chemoradiotherapy
6	Alexiou et al.[5]	2013	2 m	Male	Cervicothoracic	Disease-free survival	9 months	N/A
7	Gollard et al.[13]	2011	21	Female	Thoracic	Disease-free survival	11 years	Chemotherapy
8	Ellis et al.[10]	2011	27	Female	Cervical	Disease-free survival	28 months	Chemotherapy
9	Benesch et al.[6]	2010	1.5	Female	Cervical	Died of disease	6 months	Chemotherapy
10	Benesch et al.[6]	2010	10 m	Female	Thoracic	Died of disease	6 months	Chemotherapy
11	Benesch et al.[6]	2010	2	Male	Thoracic	Disease-free survival	40 months	Chemotherapy
12	Tsutsumi et al.[31]	2010	39	Male	Thoracic	Died of disease	11 months	Radiotherapy
13	Otero-Rodríguez et al.[30]	2009	1.5	Male	Thoracic	N/A	6 months	Chemoradiotherapy
14	Han et al.[15]	2008	17	Male	Conus Medullaris	Died of disease	24 months	N/A
15	Kumar et al.[23]	2007	18	Male	Whole Spine	Alive with disease	6 months	Chemoradiotherapy
16	Kumar et al.[23]	2007	9	Female	Thoracolumbar	Died of disease	18 months	Chemoradiotherapy
17	De Tommasi et al.[8]	2006	38	Male	Thoracic	Died of disease	18 months	Radiotherapy
18	Jain et al.[16]	2006	54	Female	Cervical	N/A	N/A	Radiotherapy
19	Kampman et al.[19]	2006	3	Male	Cervical	Died of disease	7 days	None
20	Kim et al.[21]	2004	17	Male	Thoracolumbar	Disease-free survival	4 months	Radiotherapy
21	Albrecht et al.[4]	2003	29	Female	Thoracic	Disease-free survival	17 months	Chemoradiotherapy
22	Mawrin et al.[26]	2002	69	Male	Cervicothoracic	Died of disease	3 months	Radiotherapy

(continued)

Table 1. Continued.

No	Author	Year	Age	Sex	Location	Outcome	Follow-up	Adjuvant therapy
23	Mottl et al.[28]	2002	17	Female	Cervical Lumbar	N/A	N/A	Radiotherapy
24	Meltzer et al.[27]	1998	25	Male	Cervical Conus Medullaris	Died of disease	60 months	Chemoradiotherapy
25	Deme et al.[9]	1997	22	Female	Thoracolumbar	Alive with disease	15 months	Chemoradiotherapy
26	Kwon et al.[24]	1996	3 m	Female	Thoracolumbar	Alive with disease	15 days	Chemotherapy
27	Ogasawara et al.[29]	1992	16	Female	Lumbar	Disease-free survival	29 months	Chemoradiotherapy
28	Freyer et al.[11]	1989	7	Male	Thoracolumbar	Died of disease	20 months	Chemoradiotherapy
29	Jaksche et al.[17]	1988	26	Male	N/A	Died of disease	18 months	Chemoradiotherapy
30	Jaksche et al.[17]	1988	15	Female	N/A	Died of disease	36 months	Radiotherapy
31	Kosnik et al.[22]	1978	N/A	N/A	N/A	N/A	N/A	Chemoradiotherapy

Note: Age is listed in years unless otherwise stated.
m, months; N/A, not available.

remarkable active residual focus. The PET scan findings suggested a complete metabolic response; thus, the disease was considered cured.

On physical examination at the current presentation, the patient's muscle strength in her lower extremities was as follows: right: 3/5 and left: 1/5 (proximal/distal, respectively). There was a sensory level in the thoracic region (T6), and spastic hyperreflexia was present in the lower extremities (American Spinal Injury Association (ASIA) impairment scale C). Urinary sphincter function was intact, but the patient complained of incomplete evacuation of the bladder. A urodynamic study revealed a spastic bladder. Bilateral Babinski reflexes were upward. Emergency whole-spine contrast-enhanced magnetic resonance imaging (MRI) revealed a huge intramedullary thoracic lesion extending from T5 to T8 that was hyperintense in T2-weighted images (Figure 1a), with scattered enhancement after the injection of contrast (Figure 1b). Additionally, there was a round intradural extramedullary mass lesion at the L1 and L2 levels causing a compression effect on the conus medullaris that was isointense in T2-wighted images (Figure 1c), with homogenous enhancement after the injection of contrast (Figure 1d). The patient was diagnosed with multiple intramedullary and extramedullary thoracic and lumbar lesions, and surgery was scheduled to decompress the affected spinal segments. Surgery was performed with intraoperative neuromonitoring comprising motor evoked potentials (MEP), sensory evoked potentials (SEP), and electromyography (EMG). No neuronavigational instruments were used, and the affected spinal levels were cleared of lesions with the aid of intraoperative fluoroscopy (C-arm). The patient underwent laminectomy from T5 to T8.

The dura was opened, and midline myelotomy was performed in the thoracic

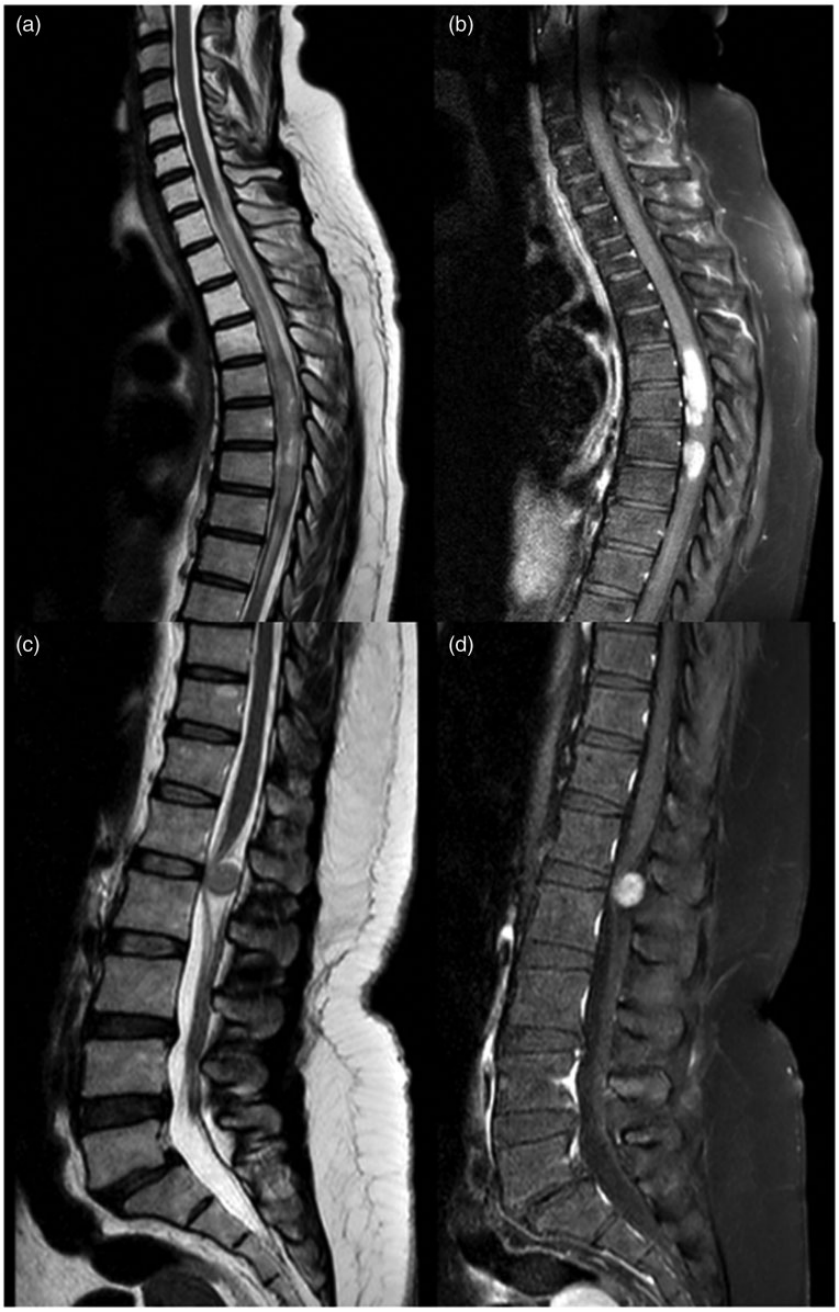

Figure 1. Sagittal T2-weighted MRI of the patient demonstrating an intramedullary lesion extending from T5 to T8 with hyperintense signals associated with increased spinal cord diameter (a) with scattered enhancement after gadolinium injection (b). Sagittal T2-weighted MRI of the lumbosacral spine showing a round intradural extramedullary isointense lesion compressing the conus medullaris (c) with homogenous enhancement after gadolinium injection.
MRI, magnetic resonance imaging.

region. The lesion was microscopically resected as much as possible (intralesional resection and multiple biopsies of the lesion). There was a soft, gray-to-white mass with poorly defined margins and poor consistency. Because the patient had appropriate spinal sagittal balance preoperatively, and we performed a limited laminectomy for tumor resection, no spinal fixation and fusion was performed. The patient's neurological status did not improve after the surgery (ASIA impairment scale C), and she began rehabilitation and physical therapy postoperatively. She also received temozolomide (100 mg/m^2/day orally) and irinotecan (40 mg/m^2/day intravenously) in accordance with the standard protocol as adjuvant therapy for recurrent or metastatic Ewing sarcoma. Postoperative imaging of the thoracic spine revealed appropriate decompression (Figure 2a and b) and showed that the lumbosacral lesion had completely resolved (Figure 2c).

Histopathological examination of the thoracic lesion revealed small round cells with moderate pleomorphism, nuclear atypia, and some true rosette formations in a pauci-vascular background (Figure 3a). Immunohistochemical staining of the tumor showed diffuse, strong immunoreactivity for cluster of differentiation (CD)99, and a high proliferative index (Ki-67: 85%–90%), with negative staining for leukocyte common antigen (LCA) (Figure 3b–d). The diagnosis of Ewing sarcoma was confirmed accordingly. At the 10-month follow-up, the patient's neurological status had worsened, with bilateral lower extremity muscle strength scores of 0/5 (proximal/distal, respectively; ASIA impairment scale B), with spasticity and urofecal retention. She was alive with disease at the time of this report. The patient provided written

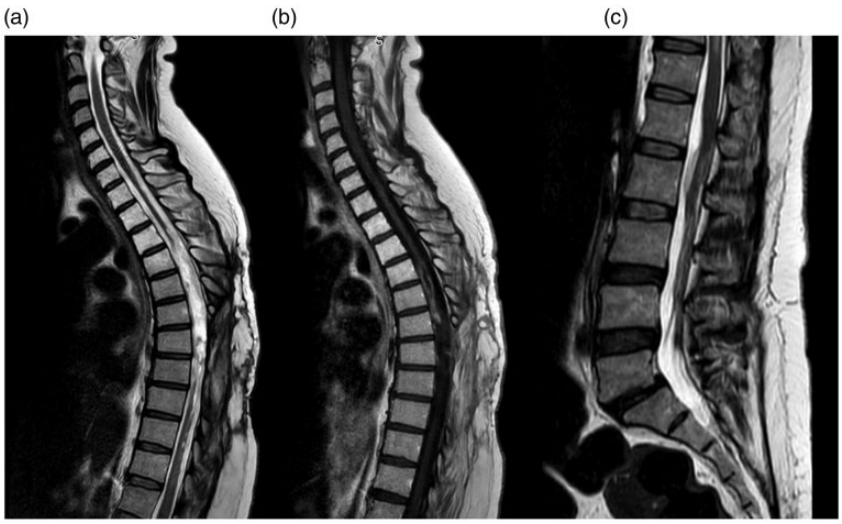

Figure 2. Postoperative sagittal T2-weighted cervicothoracic MR image of the patient showing an intramedullary lesion extending from T5 to T8 with hyperintense signals (a). Postoperative T-1 weighted MR image with gadolinium enhancement showing the lesion after internal resection and decompression (b). Post-chemotherapy T2-weighted MR image of the lumbosacral spine showing complete resolution of the extradural lesion (c).
MR, magnetic resonance.

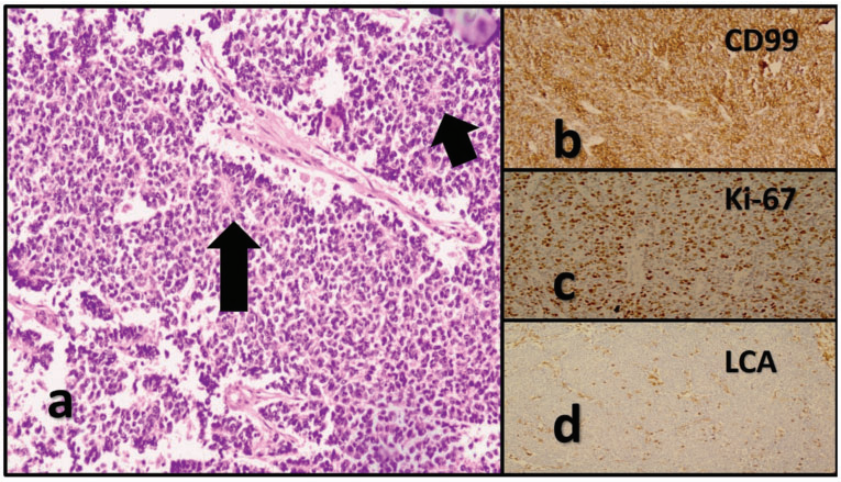

Figure 3. (a) Histopathological sections showing sheets of small round cells with moderate pleomorphism, nuclear atypia, and some true rosette formations (arrows) in a pauci-vascular background (×400, H&E) and (b–d) Immunohistochemical staining of the tumor showing diffuse, strong immunoreactivity for CD99, and a high proliferative index (Ki-67) with negative staining for LCA (×100, ×100, ×40; b–d, respectively).
H&E, hematoxylin and eosin; CD99, cluster of differentiation 99; LCA, leukocyte common antigen.

informed consent for the publication of her case, and the reporting of this study conforms to the CARE guidelines.[38]

Discussion

Metastasis of Ewing sarcoma to the spinal cord is extremely rare, and the management and clinical significance is controversial, with no appropriate level of evidence.[2] Ewing sarcoma accounts for 10% to 15% of malignant bone tumors and 40% to 45% of all malignant tumors in children.[1,2] Ewing sarcoma can arise in any bone or soft tissue; however, tumors in the spinal cord are rare.[2] We reviewed the current literature and identified 36 cases of intramedullary Ewing sarcoma that were either primary or metastatic (Table 1). Most of the cases were primary, and only five recurrent cases from other organs have been reported. We herein report a unique and rare case of metastatic Ewing sarcoma to both intramedullary and extramedullary sites in a single patient. The thoracic lesion was histologically revealed to be Ewing sarcoma in accordance with the immunohistochemical examination findings. The patient had been diagnosed with Ewing sarcoma 2 years prior to presentation and underwent surgery and chemoradiotherapy. She had no evidence of the disease 6 months prior to the current presentation. However, subsequently, she experienced a very progressive and malignant course of metastasis to the spinal cord. To the best of our knowledge, this is the only reported case of both intramedullary and extramedullary metastasis of Ewing sarcoma.

We performed a systematic review of the published literature in accordance with the preferred reporting items for systematic reviews and meta-analyses (PRISMA) guideline.[39] We searched the electronic databases, MEDLINE/PubMed, Embase, Scopus, Cochrane Library, and Web of Science (WOS) to 10 January 2022. We used the

following Medical Subject Heading (MeSH) search terms, text words, and keywords: ['Ewing sarcoma' or 'cord Ewing' or 'intramedullary Ewing' or 'extramedullary Ewing'] AND ['recurrent' or 'metastatic'] in the English language, without date restrictions. We also manually searched the bibliographic lists of previously published review articles to identify related studies. All reference management was conducted using EndNote X8 for Windows (Clarivate Analytics, Philadelphia, PA, USA). We identified 36 articles reporting intramedullary or extramedullary Ewing sarcoma and these were included in the current systematic review.

The identified cases showed that intramedullary Ewing sarcoma was located mainly in the thoracic and lumbar regions, although cervical and sacral locations were also reported.[4–36] The current case had a thoracic intramedullary lesion and a lumbar extramedullary lesion, both causing neural compression and deficits.

The clinical findings of spinal cord metastasis of Ewing sarcoma are similar to those of other lesions of the spinal cord, namely muscle weakness, sensory level, hyperreflexia, and involvement of sphincters in the form of spasticity. Chronic lesions cause spasticity and muscle wasting.[3] A review of the literature and a meta-analysis of the extramedullary cases of Ewing sarcoma by Saeedinia et al.[37] revealed that Ewing sarcoma should be considered among the differential diagnoses of intradural lesions despite their rarity, especially in patients with a previous history of the disease.

Gadolinium-enhanced spinal MRI remains the modality of choice for the diagnosis of Ewing sarcoma.[40] The lesions are mainly hyperintense in T2-weighted images and isointense in T1-weighted images. After contrast injection, segmental and scattered enhancement are observed.[3,35] However, histology remains the gold standard for the diagnosis of Ewing sarcoma, and the examination should include immunohistochemical staining and molecular analysis comprising fluorescent in situ hybridization (FISH) and real-time polymerase chain reaction (RT-PCR) to identify and characterize *EWSR1* translocation.[41]

The treatment of Ewing sarcoma of the spinal cord lacks a standard and specific protocol, and the standard of care remains elusive.[12] Treatment comprises surgical resection (as much as possible while maintaining the neural anatomy) followed by chemoradiotherapy.[2] The prognosis and survival rate for patients with spinal cord Ewing sarcoma is not well known because of the rarity of the condition; however, based on previous cases (Table 1), the survival rate was 45% to 50% in primary cases and 30% to 35% in recurrent and metastatic cases.[4–36] In surviving cases, rehabilitation and physical therapy should be considered part of the treatment program to improve the patient's neurological status. In the current case, the patient improved markedly after 4 months of physical therapy and rehabilitation.

Conclusion

This is the first case of intramedullary and extramedullary spinal cord metastasis of Ewing sarcoma in a single patient. The patient was treated by surgical resection followed by adjuvant chemotherapy, physical therapy, and rehabilitation. Although rare, Ewing sarcoma should be considered among the differential diagnoses of spinal cord lesions in patients with neurological deterioration, especially in those with a history of Ewing sarcoma. Surgical resection and chemoradiotherapy are the main treatments; however, there is no standard guideline for the treatment of spinal cord Ewing sarcoma.

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Declaration of conflicting interest

The Authors declare that there is no conflict of interest.

Ethics statement

As a case report, no institutional review board (IRB) approval was required. The patient provided informed written consent to publish her case.

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