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SNI: Spine

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Case Report

Primary spinal epidural rhabdomyosarcoma: A case report

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ABSTRACT

Background: Rhabdomyosarcoma (RMS) is a malignant childhood tumor that most commonly involves the skeletal muscles of the head and neck, genitourinary tract, limbs, and, rarely, the spine.

Case Description: A 19-year-old male presented with cauda equina symptoms. Magnetic resonance imaging showed a C7/T1 homogeneously enhancing lesion, causing pathological fracture of the T1. Similar lesions were found on T3 and S1-S2 levels. CT-guided biopsy and immunohistochemistry confirmed the diagnosis of highly malignant alveolar RMS. He underwent multi-level laminectomies with partial tumor debulking but was paraplegic postoperatively.

Conclusion: Spinal RMS rarely involves the soft tissues of the spine and should be surgically resected if feasible. Nevertheless, the long-term prognosis is poor regarding tumor recurrence and metastases.

Keywords: Chemotherapy, Neurosurgery, Radiotherapy, Rhabdomyosarcoma, Spine

INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant tumor originating from mesenchymal cells that commonly occur in the head and neck region, followed by the genitourinary tract and extremities but rarely the spine of children. It is the third most common extracranial tumor after Wilms' tumor and neuroblastoma. It occurs in 4–7 cases/one million children under the age of 15 years/per year. Sixty-five percentages of cases are diagnosed in children before age six, with very few reported in adults.[2,9] Here, we summarized our diagnosis and management of a 19-year-old male with multifocal spinal RMS and previously reported cases in the literature in [Table1]. Included cases were supplemented by reports summarized by Wang *et al.* added in the supplementary material of their paper.

CASE PRESENTATION

A 19-year-old male presented with a 1-month onset of numbness/weakness in both hands and progressive low back pain, gait disturbance, and 4 days of urinary incontinence. On examination, he exhibited mild quadriparesis.

Investigations

Magnetic resonance imaging (MRI) studies demonstrated multifocal involvement of the spine, likely due to metastases. The cervical MRI showed a right anterolateral preverbal/vertebral C7/

Table 1: Summary of our case and the previously reported cases of spinal rhabdomyosarcoma.

#.	Author	Sex/ Age	Tumor location	Imaging	Management	Subtype	Outcome
1.	Our Case	M/19	Anterior epidural C7-T1 and S1-S2	Hypo T1, Hyper T2 Homogenous enhancement	CT guided biopsy partial resection+CT	Alveolar	Slight improvement after 2 months
2.	Khalatbari *et al.*	F/13	Posterior epidural T11-T12+L3 vertebral body	Isointense T1 and T2 Homogenous enhancement	Total resection+RT+CT	Alveolar	No tumor recurrence or metastasis after 1 year
3.	Wang *et al.*	M/17	C3	N/R	Radical resection+CT	Embryonal	Died after 4 months
4.	Wang *et al.*	F/49	Posterior epidural L5	Hypo T1, Hyper T2 Homogenous enhancement	Radical resection+CT+RT	Pleomorphic	No relapse or metastasis after 8 months
5.	Wang *et al.*	F/15	Anterior epidural T9-T11	Hypo T1, Hyper T2 Homogenous enhancement	Radical resection+CT+RT	Embryonal	Died after 18 months
6.	Wang *et al.*	M/57	L5	Hypo T1, Hyper T2 Homogenous enhancement	Radical resection+CT+RT	Pleomorphic	No relapse or metastasis after 8 months
7.	Wang *et al.*	M/32	Intraspinal T2-T7	Hypo T1, Hyper T2 Homogenous enhancement	Radical resection+CT+RT	Alveolar	Died after 5 months
8.	Wang *et al.*	M/45	T10-T12	Hypo T1, Iso T2 Homogenous enhancement	Radical resection+CT+RT	Pleomorphic	Died after 9 months
9.	Wang *et al.*	M/5	L5-S2	Hypo T1, Hyper T2 Homogenous enhancement	Radical resection+CT+RT	Embryonal	Died after 7 months
10.	Wang *et al.*	M/45	T9-11	N/R	Paliative resection+CT	Sclerosing	Died in 8 months
11.	Wang *et al.*	F/21	T7	N/R	Palliative resection+CT+RT	Alveolar	Died in 10 months
12.	Wang *et al.*	M/42	T11	N/R	Radical en Bloc resection+CT+RT	Embryonal	Died in 11 months with metastasis
13.	Wang *et al.*	M/52	L5	N/R	Palliative resection+CT	Pleomorphic	No relapse or metastasis in 14 months
14.	Rumboldt *et al.**	M/6	Posterior epidural C5-T2.	Hypo T1, Hyper T2 Homogenous enhancement	Total resection+RT+CT	Embryonal	Diffuse meningeal metastases
15.	Tsitsopoulos *et al.**	M/52	T1-T3 epidural	N/R	Laminectomy, total resection+RT	N/R	No recurrence within 1 year
16.	Haisa *et al.**	F/77	C5-C7 epidural	N/R	Laminectomy+ subtotal resection+CT	Alveolar	Partial remission
17.	Saito *et al.**	F/7	L3 vertebral body	N/R	L3 en bloc total resection+ Pre-operative CT+ Intra-operative RT+ post-operative CT	Embryonal	No recurrence within 6 years

(Contd...)

Table 1: *(Continued).*

#.	Author	Sex/ Age	Tumor location	Imaging	Management	Subtype	Outcome
18.	Wagner *et al.**	F/2	L5-S2 intramedullary	N/R	Biopsy and sectioning of filum terminale+ CT	Embryonal	Slow but marked improvement of lower limb
19.	Ogzu *et al.**	F/9	L4-L5 epidural	N/R	Macroscopic total resection+ CT+RT	Embryonal	N/R
20.	Fountas *et al.**	M/47	T8-T9 epidural	N/R	Decompressive laminectomy+ subtotal resection	Pleomorphic	Diffuse metastasis of all vertebrae after 6 weeks
21.	Spalteholz and Gulow	F/58	T11-T12 vertebral body	N/R	Total enbloc resection+CT+RT	Pleomorphic	No recurrence after 6 months
22.	Sequea *et al.**	F/69	L2-L4 epidural	N/R	Laminectomy+ resection	Spindle-cell	Discharged to palliative care after 8 months
23.	Hakozaki *et al.**	F/16	S2-S3	N/R	Incisional biopsy+CT+RT	Embryonal	Died after 17 months
24.	Sundaresan *et al.**	F/44	Para-spinal tumor T12-L3	N/R	Pre-operative CT+surgery	Pleomorphic	Disease free and neurologically intact
25.	Watanabe *et al.**	F/1	C3-5 epidural (primary)+ T12 subdural (metastasis)	N/R	Biopsy CT+RT	Embryonal	Died after 3 months
26.	Shapeero *et al.**	F/12	Thoracic and lumbar spine, pelvis, ribs, clavicle (no primary)	N/R	Biopsy+CT+RT	Alveolar	Died after 3 months
27.	Shapeero *et al.*	F/15	Thoracic and lumbar spine, pelvis, femur (no primary)	N/R	Biopsy CT+RT	Alveolar	Living 14 months later with diffuse metastasis
28.	Das *et al.**	M/13	T9-12 vertebral and intradural, epidural space (Hand)	N/R	Decompression laminectomy+ excisional biopsy CT+RT	Alveolar	Died a few months later
29.	Raney *et al.**	F/8	T1-7 intradural (retropharyngeal)	N/R	Decompression laminectomy+ excisional biopsy CT+RT	Embryonal	Died 2 months after discovery of the CNS extension
30.	Sarkar *et al.**	M/18	Diffuse metastasis including spine (pelvis)	N/R	Biopsy CT+RT	Alveolar	Died 3 weeks after the initiation of chemotherapy
31.	Sparreboom *et al.**	M/50	Bony metastatic disease throughout the lumbar spine (thigh)	N/R	CT-guided biopsy+RT	Pleomorphic	Died 1 week after diagnosis
32.	Yavas *et al.**	F/6	Spinal cord intramedullary (para-meningeal)	N/R	Biopsy+CT+RT	Not reported	Died 2 weeks after initiation of RT
33.	Keorochana *et al.**	F/16	spinal and bilateral breast metastases (Hand)	N/R	Decompression+ instrumentation CT+RT	Embryonal	Died at 3 weeks after surgery
34.	Thomas *et al.**	M/10	Thoracic and lumbar spine	N/R	Biopsy CT	Alveolar	Died 12 months after CT began

M: Male, F: Female, C: Cervical, T: Thoracic spine, L: Lumbar spine, S: Sacral, N/R: Not reported, CT: Chemotherapy, RT: Radiation therapy. *: Included cases were supplemented by reports summarized by Wang *et al.* added in the supplementary material of their paper.

T1 homogeneously enhancing lesion, a pathologic fracture of the T1 body, and epidural compression [Figure 1]. A similar lesion was found at the S1-S2 level, contributing to the cauda equina compression [Figure 2]. A third small lesion was found in the right T3 vertebral pedicle [Figure 1]. The differential diagnoses included multifocal spinal lymphoma versus multiple metastases.

CT-guided biopsy

A CT-guided biopsy of the sacral lesion confirmed the diagnosis. Furthermore, the immunohistochemistry revealed a highly malignant RMS [Figure 3]. The primary spinal origin of the RMS was confirmed by negative CT scans of the chest, abdomen, and pelvis.

Management

Before the initiation of planned chemotherapy/radiation, the patient abruptly developed paraplegia and underwent multi-level decompressive laminectomies for partial debulking of the cervicodorsal and sacral lesions. He remained paraplegic postoperatively and failed to improve over the next 2 months.

DISCUSSION

RMS is the most common soft-tissue sarcoma in the pediatric population, accounting for approximately 5% of all pediatric cancers.[5,9] RMS is the third most common type of extracranial solid tumor found in children, following Wilms' tumor and neuroblastoma.[4] The peak incidence occurs in children under the age of six. Primary spinal RMS can occur in any section of the spine, from the cervical to the sacral region; however, the lumbar region is the most common site of involvement.[8] Primary spinal RMS is exceedingly rare; 20% experience distant metastases, primarily in the lungs (39%), bone marrow (32%), lymph nodes (30%), and bones (27%).[1] Symptoms/signs associated with RMS reflect

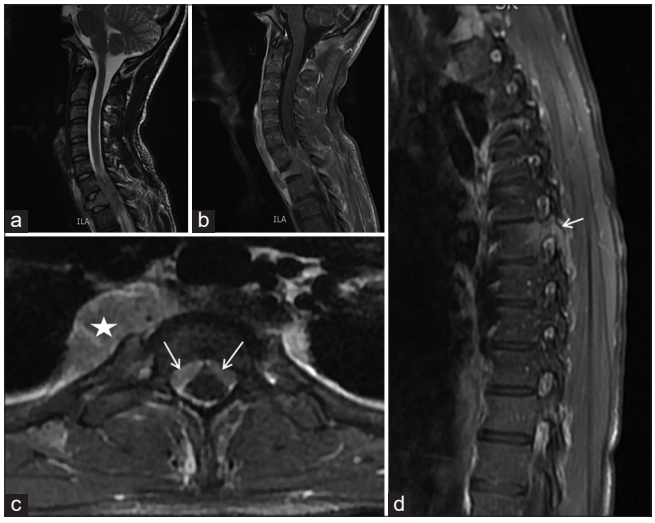

Figure 1: Cervical and dorsal magnetic resonance imaging in sagittal T2-weighted sequence (a) and T1-weighted sequence with gadolinium (b and d), and in axial T2-weighted sequence at the level of the first dorsal vertebra (c). It shows the pathologic fracture of the latter with the epidural extension of the rhabdomyosarcoma (a and b), also the prevertebral right-sided part of the tumor (star) and its anterior bilateral invasion of the canal causing cord compression (arrows). Note the infiltration of the right pedicle and lower facet of the third dorsal vertebra (d arrow).

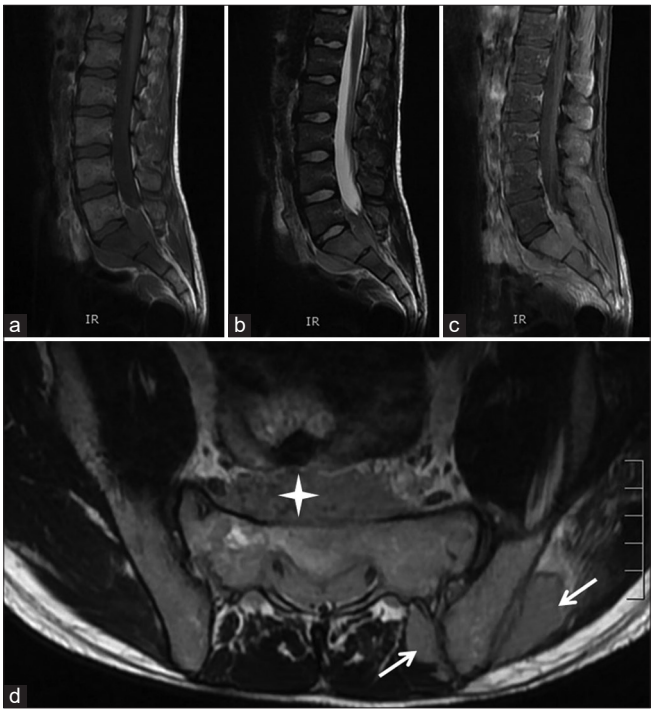

Figure 2: Lumbosacral magnetic resonance imaging in sagittal cuts T1-weighted sequence (a), T2-weighted sequence (b), and T1 with gadolinium (c), and axial cut T2-weighted sequence, focused on the sacrum. It shows the tumor starting in the pre-sacral space (star), infiltrating the sacrum mainly to the left ala with sacral canal invasion and gluteal muscle infiltration (d arrows).

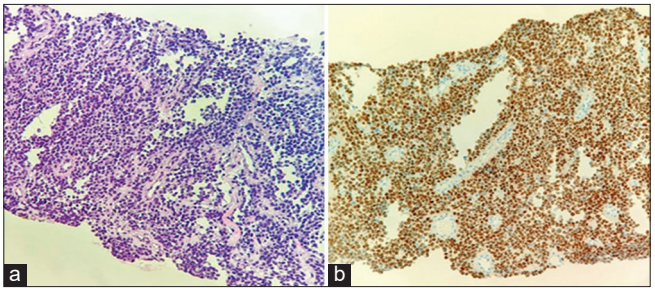

Figure 3: Photomicrographs of our patient (a) with H-E staining, magnification ×20, showing high cellular proliferation of primitive round cells with nested and alveolar patterns. (b) immunohistochemistry study, showing extensive nuclear positivity of tumor cells to myogenin.

the involved spinal levels. The diagnosis of RMS is based on clinical, radiographic, and histopathologic studies.

Radiographic

CT and MRI are the most commonly used imaging modalities in evaluating RMS; however, the findings are variable and non-specific for RMS.[8] The lesion is typically hypointense to iso-intense on T1-weighted images and hyperintense on T2-weighted images while enhancing homogeneously with contrast material in all reported cases. The primary RMS usually infiltrates the vertebral bodies and invades the spinal cord, eventually resulting in pathological fractures associated with cord compression.[3,6]

CT-guided biopsy/pathology

CT-guided biopsy or surgical excision can obtain tissue sampling for the histopathologic examination. According to the current World Health Organization classification, RMS is classified into four subtypes: Embryonal, spindle cell, alveolar, and pleomorphic. The embryonal type is the most common in primary RMS. However, in metastatic RMS, the alveolar subtype is typically found. The alveolar type represents 20% of RMS cases and is associated with a poorer prognosis.[3,7,8]

Management

Treatment typically involves a combination of surgery, radiation therapy, and chemotherapy. At present, there is no consensus on the most effective treatment. Few studies have documented the efficacy of surgery in managing spinal RMS. Wang *et al*. evaluated 11 primary and secondary RMS cases with posterior locations (eight of the 11 cases); complete surgical resection and spinal fixation with adjuvant radiotherapy provided a longer median overall survival versus palliative surgery/conservative treatment. Therefore, surgery is recommended to treat spinal RMS to reduce the neoplastic mass and allow for a better response to radiation therapy and chemotherapy.[6,8]

Radiation therapy is indicated after surgery to eliminate all residual tumor cells and reduce the risk of local recurrence. Chemotherapy is typically given before or after surgery or in combination with radiation therapy.[2]

CONCLUSION

RMS is a malignant tumor that is exceedingly rare in the spine. Here, a 19-year-old male with primary multifocal/metastatic spinal RMS (C7/T1, T3, and S1/S2) developed paraplegia before radiation and/or chemotherapy could be instituted.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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