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Case Report

Synovial sarcoma of the spine: A case report and review of the literature

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ABSTRACT

Background: Synovial sarcoma (SS) of the spine is a rare malignant soft-tissue tumor, and there are few reported cases. The aim of this paper is to report a rare case of spinal SS involving the paraspinal muscles, and to review all such cases reported in the literature.

Case Description: In this paper, we report a rare case of spinal SS involving the paraspinal muscles in a 12-year-old girl. The patient underwent surgical excision of the mass with adjuvant radiation and chemotherapy. At the 1-year follow-up, there was no evidence of local tumor recurrence, and the patient's symptoms had improved. In addition, we identified and reviewed 33 reported cases of SS involving the spine.

Conclusion: Due to the limited number of reported cases in the literature, it is difficult to predict the outcomes of spinal SS. Further, different treatment modalities have been used to treat spinal SS. However, most of the reported cases had poor outcomes. Therefore, prospective multi-center studies are needed to further investigate the treatment strategies and outcomes for patients with spinal SS.

Keywords: Hemangiopericytoma, Paraspinal, Spinal synovial sarcoma, Spinal tumor, Spinal tumors

INTRODUCTION

Synovial sarcoma (SS) is a rare malignant soft--tissue tumor accounting for 5–10% of all soft-tissue tumors.[16] Approximately 60–80% of all SS patients are young adults and adolescents, with a higher prevalence in men.[22] SS can occur anywhere in the body, including the brain, prostate, and heart. Approximately 80% of SSs arise in the deep soft-tissue of the extremities, but SS rarely arises in the spine, with spinal SS accounting for <5% of all reported SS cases. In addition, SS can be intradural or extradural, but intradural SS is very rare.[11] Histologic confirmation is required for the diagnosis of SS because differentiation from other types of spinal tumors is difficult.[3,4] Chromosome translocation characteristics are found in most SSs. This involves the fusion of the *SYT gene* to one of *SSX genes* (*SSX1* and *SSX2*). These gene translocations allow for several histopathological variants of SS, including monophasic, biphasic, and poorly differentiated forms.[21] However, there are only a few reported cases of spinal SS. The aim of this paper is to report a rare case of spinal SS involving paraspinal

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muscles and to review all the cases reported in the literature.

CASE DESCRIPTION

A 12-year-old female presented to our neurosurgery clinic with mid-lower back pain radiating to the left frontal aspects of the thigh. Four months before the presentation, the patient discovered a lump in the lower aspect of her back. The lump was small and increased in size. Her medical and surgical history was unremarkable. Physical examination revealed a large paravertebral lump (~4 × 5 cm) in the mid-lower aspect of the back. The lump was tender to light palpation and hard inconsistency, with irregular borders and no skin changes or muscle atrophy. Neurological examination of the lower limbs revealed normal tone, power, and intact sensation. Imaging studies (spinal magnetic resonance imaging [MRI] and computed tomography [CT]) revealed a left paraspinal soft-tissue mass extending from the T12–L1 level to the L4 vertebral level [Figures 1 and 2]. The patient underwent surgery for an excisional biopsy. The tumor was heavily vascular, and approximately 3.5 L of blood was collected intraoperatively. Postoperatively, the patient was neurologically intact and hemodynamically stable. Histopathology revealed multiple pieces of gray-tan, soft-tissue measuring 13 × 12 × 3 cm in total. The largest piece measured 8 × 7.5 × 2 cm and exhibited areas of hemorrhage and cystic degeneration, and areas covered by membranous tissue. Microscopic examination revealed malignant spindle cell proliferation forming sheets of cells with a prominent staghorn (hemangiopericytoma) vascular pattern [Figure 3]. Molecular analysis revealed a hybridization pattern of break-apart SYT-specific probes, indicating an SYT rearrangement; thus, a diagnosis of monophasic SS was made. The patient then received adjuvant chemotherapy with ifosfamide and doxorubicin hydrochloride in addition to radiation therapy. At the 1-year follow-up, there was no evidence of local tumor recurrence, and her symptoms had improved.

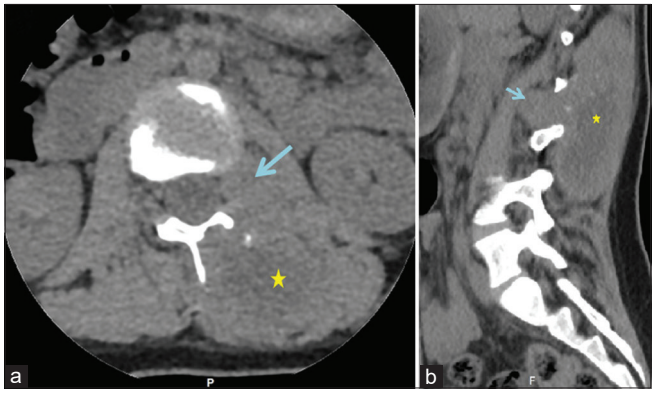

Figure 1: Selected computed tomography images of the spine. Axial (a) and sagittal (b) views demonstrate the left paraspinal soft-tissue mass (*) with an area of calcification that is extended from the level of T12/L1 down to the L4 level vertebra. Note the bone erosion changes in the lamina at the axial plane and the extension into the L1/L2 left neural foramina (arrows).

DISCUSSION

Sarcomas are a group of heterogeneous tumors that predominantly arise from the embryonic mesoderm. One type of sarcoma is soft-tissue sarcoma (STS), which can occur nearly anywhere in the body, but most commonly in the extremities. A rare histological subtype of STS is SS, which accounts for 10% of all STS cases. Despite its name, it does not arise from the synovial membrane.[1,5] SS primarily affect younger adults, and is more prevalent between the ages of 15 and 40 years, peaking in the third decade of life, which is unusual as most STSs appear in the 50s.[11,21,22]

Primary SS of the spine is particularly rare, with few case reports in the literature, and its etiology remains unclear. It can arise from the paraspinal muscles, paravertebral regions, or epidural spaces.[22] Histologically, there are three distinctive types of growth in SS: Monophasic, biphasic, and poorly differentiated. The monophasic type has only a spindle cell

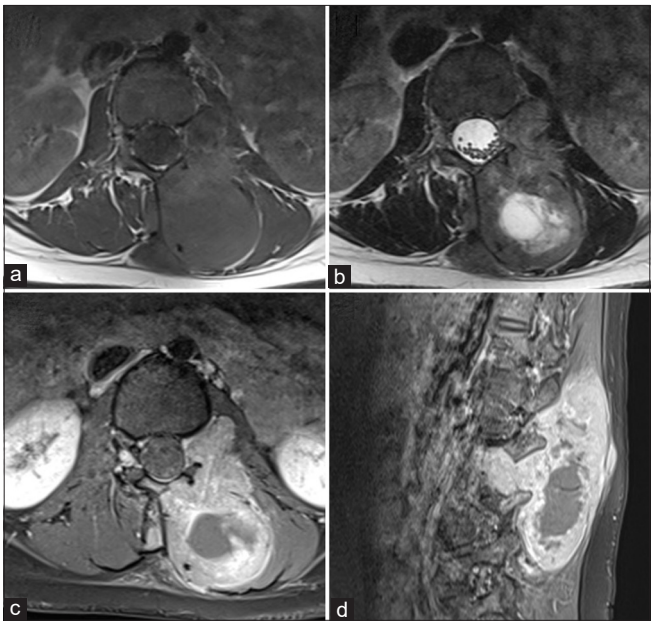

Figure 2: Selected magnetic resonance images of the lumbosacral spine. Axial T1- and T2-weighted images at the L1/L2 level in the left neural foramina (a and b) demonstrate the left paraspinal soft-tissue mass causing erosion of the left side lamina and the spinous process of the L2 vertebra with extension through the neural foramina. No involvement of the cauda equina terminal nerve roots is noted. Axial and Sagittal T1 FS-weighted images (c and d) at the same level demonstrate soft-tissue mass enhancement.

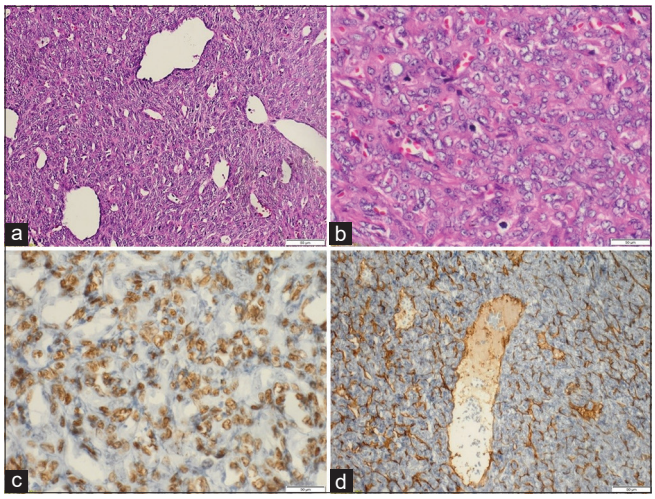

Figure 3: Histopathological description. Malignant spindle cell proliferation, forming sheets of cells with a prominent staghorn (hemangiopericytoma) vascular pattern (a). The cells display plump spindles to ovoid monomorphic nuclei with open chromatin. Mitotic figures are noted (b). Tumor cells are diffusely positive for TLE 1 (c). CD34 and smooth muscle actin highlight the prominent vascular network (d).

component, whereas the biphasic type has a characteristic mixture of glandular and spindle cell components.[5,12,22]. On plain films, there are no findings in up to 50% of patients unless the adjacent bony structures are eroded, or if there is visible calcification in the soft-tissue mass, which can be observed in up to 33% of patients.[17,21] CT can identify subtle calcification or local bony changes. Moreover, it is useful in the assessment of complex boney areas such as the spine and its relation to the SS. On CT imaging, SS can appear as a hypodense, well-demarcated mass with heterogeneous or homogenous enhancement, which makes distinction from other benign or malignant lesions difficult.[17,21] In our case, calcifications were noted on CT in addition to bony erosions [Figure 1]. MRI is generally considered the modality of choice for the staging and detection of soft-tissue tumors. On MRI, SS may appear hypointense or isointense relative to muscles and slightly hyperintense relative to fat, with a vaguely oval shape with sharp margins. The cystic appearance may lead to misdiagnosis of hematoma or other similar benign cystic masses.[17]

Nerve sheath tumors such as neurofibroma, schwannoma, or malignant peripheral nerve sheath tumor (MPNST) are among the differential diagnosis in the presence of solid lesions.[25] Schwannomas may be distinguished from SS by their eccentricity and encapsulation within the nerve bundle and can exhibit a mottled appearance on T2-weighted MRI. Neurofibromas are centrally positioned in relation to the nerve bundle with poorly defined borders and have nonhomogeneous signal intensity on T2-weighted MRI.[24,26,27] Intraneural SSs have been misdiagnosed as MPNST due to their irregular margins and association with individual nerves.[17]

SS of the spine presents in several histopathological forms. Approximately 70% of all cases reported in the literature are monophasic, and the remaining 30% are either biphasic or poorly differentiated SS. Some cases, including the current case, exhibited characteristics of hemangiopericytoma patterns, and this may explain the high intraoperative bleeding.[2,9,19,20] Approximately 80% of SS cases exhibited a positive reaction to cytokeratin, vimentin, CD99, and epithelial membrane antigen in immunohistochemical analysis. In addition, BLC2, CD56, and EMA were positive in several cases.[12,14,18] Negative results for CD34, S100, muscle-specific actin, and desmin make the diagnosis of fibrous, neural, skeletal, or smooth muscle tumors unlikely. In the cytogenetic analysis, almost all reported cases had a characteristic chromosomal translocation, t(X; 18) (p11.2; q11.2), with the resultant fusion of the *SYT* and *SSX* genes, making the diagnosis of SS highly likely.[6,7,8,10]

Surgical excision with negative margins of the tumor is considered the most effective treatment for SS; however, excision cannot be performed in most of the cases due to the important adjacent structures such as the spinal cord and spinal nerves. The use of adjuvant radiation and chemotherapy has been demonstrated to reduce local recurrence in several cases.[5,13,15,16] Most of the cases reported in the literature underwent surgical excision with either radiation or chemotherapy alone or in combination.

There was no significance difference in the outcome among different treatment strategies. However, patients who underwent surgical excision alone experienced greater recurrence with distal metastases compared to other combined treatment strategies (most commonly lung metastases).[12,16,23] Six months follow-up was the most common follow-up period reported. Most of the cases had no evidence of tumor recurrence, and their symptoms improved at 6 months. The disease-free survival period varies among reported cases. The average follow-up period in all cases was 24 months, with the exclusion of two cases with 11- and 22-years follow-up, which may suggest a different diagnosis. Almost all cases had poor outcomes with either death from the disease or distant metastases [Table 1].

CONCLUSION

Due to the limited number of reported cases in the literature, it is difficult to predict the outcomes of SS of the spine. Different treatment modalities have been used to treat spinal SS. However, most of the reported cases had poor outcomes. Therefore, prospective multi-center studies are needed to further investigate the treatment strategies and outcomes for patients with spinal SS.

Table 1: Review of spinal synovial sarcoma at our institution and reported cases in the literature by year.

Author/year	Age (years)/Sex	Location	Treatment	Outcome
Our case report 2020	12/F	T12–L4	Surgery+Chemotherapy+Radiotherapy	At 1-year follow-up, no evidence of local tumor recurrence
Rose 2018[16]	59/F	T4–T6	Surgery+Chemotherapy+Radiotherapy	No evidence of tumor recurrence at 67 months
Rose 2018[16]	54/ F	T10	Chemotherapy	The patient died after 4 months
Rose 2018[16]	32/F	T1–T2	Surgery only	At 6-month follow-up, lung metastasis was noted
Subramanian 2018[20]	46/F	T7–T8	Surgery+Chemotherapy+Radiotherapy	At 6-month follow-up, no evidence of tumor recurrence. Symptoms improved
Najib 2018[13]	44/M	T12	No information available	No information available
Yang 2016[24]	20/M	C2	Surgery	Refused treatment and died 1 month later
Guo 2016[6]	10/M	T9–T10	Surgery+Chemotherapy+Radiotherapy	At 6-month follow-up, no evidence of tumor recurrence. Symptoms improved
Peia 2013[15]	7/M	L4–L5	Surgery+Chemotherapy+ Radiotherapy	At 5-year follow-up, no evidence of tumor recurrence. Symptoms improved
Kim 2013[8]	29/M	C2–C3	Surgery+Radiotherapy	At 6-month follow-up, no evidence of tumor recurrence. Symptoms improved
Yonezawa 2012[26]	23/F	L3–L4	Surgery+Radiotherapy	At 5-year follow-up, no evidence of tumor recurrence. Symptoms improved
Kim 2012[7]	17/F	C3	Surgery+Chemotherapy	Authors did report follow-up for the patient
Zairi 2011[27]	36*	C1–C2	Surgery+Radiotherapy+Chemotherapy	Patients died 6 years later, after recurrence of disease
Naphade 2011[14]	14/M	C6–C7	Surgery only	At 6-month follow-up, no evidence of tumor recurrence. Symptoms improved
Verbeke 2010[23]	50/M	T12	Surgery only	Patient died after 22 years from local recurrence and metastasis
Verbeke 2010[23]	21/F	Sacrum	Surgery only	At 5-year follow-up, no evidence of tumor recurrence
Verbeke 2010[23]	40/M	Sacrum	Surgery only	At 5-year follow-up, no evidence of disease
Verbeke 2010[23]	31/F	L4	Surgery only	At 4-year follow-up, no evidence of disease
Verbeke 2010[23]	44/M	Sacrum	Chemotherapy+Radiotherapy	Patients died 7 years later
Verbeke 2010[23]	55/F	Sacrum	Radiotherapy only	At 11-year follow-up, no evidence of tumor recurrence
Liu 2010[10]	12/M	S2	Surgery+Radiotherapy	Patients died 2 years later, after recurrence of disease
Ravnik 2009[18]	32/M	T12–L1	Surgery+Chemotherapy+Radiotherapy	Patients died 9 months later, after recurrence of disease
Koehler 2009[9]	60/M	T7–T10	Surgery+Radiotherapy	At 9-month follow-up, no evidence of tumor recurrence
Barus 2009[1]	14/F	L2–S1	Surgery+Chemotherapy+Radiotherapy	At 5-year follow-up, no evidence of tumor recurrence
Mullah 2008[12]	14/F	L3–L4	Surgery+Radiotherapy+Chemotherapy	Pulmonary metastases noted after six cycles of chemotherapy
Sakellaridis 2006[18]	36/F	Lumbar	Surgery+Radiotherapy+Surgery	Patient died of the disease 1.5 years later after final surgery
de Ribaupierre 2006[4]	11/F	C6–C7	Surgery+Radiotherapy+Chemotherapy	Local recurrence 3 years later. No outcome reported
Greene 2006[5]	11/F	L2–L4	Surgery+Radiotherapy+Surgery	Patient died of the disease 14 months after diagnosis
Suh *et al.* 2005[21]	44/M	L4–L5	Surgery+Radiotherapy	Patient's symptoms improved at time of report. No long-term follow-up
Chu 2004[3]	11/F	C6–C7	Surgery+Radiotherapy	Local recurrence 3 years later. No outcome reported
Morrison 2001[11]	53/F	C7–T3	Surgery only	No outcome or follow-up reported
Signorini 1986[19]	59/M	T2	Radiotherapy+Surgery	Died 3 months later, after lung metastasis
Treu 1986[22]	21/M	C1	Surgery only	At 25-month follow-up, patients developed metastasis – no outcome reported
Treu 1986[22]	18/M	L4–L5	Surgery only	No outcome or follow-up reported

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Conflicts of interest

There are no conflicts of interest.

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