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. Approximately 60–80% of all SS patients are young adults and adolescents, with a higher prevalence in men. 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. Histologic confirmation is required for the diagnosis of SS because differentiation from other types of spinal tumors is difficult. Chromosome translocation characteristics are found in most SSs. This involves the fusion of the SYT gene to one of the SSX genes (SSX1 and SSX2). These gene translocations allow for several histopathological variants of SS, including monophasic, biphasic, and poorly differentiated forms. 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...
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.

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 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).

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.
Schwannomas may be distinguished from SS by the characteristic features of the disease or distant metastases. Approximately 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. 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. 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.

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. 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). 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.

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.
| 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[24] | 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 6-month follow-up, no evidence of tumor recurrence. Symptoms improved |
| Kim 2013[3] | 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] | 35/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[16] | 32/M | T12–L1 | Surgery + Chemotherapy + Radiotherapy | Patients died 9 months later, after recurrence of disease |
| Koehler 2009[16] | 60/M | T7–T10 | Surgery + Radiotherapy | At 9-month follow-up, no evidence of tumor recurrence |
| Barus 2009[16] | 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[15] | 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[29] | 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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