Thoracic spinal cord compression secondary to metastatic synovial sarcoma: case report

Compressão da medula espinhal torácica por metástase secundária de sarcoma sinovial: relato de caso

Compresión de la medula espinal torácica por metástasis secundaria de sarcoma sinovial: relato de caso

Paul M. Arnold
Michael C. Park
Kathy Newell
John J. Kepes
J. Brantley Thrasher

ABSTRACT
Synovial sarcoma is an uncommon malignant soft tissue neoplasm, occurring primarily in adolescents and young adults. It is prevalent in the periarticular soft tissues near large joints of the extremities and rarely involves the trunk. Metastases are not uncommon and usually involve the lungs; metastasis to the thoracic spine is rare. We report the case of a 47-year-old man with a history of synovial sarcoma of the lower back, with subsequent metastases to the lung, penis, and perineum (all previously resected), presenting with a 3-month history of low back pain and lower extremity paresthesias. Magnetic resonance imaging (MRI) demonstrated multiple lesions involving multiple contiguous vertebral bodies, with the mass at T12 compressing the spinal cord. The patient underwent T11-T12 laminectomy, transpedicular decompression, tumor debulking, and posterior fixation and fusion. The patient died six months later due to disease progression. Although

RESUMO
O sarcoma sinovial é uma neoplasia rara dos tecidos moles que afeta adolescentes e adultos jovens. A sua maior prevalência é nas grandes articulações das extremidades e raramente acomete o tronco. As lesões metastáticas são raras e geralmente acometem os pulmões, e as metástases para a coluna torácica são raras. Relata-se o caso de um paciente de 47 anos de idade com 3 meses de história de dor lombar e que apresentava metástase de sarcoma sinovial na coluna lombar. A ressonância magnética demonstrava lesões contíguas do corpo vertebral e compressão do canal vertebral ao nível de T12. O paciente foi submetido à laminectomia de T11-T12, descompressão transpedicular, remoção tecida tumoral e artrodese e fixação posterior. O paciente foi a óbito após seis meses devido à progressão da doença. Embora a descompressão e estabilização cirúrgica do canal vertebral não sejam curativas,

RESUMEN
El sarcoma sinovial es una neoplasia rara de los tejidos blandos que afecta a adolescentes y adultos jóvenes. Su mayor prevalencia es en las grandes articulaciones de las extremidades y raramente ataca el tronco. Las lesiones metastásicas son raras y generalmente atacan los pulmones, siendo que las metástasis de columna torácica son raras. Será relatado el cuadro clínico de un paciente de 47 años de edad con tres meses de historia de dolor lumbar y presentando metástasis de sarcoma sinovial en la columna lumbar. La resonancia magnética demostraba lesiones contiguas del cuerpo vertebral y compresión del canal vertebral al nivel de T12. El paciente fue sometido a la laminectomía de T11-T12, descompresión transpedicular, remoción de tejido tumoral y artrodesis con fijación posterior. El paciente fue a óbito después de seis meses debido a la progresión de la enfermedad. Aunque la descompresión y estabilización...
not curative, decompression and stabilization of the spine are often necessary in patients who present spinal cord compression.

KEYWORDS: Sarcoma, synovial; Neoplasm metastasis; Spinal cord compression; Spinal neoplasms/secondary; Thoracic vertebrae/pathology; Laminectomy; Spinal fusion; Case reports

INTRODUCTION

Synovial sarcoma is a rare malignant neoplasm of the soft tissue that usually arises near a large joint in the extremities (especially the knee), most often in adolescents or young adults. Rarely, it is also found in the head and neck region, the chest, the abdominal wall, and the lower back. Metastasis of synovial sarcoma is not uncommon, and affects the lung more frequently. We present a case of synovial sarcoma first appearing in the lower back, with subsequent separate metastases to the lung, penis, prostate and surrounding fibroadipose tissue, then finally to the thoracic spine, with the mass at T12 compressing the spinal cord.

CASE REPORT

A 47-year-old man presented with a three-month history of low back pain radiating to the right side of the midline. He had a history of synovial sarcoma of the right lower back, originally diagnosed in 1992 at 38 years old, and staged as III a,b (T1, N0, M0, GIII) when initially resected. This sarcoma was followed by external beam radiation treatment for seven weeks. The patient presented, in 1997, metastasis in the left upper lobe of the lung and subsequently underwent thoracotomy with lobectomy, followed by a second surgery for recurrence four months later.

In 1998, the patient was found to have a penile lesion, which was biopsied and found to be consistent with synovial sarcoma. He subsequently received a 70 Gy radiation treatment. In 2000, he underwent total penectomy and perineal urethrostomy for recurrent synovial sarcoma, with an uneventful postoperative course.

The patient began complaining about low back pain seven months later, which was initially controlled by anti-inflammatory medication and morphine. The patient also complained about lower extremity paresthesias, but did not complain about any bowel or bladder incontinence. Neurological examination was normal. Magnetic resonance imaging of the thoracolumbar spine demonstrated lesions in five contiguous segments with circumferential compression at T11-T12, with the mass at T12 compressing the spinal cord. (Figure 1)

The patient underwent T11-T12 laminectomy, transpedicular decompression, tumor debulking, T7-L3 transverse process fusion with iliac crest bone graft, and pedicle screw fixation (Figure 2). The tumor, which appeared to
be encapsulated, could be seen ventral and lateral to the spinal cord. Much of the tumor had a liquid consistency and was easily suctioned out.

Pathological evaluation revealed synovial sarcoma consistent with previously recovered tissue (Figure 3), representing a highly cellular small cell malignant neoplasm with tumor cells having scant cytoplasm, occasionally demonstrating eccentric small hyperchromatic nuclei, seemingly pushed aside by eosinophilic cytoplasm lending some of the tumor cells a somewhat “rhabdoid” shape. The tumor cells also could be seen as compressing capillary blood vessels to a narrow slit; the endothelial lining cells of those vessels, however, appeared to be normal, though slightly elongated under the pressure. The close apposition of tumor cells to the small vessel walls extending to the immediate subependymal layer of the vessels is highly reminiscent of the pattern seen in hemangiopericytomas cases. This subgroup of synovial sarcomas having (at least focally) a pattern very similar to that of hemangiopericytomas, has been previously reported. In addition, in many areas of the excised tumor, numerous intraluminal tumor cells could also be observed in the blood vessels (mostly capillaries and venules).

The patient’s postoperative period went well, as he was discharged to home on the fourth postoperative day. Subsequent postoperative X-rays showed the hardware in place. He remained neurologically normal. However, the patient died six months after surgery due to progression of disease.

**DISCUSSION**

Metastatic disease involving the spinal column is a significant source of morbidity in patients with cancer, causing compression of the spinal cord, cauda equina, and nerve roots. Epidural spinal cord compression (ESCC) develops in 5% of all cancer patients, and most often occurs in the thoracic spine (60%), followed by the lumbo-sacral spine (30%) and cervical spine (10%). Vertebral body metastases may cause local, radicular, or axial pain, in addition to a variety of neurologic deficits from mild radicular weakness to paraparesis. In autopsy studies of cancer patients, metastatic deposits in the spine have been observed in up to 90% of the patients, and symptomatic disease will develop in approximately 30% of patients. The thoracic region of the spine is involved in approximately 70% of cases.

Synovial sarcomas are uncommon malignant soft tissue neoplasms, representing less than 1% of all cancers and accounting for an estimated 5 to 10% of all soft tissue sarcomas. It is the fourth most common soft-tissue sarcoma in adults after malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma. Synovial sarcoma occurs primarily in adolescents and young adults between 15 and 40 years of age, affecting males more frequently than females. It usually arises in the periarticular regions near large joints, with 75 to 90% of cases involving the extremities, especially the lower extremity. A wide variety of other anatomic primary sites have been reported, including heart, lungs, small intestine and peripheral nerves. Only 5 to 15% of cases involve the head and neck region, the trunk (including the abdominal wall and the retroperitoneum) and the lower back.

More than 95% of synovial sarcomas have a characteristic chromosomal translocation with a resultant fusion gene of SYT and one of 3 SSX genes. Despite the understanding of the molecular biology underlying synovial sarcoma, its cells of origin remain unknown. Synovial sarcoma is histogenetically unrelated to synovium; the term “synovial” sarcoma is
due to the synovial differentiation of the tumor which is believed to originate from multipotential mesenchymal cells; thus, synovial sarcoma is a mesenchymal tumor of unknown histogenesis that does not arise in synovial membranes.

Synovial sarcoma is characterized by epithelial-like and spindle cell components arranged in a biphasic or monophasic pattern; there is also a poorly differentiated (PD) form. Synovial sarcomas are microscopically divided into four histological subtypes: biphasic (the most predominant type); monophasic fibrous; monophasic epithelial; and poorly differentiated; the biphasic type contains a glandular component in addition to the monophasic spindle cell.

Synovial sarcoma remains one of the most aggressive soft-tissue sarcomas, despite improvements in staging, surgical technique, and adjuvant therapies. According to Baptista et al., most pathologists consider synovial sarcoma as a high-grade soft-tissue sarcoma, though they consider the biologic behavior, in some cases, to be more benign than others. According to de Silva, development of local recurrence carries an increased risk for development of metastases and tumor-related death. Metastatic recurrence is determined by three independent factors: tumor grade, tumor size (>5 cm), and histopathology. Disease-free survival (freedom from local, lymph node, and metastatic recurrence) correlates with tumor grade, tumor size, microscopic resection margin, presentation, tumor site, patient age, and histologic subtype. Disease-specific survival (reflecting the effects of disease recurrence and the outcome of salvage therapy) correlates with tumor grade, tumor size, tumor site, histopathology, patient age, and resection margins. Tumor grade, size, and histopathology exerted their effect largely through their influence on metastatic recurrence, while tumor site, patient age, and resection margins exerted their effect through predisposing to local recurrence.

Although metastases of synovial sarcomas are not uncommon, with an involvement of more than half of all cases, the organ that is most frequently affected by synovial sarcoma metastases is the lung, followed by the liver and skeletal system. Synovial sarcoma with metastasis to the thoracic spine is unusually rare. In a series of 59 patients, Bilsky noted three spindle cell synovial sarcoma metastases to the spine, but did not specify a level. Meriinsky et al., in a series of 19 cases, reported one patient with a T7-8 synovial sarcoma metastasis. This patient was treated with radiation therapy. This patient’s pain resolved but not her motor and sensory disturbances. Suh et al. reported an epidural synovial sarcoma metastasis at L4-5. This patient underwent an L4-5 hemilaminectomy with tumor resection; the tumor was noted to be extending through the right L4 foramen and into the soft tissues. The patient had symptomatic improvement at the fifth month of follow-up. Otsuka et al. reported an L5 metastasis that was also surgically resected. Signorini et al. resected a T2 synovial sarcoma following paraplegia. Following a subtotal resection, the patient died three months later. Sakellaridis resected a hemangiopericytoma-like synovial sarcoma from the lumbar spine of a 36-year-old woman; she died eighteen months later due to multiple recurrences.

There were also rare reports of intradural synovial sarcoma metastasis. Greene et al. reported an 11-year-old girl who had multiple intradural extramedullary metastases, most notably at L2-4. This was the initial manifestation of her disease. The patient had this tumor resected but died 14 months after diagnosis. Scollato et al. reported the case of a 59-year-old man with intramedullary metastasis at C3-5.

Clinical management of spine tumors, including metastatic disease, has undergone drastic evolution in the past few years. Treatment of spinal column metastases is seldom curative, thus the goal in most cases is palliation. Median survival after ESCC diagnosis is four to six months.

Advances in magnetic resonance imaging, sophisticated instrumentation for spinal stabilization, and an increased understanding of the biomechanics of the spine have led to increased treatment options for spine metastases. The advent of spinal instrumentation, as well as transpedicular or ventral decompression, has allowed favorable outcomes in treating and possibly reversing significant neurological deficit in comparison to previous treatment methods of radiation, decompressive laminectomy without stabilization, or combined radiation and laminectomy. Several authors reported rates of neurological improvement nearing 70% compared to traditional therapies, after the refinement of anterior spinal approaches and the introduction of more reliable segmental spinal stabilization systems for both anterior and posterior reconstructions. These same authors also demonstrated that superior rates of pain relief could be achieved when anterior or posterior stabilization was combined with neural decompression to eliminate tumor-related axial spine instability. Patchell et al. has shown that surgery followed by radiation yields better neurologic outcomes than radiation alone.

Synovial sarcoma is an uncommon malignant soft tissue tumor, which rarely metastasizes to the spine. We report an unusual patient who presented multiple thoracic spine lesions, and was successfully treated with decompression, fusion, and fixation.

ACKNOWLEDGEMENTS

The authors thank Karen K. Anderson for her editorial assistance in the preparation of this manuscript.
REFERENCES


Correspondence:
Paul M. Arnold
Spinal Cord Injury Center of the Department of Neurosurgery of University of Kansas Medical Center
3901 Rainbow Boulevard, MS 3021
Kansas City KS 66160
Tel.: (913) 588-7587
Fax: (913) 588-7596
E-mail: parnold@kumc.edu