Case Report

Synovial sarcoma of the thoracic spine

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Received 18 May 2009; revised 29 July 2009; accepted 21 August 2009

Abstract

BACKGROUND CONTEXT: Synovial sarcoma is an uncommon malignant neoplasm occurring chiefly in young adults. It often presents as a solid well-circumscribed soft-tissue mass in the extremities of young adults. Despite its proximity to joints, it has been well established that the tumor cells do not display features of synovial differentiation but instead appear to have a primitive epithelial phenotype. There is no report of a lower thoracic paravertebral synovial sarcoma in an adult male.

PURPOSE: To describe our management in a patient with a synovial sarcoma of the thoracic spine and to review previously published cases.

STUDY DESIGN: Case report.

METHODS: A 60-year-old man presented with a 5-month history of right upper quadrant abdominal pain radiating to his back in a band-like fashion; shortness of breath on exertion; and increasing pain when standing, sitting, or walking. Magnetic resonance imaging (MRI) demonstrated a large right-sided paraspinal mass sitting on the eighth and ninth ribs, pressing on the T9 vertebra and abutting the T7 and T8 vertebral level exhibiting “Triple Intensity.” Plain films demonstrated a right-sided paraspinal mass extending from the T7–T8 level to T10. Bone scintigraphy showed increased uptake on the right thoracic spine at T7–T8 to T10. Computed tomography (CT) imaging revealed a right paraspinal mass with lytic changes in the T9 vertebral bodies. A right-sided thoracotomy was performed, and the patient underwent subsequent radiation therapy. Absence of the tumor was shown by an MRI scan after the operation.

RESULTS: Complete resolution of the patient’s complaints was achieved. The diagnosis is supported by plain radiographs, bone scintigraphy, magnetic resonance and CT imaging studies, and histologic and immunohistochemical evidence.

CONCLUSIONS: Synovial sarcomas are rarely present in the paravertebral region of the thoracic spine. A careful radiographic study of the tumor permitted early preliminary diagnosis, confirmed upon histopathologic analysis. Despite lytic changes, removal of a periosteal layer permitted sparing of the vertebral bodies.

Keywords: Synovial sarcoma; Paravertebral; Thoracic spine; Spine; Magnetic resonance imaging

Introduction

Synovial sarcoma is an uncommon malignant neoplasm occurring chiefly in young adults. It often presents as a solid well-circumscribed soft-tissue mass in the extremities of young adults. Despite its proximity to joints, it has been well established that the tumor cells do not display features of synovial differentiation but instead appear to have a primitive epithelial phenotype [1–5]. In this report, we describe a pathologically proven monophasic type synovial sarcoma presenting as a paravertebral mass of thoracic vertebrae T7–T10. To our knowledge, there is no report of a lower thoracic paravertebral synovial sarcoma in an adult male. Thus, we report the case of a 60-year-old man with a monophasic synovial sarcoma at T7–T10 who had plain radiography, bone scintigraphy, magnetic resonance imaging (MRI), and computed tomography (CT) as well as intraoperative and histopathologic correlation. The patient was...
informed that data concerning the case would be submitted for publication and consented.

Case report

A 60-year-old man presented with a 5-month history of a gradual onset of right upper quadrant abdominal pain radiating to his back in a band-like fashion; shortness of breath on exertion; and increasing pain when standing, sitting, or walking. On examination, he was slightly tender to palpation in the right upper quadrant; exhibited full strength bilaterally in upper and lower extremities; no abnormal reflexes, no clonus, normal plantar reflex, full range of motion of cervical, thoracic, and lumbar spine; full motion of thorax, as well as lateral bending and lateral rotation of the thoracic spine.

Magnetic resonance images 2 months after the onset of pain demonstrated a large right-sided paraspinal mass sitting on the eighth and ninth ribs, pressing on the T9 vertebrae and abutting the T7 and T8 vertebral level. The ninth thoracic vertebrae showed slight bony erosion where the mass was sitting, but there was no evidence of any bony instability or neural compromise (Fig. 1). Calcifications were identified within the mass accompanied by increased signal intensity at the center of the mass (Fig. 2). Plain films 3 months after the onset of symptoms demonstrated a radiopaque right-sided paraspinal mass extending from the T7–T8 level to T10 (not shown). Bone scintigraphy also performed 3 months after symptoms showed increased uptake of radiolabeled diphosphonates on the posterior view of the right thoracic spine at the levels of T7–T8 to T10 (not shown). Computed tomography images of the thoracic spine without contrast 3 months after the onset of symptoms were obtained with overlapping reconstructions. A 4.5×6.8×5.8-cm, right-sided, paraspinous mass extending from the T7–T8 level through the mid aspect of T10 was identified (not shown). Lytic changes were identified by MRI and not confirmed by CT. There was no evidence of foraminal or intracanalicular tumor extension. The lesion had a broad base against the right T9 rib and invaginated into the intercostal space.

At surgery, a right-sided thoracotomy was performed. The mass was identified at T8–T9 without significant adhesion to the right lung. After complete exposure of the mass, an incisional biopsy was performed and sent for frozen pathology. On confirmation that the mass was a malignant neoplasm, a wide resection was performed with care taken to obtain negative margins. All feeding vascular and neural supply were ligated including the right T9 nerve root. Intraoperatively, the decision was made not to perform any bony resection of the vertebral bodies because a thick periosteal layer was removed with the mass.

Gross pathology demonstrated a nodular mass of friable pink-tan soft tissue partially surfaced by pink-tan skin. There was no hematoma present. Microscopically, the mass consisted of spindle cells with variable mitotic rates and a prominent hemangiopericytomatous vascular background (Fig. 3, left). Immunohistochemical studies showed that the tumor was strongly positive for vimentin and bcl-2 (Fig. 3, right) and ethidium monoazide. The tumor was negative for actin, desmin, S-100, CD34, CK7, CMA 5.2, calretinin, and p63. The tumor was identified as a monophasic synovial sarcoma. Cytogenetic analysis was deemed unnecessary by the pathologist due to an obvious diagnosis based on histology and immunohistochemical studies. The resection margins were negative for the presence of tumor cells. Due to the adherence of the tumor to the vertebral bodies, radiation therapy was recommended. The patient agreed and underwent such treatment. Follow-up of MRIs 9 months after surgery and radiation therapy demonstrated no recurrence (not shown).

Discussion

Synovial sarcoma is an uncommon malignant neoplasm occurring chiefly in the extremities of young adults, with a median age of 35 years [1–9]. This is unusual, as most other soft-tissue sarcomas typically appear in the 50s [10,11]. Incidence ranges from 6% to 10% of all soft-tissue sarcomas occurring in adults [1,12]. Synovial sarcomas are the third most common extremity soft-tissue sarcoma,
accounting for 12% to 15% of sarcomas reported [10,11,13–17]. They occur equally in men and women and have no identifiable genetic or etiologic predisposing factor [6–9,13,18,19].

Synovial sarcomas are generally a moderately aggressive lesion, with a 5-year survival rate of 60% to 75% [6–9,20,21]. In a study performed by Lewis et al. [7], a 5-year local recurrence rate was reported as 12%, much lower than contemporary studies that range from 18% to 30% [8,9]. The distant recurrence rate has been reported as 39%, with most occurring in the lung [7]. Importantly, the 10-year survival rate has been reported to be 34%, indicating that most deaths occur between 5 and 10 years, thus long-term follow-up is vital in patients with synovial sarcomas [6].

Although most synovial sarcomas are encountered in the extremities, 5% to 15% of all synovial sarcomas have been reported in the body axis [1,12,22]. In the body axis, they have been reported as arising from the trunk (~8%), retroperitoneum (~7%), and head and neck (~5%) [9,13,20–24]. Reports of synovial sarcomas located in the paravertebral space have been extremely rare. Gualtieri and Calderoni [25], Treu et al. [26], Wu et al. [27], and Suh et al. [28] reported the presence of lumbar paravertebral synovial sarcomas. Suster and Moran [29] reported two monophasic synovial sarcomas located in the posterior mediastinum that were paraspinal. The specific locations were not provided. There are only two incidences in the literature of a thoracic paravertebral synovial sarcoma: Morrison et al. [5] reported the presence of such a neoplasm extending from C7 to T3, and Signorini et al. [3] reported the presence at the level of T2. To our knowledge, there is no report of a lower thoracic paravertebral synovial sarcoma.

Despite their name and often being proximal to joints, synovial sarcomas hardly ever arise within synovial joints and do not display features of synovial differentiation. The name stems from early literature, which frequently reported para-articular locations and microscopic resemblance to developing synovium [18]. Instead, synovial sarcomas have a primitive epithelial phenotype based on ultrastructural studies and immunohistochemistry [1,30–33]. There are three encountered histologic varieties depending on the combination of spindle and epithelial tumor cells, the classic biphasic synovial sarcoma, the monophasic type,
and newly identified poorly differentiated type [18]. The biphasic variant is characterized by epithelial cells mixed with spindle cells; the monophasic is exclusively composed of spindle cells; and the poorly differentiated are composed of uniform, densely packed, small ovoid blue cells that resemble other round blue cell tumors [5,18,28,34]. Pure poorly differentiated synovial sarcomas are quite rare, but up to 20% of biphasic and monophasic may contain areas with poor differentiation [32,35].

Monophasic synovial sarcomas, such as that identified in this report, are typically composed exclusively of spindle cells. Only extremely rare cases of purely epithelial synovial sarcomas have been reported. The spindle cells are typically very uniform in appearance and exhibit very little cytoplasm, darkly staining nuclei, and indistinct cell borders. The cells are arranged in sheets or short fascicles resembling fibrosarcoma. Compact areas frequently alternate with areas of myxoid change or more loosely packed lighter staining areas. The background often contains a large number of blood vessels with a hemangiopericytomatous appearance, as seen in this case. Occasionally, the background may contain abundant collagen or exhibit calcification or ossification. The biphasic variant comprised a spindle component identical to that of the monophasic subtype but additionally contains a population of larger paler epithelial cells with distinct cytoplasmic borders and more vesicular nuclei. Poorly differentiated synovial sarcomas comprised cells with more marked nuclear atypia, pleomorphism, necrosis, and mitotic activity. Although the nuclear features present in this case are somewhat more atypical than might be expected in a monophasic synovial sarcoma, the totality of findings did not meet criteria for the poorly differentiated subtype.

Synovial sarcomas are typically present as a slow-growing mass, simulating a benign neoplasm [36]. Additionally, these lesions are frequently painful, illustrated by this case in which the patient reported gradual onset of upper right quadrant pain with radiation [37]. The size of the synovial sarcoma resected in this case is also in accord with the literature, which reports that 85% synovial sarcomas tend to be large and >5 cm [36].

On plain films, synovial sarcomas are usually well-defined or a lobulated soft-tissue mass with up to one-third of cases demonstrating calcification [18,37]. Because the growth of the tumor is slow, it is also possible to identify superficial pressure erosions, periosteal reaction, and osteoporosis on plain radiographs. Frank osseous invasion can also occur [38]. In this case, the plain films were unremarkable for calcification or periosteal reaction.

Computed tomography imaging is very helpful in identifying the more subtle soft-tissue calcifications and local bony changes that may be present. Computed tomography also provides an accurate relationship of a soft-tissue tumor to the adjacent bones and can therefore be useful in assessing the relationship of synovial sarcomas to bone in complex areas such as the spine. Tateishi et al. [39] identified tumor calcification with CT in 27% of patients. In the CT images we report, lytic changes were identified. Additionally, we were able to determine that there was no evidence of vertebral body invasion or foraminal or intracanalicular tumor extension. There was no evidence of calcification.

Magnetic resonance imaging is considered the modality of choice for detection and staging of soft-tissue tumors. Synovial sarcomas usually appear as well-defined, nonspecific, heterogenous soft-tissue masses and may have a multilocular appearance with septation [40,41]. About 40% of lesions demonstrate high signal on both T1- and T2-weighted images, which is consistent with hemorrhage. This allows the differentiation of synovial sarcomas from other soft-tissue tumors. In this case, the tumor exhibited T1 high signal but not T2 high signal. The incidence of intratumoral hemorrhage was found to be 73% and is generally uncommon in most soft-tissue sarcomas other than synovial sarcoma [42,43]. Additionally, synovial sarcomas can be differentiated from other soft-tissue tumors by the so-called “triple signal intensity”: hyperintensity, isointensity, and hypointensity relative to fat. Approximately 35% cases of synovial sarcomas demonstrate the triple signal intensity [36,42]. The tumor in this report demonstrated the signature triple signal intensity, which was definitive for the diagnosis. Unfortunately, there is no notable difference between the MRI characteristics of monophasic and biphasic pathologic subtypes. The tumor in this case exhibited calcifications on MRI that were not confirmed by CT. A hematoma was ruled out during tumor removal at surgery.

In the thoracic location, the histologic differential diagnosis of monophasic synovial sarcoma includes primarily sarcomatoid malignant mesothelioma, solitary fibrous tumor, or sarcomatoid carcinoma [44]. Also to be considered in the differential diagnosis, although more unlikely on the basis of the anatomical location, is a fibrosarcoma, leiomyosarcoma, hemangiopericytoma, or a malignant peripheral nerve sheath tumor. In most cases, these tumors can be unambiguously identified only by immunohistochemical analysis and molecular studies.

The histology of biphasic and sarcomatoid variants of malignant mesothelioma overlaps greatly with synovial sarcoma. The clinical distribution of disease is important, as malignant mesotheliomas typically occur in an older age group than synovial sarcoma and characteristically present with diffuse pleural involvement, as opposed to a localized mass. Histologically, sarcomatoid malignant mesotheliomas generally exhibit much greater cellular pleomorphism and contain more abundant eosinophilic cytoplasm. Both are positive for epithelial markers such as cytokeratin, but sarcomatoid mesothelioma typically exhibits diffuse strong staining for keratin in contrast to the patchy focal staining typical of synovial sarcoma. Bel-2 is typically strongly positive in synovial sarcoma but is usually negative in mesothelioma. Synovial sarcomas are characterized by t(X;18)(p11.2;q11.2) translocation. Mesothelioma, in
contrast, lacks a distinctive unique translocation, but del (1p), del (3p), and del (22) have been reported [29,44].

Solitary fibrous tumor can be morphologically indistinguishable from monophasic synovial sarcoma, yet they are easily differentiated on immunohistochemical staining. Solitary fibrous tumors usually stain positive for CD34 and will not express keratin or epithium monoaude, whereas synovial sarcoma is negative for CD34 [29,44].

Histologically, sarcomatoid carcinoma shows infiltrative growth, pleomorphism, and a higher degree of nuclear atypia than synovial sarcoma. On immunohistochemistry, they are bcl-2 and CD99 negative. Additionally, they have del (3)(p14p23), making them cytogenetically distinct [29,44].

Based on the reported histologic, immunohistochemical, and clinical features of this case and their consistency, the diagnosis of a monophasic synovial sarcoma was made. Because the histologic and immunohistochemical features were consistent with a synovial sarcoma, molecular testing for the characteristic t(X;18)(p11.2;q11.2) chromosomal translocation was determined to be unnecessary. More than 90% of synovial sarcoma cases contain the translocation [45–47]. The decision that molecular testing is unnecessary is supported in the literature, which reports molecular testing as generally noncontributory in cases of histologic and immunohistochemical consistency [48].

In conclusion, we report the presence of a paravertebral thoracic monophasic type synovial sarcoma at T7–T10. The diagnosis is supported by plain radiographs, bone scintigraphy, magnetic resonance and CT imaging studies, and histologic and immunohistochemical evidence.

References


