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Spindle Cell Sarcoma of the Paraspinal Musculature with Late Pulmonary Metastases

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Male, 58
Final Diagnosis: Spindle cell sarcoma
Symptoms: Chest wall tenderness • painful paraspinal mass • shortness of breath
Medication: —
Clinical Procedure: Surgical excision with chemotherapy and radiation
Specialty: Oncology

Objective: Rare disease

Background: Sarcomas account for less than 1% of all cancers. Spindle cell sarcomas are a rare form of soft tissue sarcomas classified as undifferentiated/unclassified based on their histomorphology. These tumors have a propensity for local recurrence and distant metastases are frequently found in the lungs. The risk for metastases increases with higher-grade malignancy and the size of the primary tumor. The often-painless nature of these tumors results in a delay in diagnosis, and physicians frequently overlook sarcomas in their differentials due to their rarity, complicating the disease process.

Case Report: This article reports a case of a spindle cell sarcoma in the left paraspinal musculature in a 58-year-old Caucasian male; the tumor was excised in large pieces. There was an initial benign course, during which time the patient was undergoing regular imaging studies to evaluate for recurrence. Eight years later, the tumor metastasized to the lungs with an initial presentation of shortness of breath and pleural effusion on imaging.

Conclusions: Sarcomas are very rare soft tissue neoplasms, but they should not be overlooked in a physician's differentials, especially when evaluating an enlarging mass. Recommended treatment of choice is complete surgical excision with adequate resection margins of at least 1 cm or greater to a fascial barrier. Recurrence and late pulmonary metastases are common and metastatectomy is the recommended treatment choice if metastases are present.

MeSH Keywords: Metastasectomy • Neoplasm Metastasis • Sarcoma • Soft Tissue Neoplasms

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Background

Spindle Cell Sarcomas are rare cancers that form in many different types of connective tissues. They are a subtype of undifferentiated/unclassified soft tissue tumors that lack a specific line of differentiation. It is known that the undifferentiated/unclassified group accounts for <1% of all malignant cancers [1], but there is a significant lack of epidemiological data for spindle cell sarcomas specifically. Primary tumors are more common in the bone, skin, and soft tissue as well as the respiratory system [2]. These tumors have a propensity to metastasize to the lungs, and approximately 25% of sarcoma patients will develop distant metastases after successful treatment of their primary tumor [3]. Risk factors of this disease include genetic predisposition and an underlying inflammatory process, but little is known otherwise [4]. We present this case of a low-grade spindle cell sarcoma of the paraspinal musculature with late pulmonary metastases due to the rarity of the disease, the unique presentation of the patient in a primary care setting, and the deviation from the standard of care during treatment that resulted in future complications.

Case Report

A 58-year-old Caucasian male presented to his primary care physician with acute onset of back pain. The patient's past medical history was significant for hypertension treated with valsartan and hydrochlorothiazide. The patient used chewing tobacco and drank alcohol socially. On physical examination, there was a palpable mobile mass that measured 5–6 cm in the left paraspinal region. The patient was given a trial of Mobic 7.5 mg daily, but his pain symptoms were not relieved. Magnetic resonance imaging (MRI) without contrast showed a spindle-shaped mass that measured 6.9×3.5×5.4 cm in the left paraspinal muscle (Figures 1, 2). The mass was located from T9-T11 but did not extend into the neural foramina or central canal of the spinal cord. After this finding, an MRI with contrast was done showing the same results and no other masses were visualized.

The patient was referred to a local neurosurgeon for assessment and resection of the mass 4 months after initial presentation. The patient experienced no complications from the procedure and the mass was removed in large pieces, due to inadequate tools and malfunctions during surgery. According to the operative report, there was a piece of tumor that was attached to either an intercostal nerve or periosteum that the surgeon was not comfortable removing. At that time, the surgeon believed the tumor to be benign and the remaining portion was cauterized using bipolar electrocautery. The surgeon was comfortable leaving the remaining part of tumor behind. The pathology report indicated that sections of the mass showed a spindle cell proliferation with focally organoid architecture. Immunohistochemical staining was positive for vimentin and smooth muscle actin, but negative for S-100, desmin, and epithelial membrane antigen (EMA). Trichrome stain showed a mixture of fibrous tissue and cells with eosinophilic characteristics of the cytoplasm. Ki-67, a specific cellular marker for proliferation, was tested for and shown to be positive with activity around 10%. Many mitotic figures, up to 9 mitoses per high power field (HPF), were seen with irregular margins. Preoperative diagnosis was suspicious for a soft tissue tumor. Postoperative pathology analyzed the tumor as aforementioned and staged the mass using the American Joint Commission on Cancer (AJCC) staging guidelines as a T2bN0M0 grade II paraspinal spindle cell sarcoma.

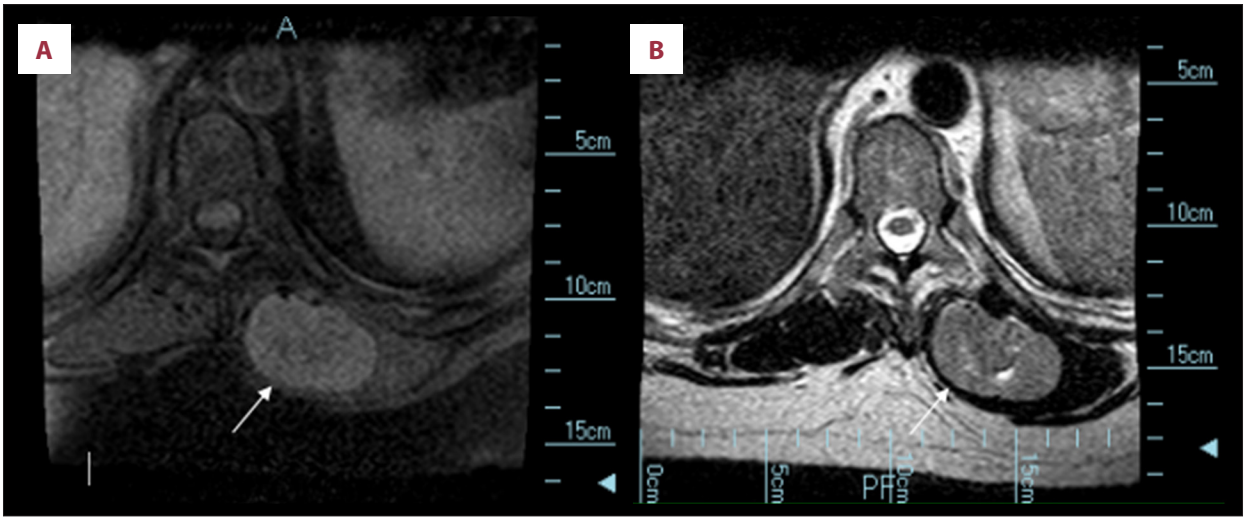

Figure 1. Transverse T2-weighted magnetic resonance imaging with (**A**) and without (**B**) gadolinium contrast showing the primary tumor in the left paraspinal musculature (arrow).

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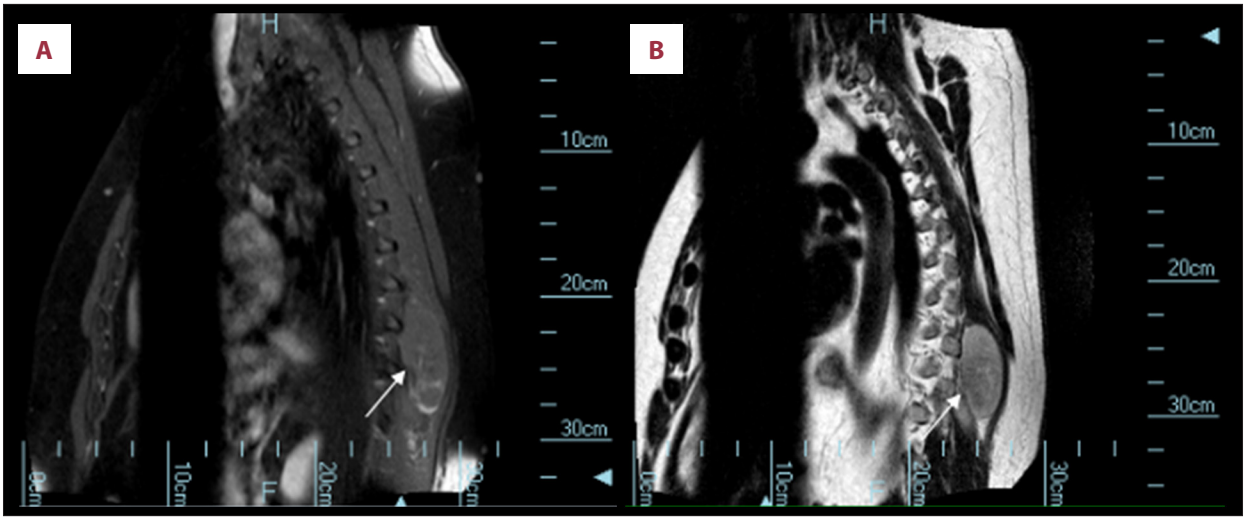

Figure 2. Lateral T1-weighted magnetic resonance imaging with fat suppression with (**A**) and without (**B**) gadolinium contrast showing the primary tumor in the left paraspinal musculature (arrow).

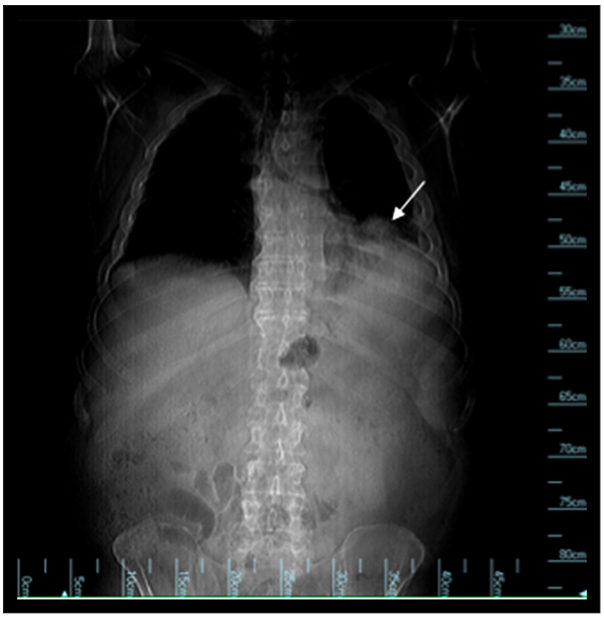

Figure 3. Anterior-posterior chest radiograph showing left lung metastasis (arrow) with large left pleural effusion.

Post-operative MRI showed complete resolution of the mass with no evidence of persistent tumor. Two months after resection of the mass, the patient was seen by a radiation oncologist to be evaluated for radiation therapy. The patient was provided with the risks and benefits to radiation treatment and consented to therapy. The oncologist decided on a total dose of 60 Gy in 30 fractions over 6 weeks. After radiation treatment, the patient underwent regular imaging every 6 months to evaluate for local recurrence or late metastases.

One year after initial presentation, the patient was seen by his primary care physician for follow-up after radiation treatment. Nodules of the thyroid were felt on physical examination and appropriate testing was ordered. Ultrasound showed multinodular goiter with hypoechoic solid appearing nodules and 2 complex cysts. The diagnosis on further work-up was found to be multifocal papillary carcinoma of the thyroid due to radiation treatment. The patient then underwent total thyroidectomy and central lymph node dissection with no complications.

One year after his initial presentation, a computed tomography (CT) of the chest showed small patchy infiltrates of the left lung with pleural effusion, which were thought to be complications from radiation treatment and no further action was taken. Follow-up imaging of the chest every 6 months was stable until 8 years after initial presentation. Chest radiograph showed an 8.8×9.4 cm left lower lung mass with extension into the chest wall and large left pleural effusion (Figure 3). The patient experienced increasing shortness of breath and chest wall discomfort at this time and was treated for what was thought to be pneumonia. The patient underwent thoracentesis to treat the pleural effusion which revealed malignant cells. The patient was to be evaluated by a university oncologist to begin chemotherapy, but the physician wanted to wait until the patient was evaluated by a local cardiothoracic surgeon first.

Repeat imaging of the mass 4 months after discovering the initial lung mass (currently the time of reporting) showed it had grown to 20×13×19 cm and a biopsy diagnosed the recurrence as a solitary fibrous tumor with spindle cell findings consistent with spindle cell sarcoma. The patient is currently undergoing chemotherapy and radiation for treatment of the lung mass.

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Discussion

Spindle cell sarcoma is a rare subtype of undifferentiated/unclassified soft tissue sarcomas that lack a specific line of differentiation. This group of sarcomas are described by their histomorphology and include the pleomorphic, round cell, and spindle cell variants. While undifferentiated/unclassified sarcomas account for <1% of all malignant cancers [1], there is a significant lack of epidemiological studies for spindle cell sarcomas, specifically. Due to their rarity, only a few sporadic case reports and retrospective case series have been published in the medical literature. These studies, however, have been largely insufficiently powerful in determining survival statistics due to their small sample sizes. More recent larger retrospective analyses have demonstrated a peak incidence in the seventh decade of life with an equal prevalence in males and females [5].

Though primary spindle cell sarcoma arises mostly in the respiratory system, cases have been noted to arise in similar locations to other undifferentiated/unclassified sarcomas, particularly the head and neck, extremities, and retroperitoneum [2]. It is also known that soft tissue sarcomas primarily metastasize to the respiratory system and late pulmonary metastases are common. Overall, approximately 25% of sarcoma patients will develop distant metastases after successful treatment of their primary tumor, and 70–80% of cases will metastasize to the lungs [3]. When the primary tumor is >5 cm, deep to the fascia, and intermediate- or high-grade, the incidence of distant metastases increases to 40–50% [6,7].

Retrospective studies have determined that adequate resection margins and avoiding compromise of the tumor's pseudocapsule, if present, are the most important prognostic factors for long-term survival [8,9]. When these tumors are excised, it is recommended that they be removed in whole and with adequate resection margins. Although studies have not been conducted to determine the ideal margin of normal tissue to excise with this tumor, it is generally accepted that a minimum of 1 cm of healthy tissue or dissection up to a fascial barrier constitutes an adequate resection margin [10]. If a pseudocapsule is present, it is important to avoid compromise because gross or microscopic residual tumor is associated with higher local failure, even if radiation therapy is used. Importantly, patients with distant metastases or local recurrence have significantly higher mortality rates [3]. If close or positive margins are noted after resection of the tumor, adjuvant radiation therapy is recommended. Preoperative radiation therapy might also be beneficial to decrease the size of the tumor prior to resection and will also allow the use of smaller radiation fields if post-resection radiation is required [11].

Because sarcomas are extremely rare, delay in diagnosis is common. They often bypass a physician's differentials and patients frequently do not seek prompt care due to the often-painless nature of the mass. The United Kingdom Department of Health has published criteria for the urgent referral of any patient with a soft tissue lesion: mass >5 cm (golf ball size), painful lump, lump that is increasing in size, lump of any size that is deep to the muscle fascia, or any lump that recurs after excision [12]. It has also been noted in the literature that masses are often excised inappropriately before a biopsy can confirm the diagnosis because physicians assume benignity. Data underscore the importance of transferring any patient with an unidentified soft tissue mass to a center that specializes in treating sarcomas so they may undergo appropriate initial resection [13]. Tumors that are inappropriately biopsied or have an inappropriate attempt at resection can result in disturbance of the tumor and increased risk of metastasis or incomplete excision.

MRI is the recommended initial imaging for all soft tissue masses of the extremities, trunk, and head and neck while CT is the recommended imaging choice for retroperitoneal and visceral masses [14]. It is also recommended that all patients diagnosed with a sarcoma undergo chest imaging with CT for evaluation of pulmonary metastases at the time of diagnosis and regularly after excision of the primary lesion. Positron emission tomography (PET) is not routinely recommended as a component of the initial staging workup of soft tissue sarcoma, however, several studies have reported that PET and integrated PET/CT using fluorodeoxyglucose (FDG) can distinguish between benign soft tissue tumors and sarcomas. These particular scans have the greatest sensitivity for high-grade sarcomas as the ability to differentiate between benign soft tissue tumors from low- or intermediate-grade sarcomas is limited [15–17]. If biopsy is performed, core needle biopsy is preferred due to its low incidence of complications and high diagnostic accuracy [18].

In patients who develop pulmonary metastatic disease, aggressive surgical resection of the metastases offers a chance for extended disease-free survival that is not possible with systemic chemotherapy. Though surgical resection is not appropriate in some patients, and is impossible in others, overall 5-year and 10-year survival in those who are able to undergo complete metastatectomy are approximately 36% and 26% respectively [19]. Following treatment, it is recommended that patients undergo regular radiographic surveillance with CT every 4 months. If pulmonary recurrence is noted and indications for metastatectomy are still met, repeat surgery should be offered as an alternative to systemic therapy or other local ablative therapy.

Conclusions

Sarcomas are very rare soft tissue cancers that have a propensity to metastasize to the lungs. When a patient presents with

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an enlarging mass, especially if located in the head, neck, trunk, extremities, or retroperitoneum, clinicians should urgently refer the patient to a center that specializes in the treatment of soft tissue tumors for appropriate biopsy and resection [12]. The treatment of choice for soft tissue sarcomas is complete en bloc surgical excision with adequate resection margins of at least 1 cm. In the patient case reported here, removing the tumor in large pieces and having microscopically positive margins post-resection likely contributed to this patient's late pulmonary metastasis. Delay in evaluation and treatment should be avoided in aggressive sarcomas as the risk of metastasis increases almost linearly as the tumor enlarges [20].

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Statement

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Conflicts of interest

None.

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