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A case of spindle cell sarcoma of the groin

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Abstract

The presence of a mass in the groin region is concerning due to its anatomical location and the wide range of possible differential diagnoses. Soft tissue sarcomas are rare tumors that should be suspected when a mass exceeds 5 cm and continues to grow independent of pain. We report a case of spindle cell sarcoma, a subtype of soft tissue sarcoma, located in the groin. A 77-year-old Caucasian male with a medical history malignant prostate neoplasm post-prostatectomy, hypertension, and chronic back pain, was referred for a painless, enlarging mass in the left upper inner thigh, just below the inguinal crease. An ultrasound revealed a 4.2 cm hypoechoic mass in the left inguinal region, with no significant lymph node enlargement. The patient underwent surgical excision of the mass under general anesthesia. Pathological analysis, including immunohistochemistry and molecular testing, confirmed spindle cell sarcoma with focal smooth muscle differentiation, grade 3. No fusion transcripts were detected. The patient is currently undergoing chemotherapy, with consideration for radiation therapy. This case highlights the rarity of spindle cell sarcoma in the groin and emphasizes the importance of early diagnosis and multidisciplinary management to improve patient outcomes.

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Introduction

Sarcomas are defined as a heterogeneous group of rare tumors that arise predominantly in the embryonic mesoderm [1] and originate from various soft tissues, such as muscles, adipose tissue, nerves, and blood vessels [2]. Soft tissue sarcomas are suspected in any patient who presents with a soft tissue mass larger than 5 cm that is growing or subfascial, independent of pain level [3]. When localized in the groin area, soft tissue sarcomas are considered a distinct subset due to their peculiar anatomical location and unique diagnostic and management challenges [4]. Soft tissue sarcomas of the groin may present a difficult problem because of misdiagnosis as groin hernias and its proximity to major neurovascular structures [4]. These tumors are further complicated by the fact that they represent at least 80 potentially malignant histological types and subtypes, 50% of which are fully malignant and commonly metastasize and 50% of which are locally aggressive with some but limited metastasis potential [5]. We present a case report of a patient with spindle cell sarcoma, a subtype of soft tissue sarcoma, in the groin region.

Report of case

A 77-year-old Caucasian male patient with a medical history of malignant neoplasm of the prostate status post prostatectomy in 2018, hypertension, and chronic back pain, was seen by urology for evaluation of a positron emission tomography-computed tomography scan showing a 1.5 positron emission tomography avid soft tissue nodule in the right hemipelvis concerning for nodal metastasis with otherwise no evidence of detectable disease in July 2024. During his evaluation with urology, he was found to have a raised, non-painful, enlarging mass in the left upper inner thigh, just below the inguinal crease. He was then referred to general surgery after an ultrasound of the bilateral groin and left upper inner thigh which revealed a hypoechoic mass present in the left inguinal region measuring 4.2 cm in greatest diameter with nonenlarged inguinal lymph nodes noted bilaterally. In August 2024, the patient had an excision of the large left groin/upper medial thigh mass under general anesthesia. During this procedure, the mass was noted to be firm and adherent to soft tissue when it was dissected off the underlying medial thigh musculature. Additionally, the mass did not appear to arise from the muscle tissue itself.

Following surgery, the excisional biopsy of tan-pink skin and underlying soft tissue measuring 6×4.6×4 cm in gross dimension was sent to pathology. The microscopic tissue specimen revealed hypercellular proliferation of cytologically malignant spindle cells arranged in sheets and broad fascicles with numerous mitoses showing >30 per 5 high power field which included atypical mitotic figures. Additionally, patchy necrosis was present. The immunohistochemical stains were patchy positive for Smooth Muscle Actin (SMA), and negative for Calponin, S100, CD34, Calretinin, and Pankeratin. Approximately 25% of tumor cell nuclei demonstrated positive nuclear immunostaining with Ki-67. At another surgical pathology laboratory, additional immunohistochemical stains performed were positive for CD99, patchy SMA, and focal Desmin, and were negative for CK5/6, Prostate Specific Antigen (PSA), SOX10, HMB45, and TLE1. In addition to immunohistochemical stains, a sarcoma fusion panel was performed by a molecular diagnostics laboratory which demonstrated that there were no fusion transcripts detected in the specimen. The interpretation of the panel also noted that the current assay used may not have detected uncommon fusion transcripts and recommended fluorescence in situ hybridization assay for translocation of a specific gene, if clinically indicated. The final diagnosis from the surgical pathology report revealed spindle cell sarcoma with focal smooth muscle differentiation, grade 3 with no fusion transcripts detected from the sarcoma fusion panel.

Within a few days of the final surgical pathology report, the patient had an appointment with the oncologist. During the appointment, the oncologist reviewed the pathology results with the patient of his nonspecific sarcoma subtype despite extensive testing and molecular testing. Moreover, different treatment scenarios were discussed with the patient. Chemotherapy was recommended and was begun after placement of a left subclavian vein port.

Discussion

As soft tissue sarcomas represent only 1% of all adult malignancies, it is difficult to make general conclusions about epidemiology [6]. Soft tissue sarcomas are distributed among all age groups, and although some reports suggest an increase in incidence among men [7], most reports suggest both genders are equally susceptible [8]. In a prior study, 3299 spindle cell sarcoma cases were identified and extracted from a Surveillance, Epidemiology, and End Results (SEER) database (1973-2017) revealing that spindle cell sarcoma most frequently occurs during the seventh decade of life with the mean age at diagnosis being 61 years old [9]. Additionally, overall race distribution includes 80.9% white, 11.4% black, and 7.8% American Indian/Asian/Pacific Islander [9]. These demographic factors demonstrate similarity to the patient's gender, age, and ethnicity.

Histologic grade currently represents the most significant prognostic factor for all soft tissue sarcomas in adults [10]. However, several limitations prevent its use: (1) grade cannot be applied to all histological types, (2) its reproducibility is not perfect, (3) a three-grade system generates an intermediate grade with undetermined prognosis, and (4) the core needle biopsy, now widely used for the diagnosis of soft tissue sarcoma, is not the best sample to assess the grade [11]. In addition to histologic grading, immunohistochemistry may be used to evaluate for the presence or absence of a mutated protein, expression of a protein in the same pathway as the altered gene, or even a protein resulting from a gene fusion [12]. Immunohistochemistry can be diagnostically helpful; however, staining may be patchy or negative in a subset of tumors and the immunoprofile of many soft tissue tumors often show overlap [12]. Due to many patients' incomplete immunoprofile, advances in our understanding of spindle cell sarcoma molecular testing are becoming increasingly essential [12]. The molecular testing done for this patient using the sarcoma fusion panel included only known gene fusions and despite no fusion transcripts detected, new fusion genes will be added to the panel in the near future.

The standard management of high-grade limb sarcomas includes surgical excision followed by postoperative radiation therapy, although occasionally amputation remains the only option [13]. The use of external beam radiation therapy has been widely advocated as a means of improving local control after surgical resection, although its effectiveness in this role has never been definitively determined [14]. According to a prospective randomized controlled trial done by Yang et al, the use of adjuvant postoperative external beam radiation therapy in addition to chemotherapy resulted in a statistically significant reduction in local recurrence as seen in 0 of 44 patients diagnosed with high-grade extremity sarcoma compared to 9 of 47 patients diagnosed with high-grade extremity sarcoma not receiving adjuvant postoperative radiation therapy [14]. In recurrent/metastatic soft tissue sarcoma patients, doxorubicin monotherapy is considered first line chemotherapy for most of the histological subtypes [15]. Since 2007, three novel agents including trabectedin, pazopanib, and eribulin, have been approved for the treatment of highgrade soft tissue sarcomas in the second-line setting after progression on anthracyclines [16]. Because soft tissue sarcomas are relatively uncommon, the fewer number of patients have limited the ability to conduct traditional clinical trials and advance drug development [16]. Further advances will require an improved understanding of the biological differences between soft tissue sarcoma subtypes, specific biomarkers to elucidate responses to treatment, and mechanisms of resistance [16]. In addition to discovering new therapeutic agents, current and future clinical trials to evaluate novel combinations, including immunotherapy combinations, will add to the growing number of treatment options for soft tissue sarcomas [16]. Innovative design of clinical trials is essential to maximize the impact of studies involving this rare disease and to understand the unique interplay between the tumor, microenvironment, and therapeutic interventions [16].

Conclusions

This case report highlights the rarity of soft tissue sarcomas, specifically spindle cell sarcoma in the groin, suggesting the complexity of management and treatment of high-grade soft tissue sarcomas. Available treatment options, including surgical resection, adjuvant radiation and adjuvant chemotherapy, have been studied and continue to be studied. However, the standard of care is still not well defined due to the different histological subtypes and immunohistochemical markers of spindle cell sarcomas.

Declarations

Informed consent: The patient in this study provided written informed consent prior to participation.

Author contributions: Madeline M. Foreman, OMS III, MS, provided substantial contributions to conception and design, acquisition of data, or analysis and interpretation of data; Madeline M. Foreman, OMS III, MS, drafted the article or revised it critically for important intellectual content; F. Jefferson Liner,

MD, gave final approval of the version of the article to be published; and all authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Ethic and legal declarations: The patient has provided consent on this case report for publication on September 9th, 2024. The consent was provided in paper form and collected by Dr. F. Jefferson Liner, MD.

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