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Spindle-Cell Rhabdomyosarcoma of the Lateral Pelvic Wall and Inguinal Canal A Case Report and Literature Review

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Abstract

Spindle cell sarcoma is a variant of Rhabdo Myo Sarcomas (RMS) which rarely presents in adulthood. Often, this entity has a predilection for head and neck. This pathology infrequently involves extra-abdominal pelvic areas. Herein, we present a case of spindle cell variant of spindle cell RMS of the lateral pelvic area and inguinal canal in a 50-year-old woman. This is the first case of its kind reported in the Englishlanguage literature. Negative margin radical resection is advised. For effective surgical planning, a biopsy-based preoperative diagnosis is essential. In the present case, a biopsy of the tumor's sluggish area resulted in an inaccurate pre-operative diagnosis. The abdominal wall and inguinal canal needed to be mesh-reconstructed after a wide resection.

Keywords: Pathology • Physicians • Abdominal wall • Surgical pathology • Diagnosis

Introduction

Rhabdo Myo Sarcoma (RMS) is a rare neoplasm that develops from undifferentiated skeletal muscle cells, of which Spindle Cell Rhabdomyosarcoma (SC-RMS) is an uncommon variant and comprises less than 10% of all reported RMS cases[1,2]. SCRMS was first described in 1992 as a prognostically favorable variant of RMS commonly encountered in the para testicular region of male pediatric patients [3,4]. Years later, physicians began to identify histologically similar cases in adults [5]. Adult cases are rarer and have worse prognoses than their pediatric counterparts [2]. Adult patients diagnosed with RMS most frequently complain of the presence of a painless, rapidly growing tumoral mass with associated symptoms related to encroachment on nearby tissue structures [2,6]. SC-RMS pathology displays a fascicular spindled-growth pattern, with immunohistochemical staining showing diffuse positivity for skeletal muscle markers [2]. Staining patterns are especially helpful to differentiate from leiomyosarcoma, a more common spindle-shaped tumor of smooth muscle origin [2]. Due to the paucity of data on outcomes of this rare disease, there is limited knowledge regarding the optimaltreatment modality for adult SC- RMS. As such, SC-RMS is treated aggressively with protocols similar to other RMS subtypes—a multimodality approach consisting of surgery, chemotherapy, and sometimes, adjuvant radiation. Here, we discuss a case of a 50-year-old female with the diagnosis of SC-RMS of the round ligament, treated with surgical resection and adjuvant chemotherapy. This report represents the first report case in the English language literature of SC-RMS in the inguinal canal of an adult female patient.

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Case Presentation

A 50-year-old peri-menopausal female presented with two-year history of slow-growing abdominal wall neoplasm. The patient experienced mild discomfort and abnormal uterine bleeding, without any other associated symptoms. Previous D and C and hysteroscopy results were negative for any malignant pathology. Follow-up MRI imaging revealed a fibroid uterus. The mass was characterized as a right-lower abdominal wall fibroid measuring 3.5 × 6.8 cm (Figure 1).

Core needle biopsy revealed a spindle cell tumor with features suggestive of leiomyoma with minimal atypia and low proliferation index. Subsequently, the patient underwent transabdominal hysterectomy and bilateral salpingectomy with simultaneous resection of the abdominal mass and complex mesh reconstruction of the lateral abdominal wall (Figure 2).

Intraoperatively, the tumor was well-encapsulated and appeared to emanate from the round ligament. Margins were negative. Surgical pathology specimen was read as spindle cell rhabdomyosarcoma pT2N0 with benign uterine findings. The patient was subsequently started on 8 cycles chemotherapy with VAI/VIT. Currently, she is without evidence of disease after a year postchemotherapy (Figure 3).

Discussion

Spindle Cell Rhabdomyosarcoma (SC-RSM) is a Rare Morphologic subtype of rhabdomyosarcoma (RMS) and most often presents in the paratesticular area of male children [2,3]. Despite its rarity, authors have detailed the presence of RMS in the adulthood, describing its difficulty in diagnosis and poor prognosis in this population [5,7]. Cavazzana, et al. reported 16 cases of SC-RMS in the adult population, presenting in both men (n=11) and women (n=5) most commonly in the head and neck region, but also within the upper and lower extremities, retroperitoneum, abdominal cavity, pretesticular region, and volva [2,3]. Our report is the first to describe spindle cell RMS in the female inguinal canal region. RMS generally presents with a rapidly growing and painless mass with additional symptomatology varying based on anatomical location affected and the involvement of metastases. In the current case, the mass involved the inguinal ligament and the patient suffered from lower abdominal discomfort. In the case of a suspected RMS, patients should undergo a CT or MRI scans of the correlated anatomical

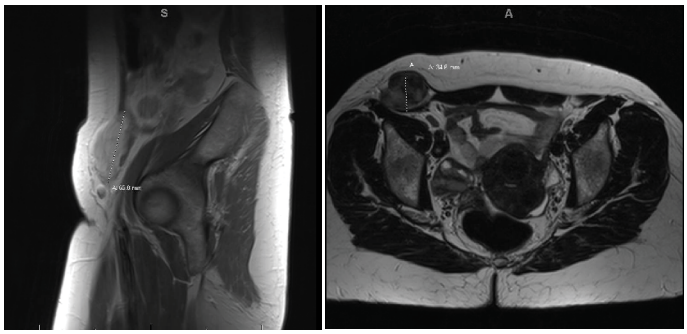

Figure 1. Sagittal (left) and coronal (right) views of the lobulated avidly enhancing soft tissue lesion measuring 3.5 × 6.8 cm within the fascia of the right lateral abdominal wall musculature demonstrating restricted diffusion.

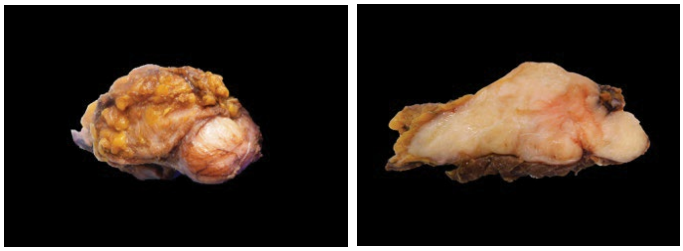

Figure 2. Pathologic gross specimen of pelvic mass

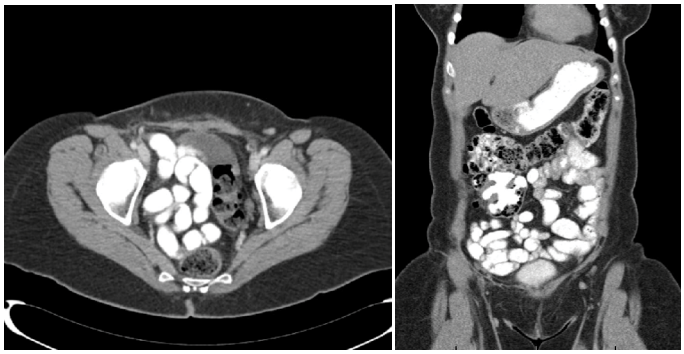

Figure 3. Most recent imaging as of July 2020 with transverse (left) and coronal (right) MRI images demonstrating no evidence of suspicious masses or lymphadenopathy in the chest, abdomen or pelvis.

region. Chest and abdominal imaging, bone scan, and bone marrow aspiration should be included in the workup for staging and evaluating potential sites of metastasis, most commonly involving the lung, bone marrow, and lymph nodes [8]. Importantly, a biopsy of the tumor is required for immunohistological diagnosis. The histologic features of SC-RMS are characterized by fascicular proliferation of spindle cells with rhabdomyoblastic differentiation and resemble features similar to fibrosarcoma, leiomyosarcoma, and malignant peripheral nerve sheath tumors [3]. Immunohistochemical staining shows diffuse positivity for skeletal muscle markers such as myf-4 & myo-d [2]. Due to its histologic similarity to benign pathology, SC-RMS may be initially misdiagnosed leading to suboptimal therapy. As such, we encourage physicians to have this entity in mind when working up its benign counterparts and assure ample tissue sampling when taking a biopsy and analyzing the pathologic features for histopathologic diagnosis. If possible, two to three random samples of the suspected lesion should be taken for pathologic diagnosis. Compared to children, adults have a seemingly less favorable prognosis. There are two hypotheses for this discrepancy. The first hypothesis is rooted in the idea that cancer cells can become less sensitive or resistant to chemotherapeutic drugs through Multidrug Resistance Proteins (MRPs) [9]. This phenomenon was investigated by Komdeur, et al., who compared the expression levels of specific multidrug resistance-associated proteins (i.e., P-glycoprotein (Pgp), Multidrug Resistance-associated Protein 1 (MRP1), and Lung Resistance-related Protein (LRP)) between children and adults with RMS. P-gp and MRP1 actively removing the drug from the cancer cell via a transmembrane pump [10]. LRP decreases the concentration of the chemotherapeutic drug within the cancer cell by distributing it away from the target site [11,12]. The expression levels of P-gp and MRP1 were found to be the same between children and adult patients with RMS [13]. However, specifically for embryonal and pleomorphic RMS types, Lung Resistance-Related Protein (LRP) was expressed more in the adult population. The presence of LRP has been shown to continuously drive chemotherapy-induced differentiation of cancer cells to express MRPs [10,14]. The additional multidrug resistance protein in adults that increases in expression throughout chemotherapy treatment may contribute to overall chemotherapy failure.

The second hypothesis focuses on the tumor location and histological variations. In adults, tumors often originate in more unfavorable locations [15]. Furthermore, children are far more likely to have embryonal and alveolar RMS subtypes, which have shown more favorable prognoses than the pleomorphic, spindle, or Not Otherwise Specified (NOS) variants, which are more common in adults [15,16]. Lastly, poorer outcomes in adults may be due to insufficient treatment [17,18]. The majority of clinical trials and treatment plans for RMS have been completed in the pediatric population and the application of these treatment plans to the adult population have been successful. However, despite this, some studies have shown that adult patients do not receive the same multimodality standard of care as children do [17,18]. This is likely due to a lack of understanding but should be further studied. Current treatment guidelines are modeled by those directed towards pediatric RMS created by the Intergroup Rhabdomyosarcoma Studies Group (IRSG) [19,20]. Treatment is primarily managed locally with the addition of chemotherapy and/or radiation therapy depending on the histological classification and pre-and postsurgical classifications. Chemotherapy follows either the VAC regimen (vincristine, actinomycin D, and cyclophosphamide) in North America or the IVA regimen (isofosfamide, vincristine, and actinomycin D) in Europe. When the neoplasm is of a higher grade and/or when a complete resection is not possible, radiotherapy is added to the treatment plan [8,20]. Adult RMS recurrence is common, and may occur with metastases, usually to the lungs so close monitoring is suggested.

Conclusion

This is the first report spindle-cell rhabdomyosarcoma of the female inguinal canal. This pathology is very aggressive in nature and requires multimodality treatment in order to decrease recurrence rates. Radical resection with negative margins is recommended. Establishing a pre-operative diagnosis via biopsy is important for adequate surgical planning. In the current case, biopsy of the indolent region of the tumor led to an incorrect pre-operative diagnosis. Wide resection required mesh reconstruction of the abdominal wall and inguinal canal. Currently, treatment modalities for adults with RMS are based on those developed for RMS in the pediatric population; further studies need to be done to establish optimal clinical guidelines for RMS in adults.

Conflict of Interest

None.

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