Spermatic cord dedifferentiated liposarcoma presenting as a recurrent inguinal hernia

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Paratesticular sarcomas are a rare entity and provide a unique clinical challenge due to their slow growing, often painless natural course. Adding to this challenge is the complex anatomy of the scrotum that allows these masses to mimic other conditions, including inguinal hernia, cysts, or fluid collections. We report such a case and our approach to an 83-year-old male with dedifferentiated liposarcoma of the spermatic cord with a history of inguinal hernia. In doing so, we highlight the need for thorough evaluation of scrotal masses and the management of these rare, though well-described, tumors.

Key Words: spermatic cord, paratesticular tumor, liposarcoma, inguinal hernia, orchiectomy

Introduction

The vast majority of urologic tumors in men originate from the testes, and paratesticular masses are exceedingly rare, comprising approximately 3% of scrotal masses. The spermatic cord is the site of origin for 90% of paratesticular masses, although determining the site of origin is often challenging.[1] Non-osseous soft tissue sarcomas (STS) account for less than 2% of all urologic tumors.[2] With respect to tumors arising within the soft tissue of the spermatic cord, sarcomas account for over a third of cases demonstrating a wide variety of differentiation. In this setting, the most commonly reported histologic subtypes of STS include liposarcoma (23%), malignant fibrous histiocytoma (21%) and leiomyosarcoma (13%).[3]

Given the complex anatomy of the scrotal contents, their proximity, and various embryologic elements, tumors arising within the scrotum are often clinically and histologically diverse. As a result, neoplasms arising in this location are heterogenous by nature, exhibit behaviors that are difficult to predict, and often blur anatomic boundaries. Such tumors may present a clinical challenge in distinguishing paratesticular from testicular site of origin, resulting in delayed or incorrect diagnosis. Though radiologic evaluation can help further delineate the anatomic site of the tumor, imaging provides little information in definitively differentiating benign from malignant tumors. As a result, such masses are generally considered malignant with radical orchiectomy and resection of involved paratesticular tissue serving both therapeutic and diagnostic purposes.

Consensus regarding effective management of paratesticular sarcomas is lacking because of their rarity, with only 212 cases of spermatic cord sarcomas reported between 1845 and 1978.[4] In this case study, we report our experience of an 83-year-old male with a history of inguinal hernia repair and review key diagnostic, treatment, management, and prognostic considerations in a case of liposarcoma of the spermatic cord.

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Case report

An 83-year-old man with coronary artery disease, diabetes mellitus type 2, and hypertension presented with concern for a mass of the right testis after experiencing intermittent, sharp scrotal pain for a year. The patient's past surgical history is significant for a previous right-sided inguinal hernia repair with a recurrence of an inguinal bulge in recent years. The patient's main concern was a suspected recurrence of his inguinal hernia. The scrotal mass was discovered on physical examination by the patient's primary care physician who obtained a scrotal ultrasound that revealed multiple solid lesions within the right hemiscrotum extending inferiorly along the spermatic cord. The aggregate size of these lesions was 6.1 cm along the longitudinal axis by ultrasonography. Due to suspected malignancy, the portion of the spermatic cord involved by the mass was excised with high en-bloc removal in addition to radical orchiectomy of the right testis.

Materials and methods

The surgical specimens were fixed with 10% neutral-buffered formalin and tissue sections were processed by standard methods and embedded in paraffin blocks. Four micrometer-thick paraffin sections were prepared and immunohistochemical staining for the following antibodies was performed: alpha smooth muscle actin and myosin (Cell Marque, Rocklin, CA, USA), in addition to vimentin, desmin, S-100, CD34, calretinin, and pancytokeratin (Ventana, Tucson, AZ, USA).

Results

On initial receipt of the resected specimens, gross examination revealed a 256 g orchiectomy specimen enveloped by a tan-pink tunica vaginalis consisting of a 4.5 cm x 2.2 cm x 3.0 cm testicle with a 7.0 cm x 1.5 cm x 1.0 cm tan-pink epididymis and an attached 3.5 cm x 0.8 cm dark red spermatic cord. A 9.5 cm x 7.0 cm x 6.0 cm partially encapsulated mass adjacent to the spermatic cord revealed tan-white firm, nodular cut surfaces with gelatinous, hemorrhagic and necrotic areas extending to within 3.5 cm of the spermatic cord margin.

Microscopic examination revealed a multinodular growth pattern with variable morphology, cellularity and degree of atypia. Overall, the tumor was solid and composed of vaguely nodular foci of spindle cells with interspersed areas of necrosis, the latter of which comprised approximately 10% of the tumor volume. Much of the tumor was composed of elongated spindle cells with pale eosinophilic cytoplasm and atypical ovoid to tapering nuclei arranged in a fascicular or patternless growth pattern within a background of collagenous stroma, Figure 1. In other areas, the tumor appeared less cellular and demonstrated prominent myxoid change, corresponding to the gelatinous foci seen on gross examination. Hypercellular areas with anaplastic morphologic, scattered bizarre cells and numerous mitotic figures, numbering up to 20 mitoses per 10 high power fields, were also present, Figure 2.

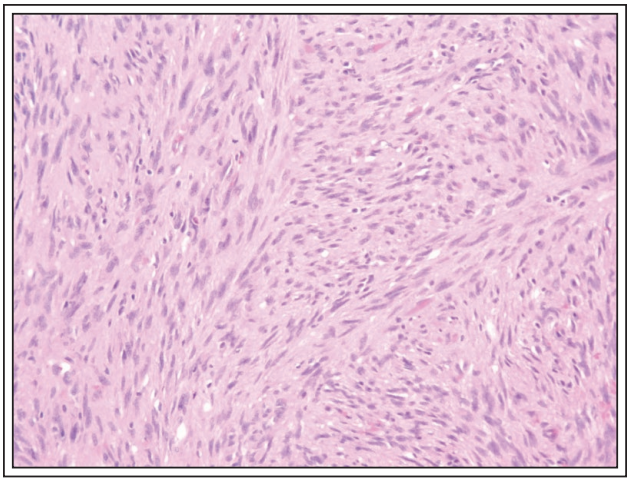

Figure 1. A significant portion of the sarcoma showed spindled growth with intersecting fascicles of elongated cells with moderate cytologic atypia. Evidence of adipocytic differentiation was virtually absent. (H&E stain, 400X).

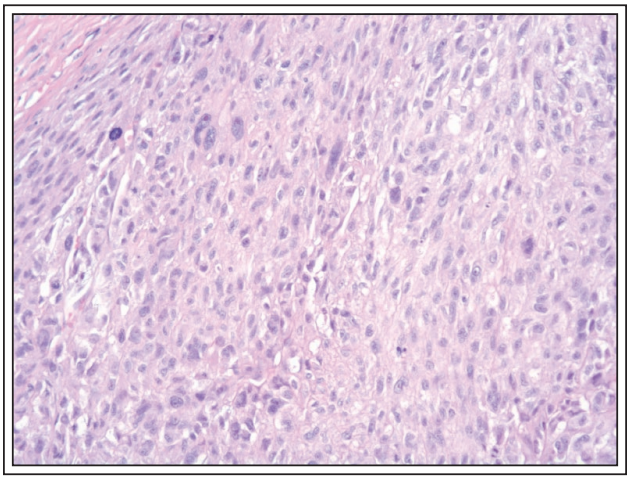

Figure 2. Foci of anaplasia were noted and are characteristic of dedifferentiated liposarcoma. These areas contained large, highly atypical cells with prominent nucleoli and frequent atypical mitotic figures. (H&E stain, 400X).

Of note, no areas of definitive adipocytic differentiation were identified.

Immunohistochemical staining showed multifocal, strong positivity for both MDM2 and CDK4 with weak immunoreactivity for smooth muscle actin and vimentin. Upon confirmation from an outside facility, the anatomic location, morphology, and immunophenotype of the 9.5 cm paratesticular mass was consistent with dedifferentiated liposarcoma.

Discussion

The majority of scrotal masses originate from the testicle, and unfortunately, malignancy is often the rule rather than the exception. Given the challenging anatomy of the scrotal contents, close proximity, and various embryological origins, differentiating between testicular and paratesticular masses may be a difficult task. The differential diagnosis for palpable scrotal masses should include inguinal hernia, hydrocele, varicocele, epididymal cyst, and both benign and malignant neoplasms.

Evaluation and management of a scrotal mass should begin with ultrasonography to elucidate solid masses from fluid collections and to identify characteristics suggestive of malignancy, such as solid, hyperechoic, and heterogenous lesions.[5] It should be reiterated that ultrasonography provides the initial location of an extratesticular mass, but cannot be used solely in distinguishing the nature of the lesion and that findings suggestive of benign masses still require more thorough evaluation, Figure 3. Any initial lesion suspicious for malignancy should be resected with subsequent microscopic and possible immunohistochemical evaluation as this is critical to establishing the definitive diagnosis.

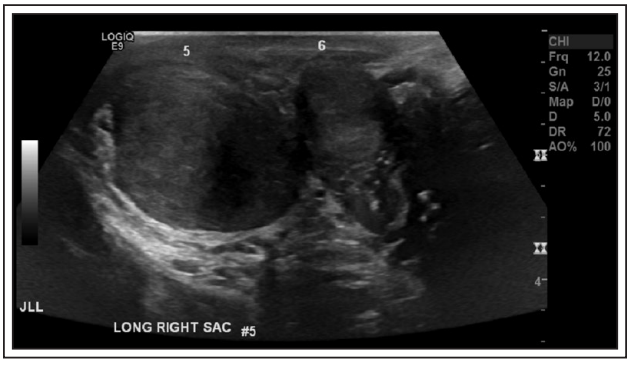

Figure 3. Ultrasonography (US) of right hemiscrotum. US reveals a well-defined, heterogenous mass with hyperechoic areas concerning for malignancy.

STS represent a heterogenous group of tumors that can be further differentiated into multiple sub-classifications. Sarcomas are the most common paratesticular tumors, accounting for approximately one-third of all cases. In the case of our patient, his presentation was typical for that of paratesticular sarcoma characterized as a slow-growing, often painless scrotal swelling most common between the fifth and seventh decades of life.[4,5] Interestingly, the prior history of inguinal hernia with repair in our patient may have contributed to his relative delay in seeking treatment due to his belief of this scrotal mass may have been a simple recurrence of prior pathology.

Liposarcoma typically occurs in the retroperitoneum, with the scrotum and spermatic cord being the third most common site for this lesion. There are five histologic subtypes of liposarcoma, including well-differentiated, lipoblastic, fibroblastic, myxoid/round cell and pleomorphic. Although liposarcomas are frequently well-differentiated, dedifferentiation may occur, a feature common to the biology of all sarcomas in general. Dedifferentiated liposarcomas most often represent de novo lesions, with the remainder developing from a preexisting well-differentiated subtype after an average of 7.7 years.[6] Compared to well-differentiated liposarcomas, dedifferentiated liposarcomas carry a poorer prognosis, but still behave less aggressively than high grade sarcomas.[7]

The proper management of paratesticular sarcomas remains controversial given the rarity of these tumors and the lack of consensus regarding treatment in the existing literature. Based on the experiences of several authors, if paratesticular or testicular tissue is involved, it is recommended that adults undergo radical orchiectomy, the procedure of choice for spermatic cord STS, with high cord ligation and wide excision of surrounding soft tissue structures within the inguinal canal as the standard of treatment.[5] Though liposarcomas are often low grade tumors, their risk of local recurrence and spread via local extension is on par with high grade sarcomas, thus necessitating aggressive complete surgical resection with negative margins to offer the best chance of cure in patients presenting with primary disease. Positive surgical margins and incomplete resection significantly increase local recurrence and mortality. In one reported series, one-half of patients experience recurrence of sarcomas, and this was attributed to inadequate perioperative radiographic staging owing to the rarity of these lesions. Overall, 75% and 55% of patients experience 5 and 10 year disease-specific survival, respectively.[5]

Debate ensues regarding the efficacy of retroperitoneal lymph node dissection (RPLND) and adjuvant radiation

therapy and chemotherapy.[4,8] Though RPLND has classically been employed for all high grade sarcomas, there is no clear survival benefit of superficial inguinal or RPLND, with the exception of the embryonal rhabdomyosarcoma variant.[6] In this case, we elected to proceed with radical orchiectomy and resection of the involved spermatic cord to achieve negative margins, as recurrences are frequent, owing to incomplete surgical removal of the tumor. In the case of liposarcoma in general, radiation therapy has been used for local control as liposarcomas are the most radio-sensitive sarcomas, though the results in paratesticular liposarcoma are not as well characterized.[9] Adjuvant radiation therapy is recommended in certain scenarios such as inadequate surgical margins, recurrence, lymphatic invasion, or high grade histology.[10] The indications for adjuvant chemotherapy remain unclear and are currently not recommended.

Conclusion

Paratesticular sarcomas are a rare entity, with most of the existing literature on this topic consisting of case reports and small series from select institutions. As a result, clinical judgment often directs ultimate management in these patients due to the lack of consensus in treatment guidelines and standard of care. Evaluation should begin with ultrasound to assess features more suggestive of malignancy. The only current agreed upon standard of treatment is aggressive surgical resection. The role of RPLND, adjuvant radiation therapy, and chemotherapy are controversial and largely unsupported. We report a case of paratesticular liposarcoma with dedifferentiated morphology and our treatment approach including aggressive surgical resection with high ligation. We hope that our experience may add to the existing literature, and that over time, the sum of the contributions, both preceding and following this report, may eliminate ambiguity and help establish a standardized approach and treatment plan for patients with paratesticular sarcomas. □

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