Primary seminal vesicle epithelioid smooth muscle neoplasm of uncertain biologic potential

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We present the case of a 73-year-old male patient who presented with obstructive urinary symptoms, pelvic pressure, and hematuria. CT imaging revealed a heterogenous prostate enlargement, and MRI demonstrated the mass to be arising from the seminal vesicle. Prostate biopsies showed benign tissue. Surgical excision was completed and pathology revealed it to be an epithelioid smooth muscle neoplasm of uncertain biologic potential. This is only the second known case of such a seminal vesicle tumour. As soft tissue sarcomas of the seminal vesicle emerge in the literature, we may develop a better understanding of their biologic behaviour and prognostic potential.

Key Words: smooth muscle neoplasm of uncertain biologic potential, seminal vesicle

Introduction

Primary neoplasms of the seminal vesicle are uncommon, with most neoplasms of the seminal vesicle being secondary to malignant spread.[1] Of the primary neoplasms documented, most can be classified as epithelial, mesenchymal, or mixed epithelial-stromal tumors, with epithelial adenocarcinomas seen most frequently.[2] Mesenchymal tumors however are rare; the largest study to date of soft tissue sarcomas of the genitourinary tract observed 188 cases over a 25 year period at a high volume institution and only two of these cases included seminal vesicles.[3] This is reflected in the larger literature, where smooth muscle tumors in particular make up a small fraction of primary seminal vesicle tumors. We performed a systematic review of the literature, which demonstrated 14 cases of leiomyomas of the seminal vesicle and 11 cases of leiomyosarcomas of the seminal vesicle as of March 2021.

There do exist smooth muscle tumors that do not fit the dichotomous criteria of either leiomyoma or leiomyosarcoma, designated as smooth muscle tumors of uncertain biologic potential.[4] To date, there is only one prior published smooth muscle tumor of uncertain biologic potential of the seminal vesicle, found incidentally on prostatectomy for a prostate adenocarcinoma.[5] We present a case report of a seminal vesicle epithelioid smooth muscle tumor of uncertain biologic potential which was found on MRI after the patient presented with obstructive symptoms. This is only the second known case of a seminal vesicle smooth muscle tumor of uncertain biologic potential and the first to accompany a grossly benign prostate. We describe herein the surgical management and follow up of this case.

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Case report

A 73-year-old male patient presented to his urologist with worsening voiding symptoms, including pressure, hesitancy, and straining. His past medical history is significant for prostatitis, BPH, dyslipidemia, and hypertension. He has a remote smoking history having quit approximately 40 years ago. His surgical history at time of presentation included a previous excision of left epididymal cyst and benign mesothelial cyst. Examination of the patient revealed a moderately enlarged symmetric benign-feeling prostate, and a subsequent flow study revealed obstructive symptoms. He was started on finasteride and tamsulosin. A few days after this presentation, the patient developed hematuria. A renal and pelvis CT showed heterogenous prostate enlargement with extension superiorly that could not be differentiated from the seminal vesicles. A follow up MRI with gadolinium in the same month allowed for distinguishing of a 5.7 cm x 5.7 cm x 5.4 cm heterogenous mass centered within the seminal vesicles, slightly left of the midline, favored to be separate from the prostate gland, Figure 1. The prostate itself was enlarged; no pathologic pelvic lymphadenopathy was seen on this image.

Core biopsies were taken to further characterize the seminal vesicle mass; core biopsies of the prostate were also taken. The prostate biopsies showed some active chronic inflammation but were negative for malignancy. The biopsies of the seminal vesicle mass demonstrated small blue cells with rounded nuclei and scant cytoplasm with a focal myxoid background, positive staining for SMA (smooth muscle actin) and desmin, with some CD99 positive staining, scarce mitoses, and very occasional nuclear atypia. We sent our specimen to a second outside institution, who confirmed the non-specific findings and the difficulty in classifying this lesion. The possibility of a prostatic stromal tumor was considered, as further staining at the outside institution demonstrated diffuse nuclear positivity with androgen receptors. However, the diffuse positive staining with desmin and negative CD34 did not support a stromal tumor. Therefore, the decision was made to label the lesion with a descriptive diagnosis, as an atypical spindle and round cell neoplasm.

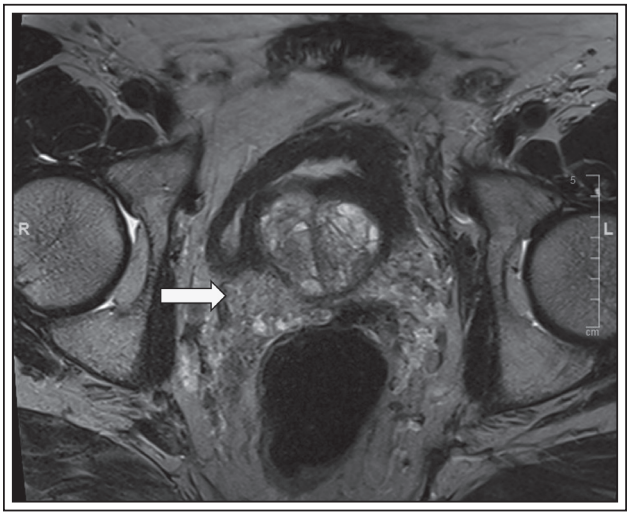

Figure 1. Axial T2 weighted 3T MRI image shows arrow pointing to lesion representing primary seminal vesicle epithelioid smooth muscle neoplasm of uncertain biologic potential. Posterior to lesion is fragment of right seminal vesicle; anteriorly one can see the prostate.

We decided to proceed with surgical excision of the lesion for diagnostic and therapeutic purposes. We consented the patient for an extensive laparotomy including the possibility of a prostatectomy, cystectomy, ileal conduit and/or colostomy. Ureteric stents were placed bilaterally for identification. We utilized an infra-umbilical midline incision and performed an extended left-sided lymph node dissection. We then proceeded to identify the lesion which extended from the left over and across the midline. We were able to dissect the mass away from the bladder anteriorly and follow the plane we created medially and then posteriorly. The stretched vas deferens came around the back of the mass so we clamped, ligated, and divided this in a left vasectomy to isolate the mass. Then we completed the dissection laterally and inferiorly and were able to elevate the mass to see the entrance of the seminal vesicle to the ejaculatory duct. We transected between the two structures to release the mass and leave the ejaculatory duct intact. The mass was removed and sent dry to pathology.

The isolated lesion was evaluated by our pathologist and was again assessed independently at a same second institution. The gross specimen consisted of a large, circumscribed mass with a tubular structure (seminal vesicle), weighing 72 grams (fresh weight) and measuring 6.2 cm x 5.5 cm x 4.5 cm, Figure 2. The lesion was found to consist of rounded epithelioid cells with pale to eosinophilic cytoplasm, fairly uniform nuclei without significant pleomorphism, and scarce mitoses. Multiple immunohistochemical stains were performed, some at our institution, and additional ones at the outside institution. The lesion stained

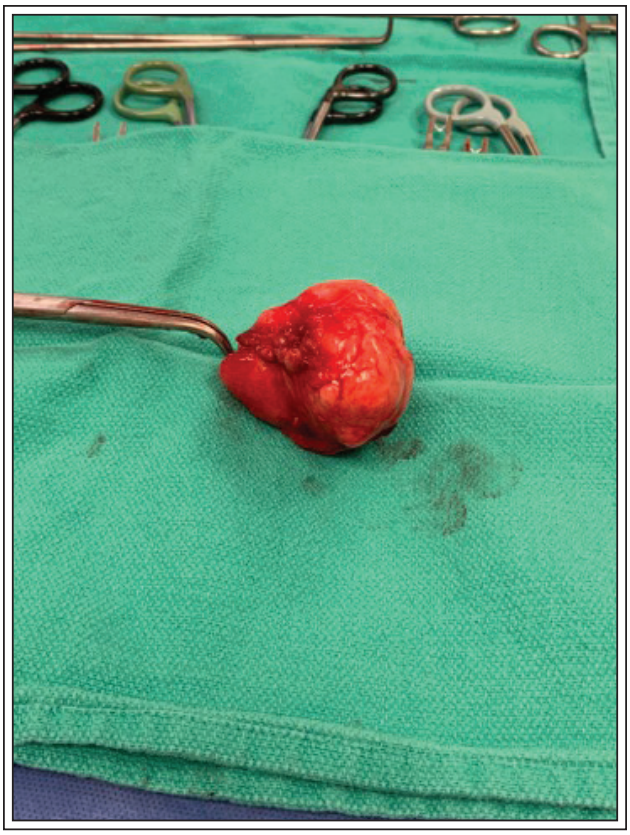

Figure 2. Gross specimen.

positive for desmin, caldesmon, SMA, vimentin, CD99, and calretinin, with diffuse nuclear positivity for androgen receptor and scattered cells positive for ER and PR, Figure 3. The lesion stained negative for CD34, MYOD1, pan-keratin, AFP, inhibin, S100 protein, GFAP, myogenin, MART-1, PLAP, and EMA. Given the findings, it was agreed that the lesion would best be considered as an epithelioid smooth muscle neoplasm of uncertain biologic potential, though without any overt features of malignancy.

The patient was admitted postoperatively, with a stable course in hospital, and discharged after 6 days. The patient was seen 1 month postoperatively with no concerns and was noted to have improvement in is urinary symptoms with diminished pelvic pressure. The lesion was reviewed widely with urology, oncology, and pathology and given the uncertain biologic potential it was decided that follow up surveillance with MRI would be appropriate. The first follow up MRI was completed approximately 6 months postoperatively and showed stable hypertrophy of the prostate, with no evidence of tumor recurrence or adenopathy.

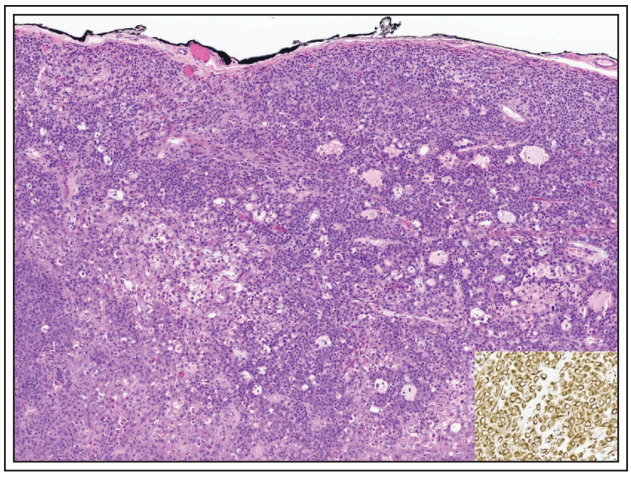

Figure 3. Histologic image (H&E staining). Right corner demonstrates positive desmin immunohistochemical stain.

Discussion

Mesenchymal tumors are rare examples of primary tumors found in the seminal vesicles. In particular, a smooth muscle neoplasm of uncertain biologic potential of the seminal vesicle has only been described once prior to our case; accordingly, the presentation and subsequent management of these lesions remains specific to the individual case. However, management may be guided by histologic features, as well as retrospective analysis of cases of other smooth muscle tumors of the seminal vesicle.

In the literature, presentation of primary seminal vesicle lesions range from asymptomatic to symptoms secondary to the seminal vesicles' close proximity to adjacent organs. Leiomyoma may present with smaller tumors and asymptomatic features; leiomyosarcoma tend to be larger, with pelvic pain and obstructive symptoms.[1] Our patient presented with obstructive symptoms including pelvic pressure which was relieved post-seminal vesiculectomy. Despite the rarity of this lesion, it is important to consider seminal vesicle neoplasms in urologic patients presenting with non-specific symptoms.

When neoplasms of the seminal vesicle are suspected, MRI remains the imaging of choice as it has the best tissue discrimination and multiplanar imaging.[6] In our case, initial CT imaging did not isolate the smooth muscle tumor of the seminal vesicle from the prostate; rather, it was use of MRI that allowed for better resolution of the soft tissue. Previous case reports of MRI imaging in a seminal vesicle leiomyoma

showed low intensity signal in T2-weighted sequences, consistent with epithelial lesions.[7] Leiomyosarcomas however tend to present on MRI as non-specific masses with low-intensity signal in T1-weighted sequences, and high-intensity signal in T2-weighted sequences.[6] Our MRI findings showed decreased signal in both the T1 and T2 weighted sequences. Leiomyosarcomas also tend to be irregular and poorly outlined, without a capsule or membrane. We were able to identify a well encapsulated mass with suspected seminal vesicle origin. Ultimately tissue sampling is required for histologic diagnosis, however MRI is important to assess the soft tissue and in our case it allowed for better surgical planning to obtain the tissue.

In establishing our differential diagnosis, we performed routine work up for secondary malignancy, however we largely relied on evaluation of this lesion through immune-histochemical staining. Though leiomyomas may commonly be found in the female genitourinary tract, they are less prevalent in the male genitourinary tract and as a result the immune-histochemical differentiation remains controversial.[8] In males, most primary seminal vesicle tumours can be classified as epithelial, mesenchymal, or MEST tumors. An epithelioid smooth muscle neoplasm is a distinct entity that lacks enough histologic features to characterize it to a subtype, rather the practical designation is to consider it as a smooth muscle tumor of uncertain biologic potential.

Our initial biopsy stained positive for SMA, desmin, and CD99, and negative for CD34, which prompted suspicion of a tumor of smooth muscle origin. The final pathology showed rounded epithelioid cells, and again stained positive for desmin and SMA, with nuclear positivity for androgen receptors, and negative staining for CD34. It was also strongly and diffusely positive for caldesmon, which helped to delineate the lesion as a smooth muscle neoplasm. There were no overt features of malignancy and the decision was made to label this as an epithelioid smooth muscle neoplasm of uncertain biologic potential. Despite the uncertainty in our sample, it is evident that use of immunohistochemistry remains important in analysis of any primary seminal vesical lesion.

In light of the mixed features of the lesion, treatment plan was widely discussed among our urology, pathology, and oncology team. For seminal vesicle leiomyomas, the risk of malignant change is quoted as less than 10%.[8] However our lesions' uncertain biologic potential as well as the patient's symptomology prompted us to perform a surgical excision. Retrospective studies show that surgical resection of genitourinary sarcomas is the most important prognostic indicator of long term survival.[3] For postoperative management, we opted for MRI surveillance, as radiotherapy or chemotherapy remains widely debated and is largely reserved for patients with positive surgical margins or conditions unamenable to surgery.[3]

Given the relative paucity of literature describing primary seminal vesicle epithelioid smooth muscle neoplasm of uncertain biologic potential, definite prognostication is limited. In the case presented, we were able to use imaging and immunohistochemistry staining to assess our lesion and plan for treatment; the patient tolerated surgical excision well and has had no local recurrence in the first year of follow up. In the future, case reports such as the one presented here may allow for development of diagnostic criteria that guide future management and treatment. □

References

1. Lopez-Beltran A, Menendezi CL, Montironi R, Cheng L. Rare tumors and tumor-like conditions in urological pathology. Springer International Publishing;2015.
2. Humphrey PA, Moch H, Cubilla AL, Ulbright TM, Reuter VE. The 2016 WHO classification of tumours of the urinary system and male genital organs-part B: prostate and bladder tumours. *Eur Urol* 2016;70(1):106-119.
3. Wang X, Tu X, Tan P et al. Adult genitourinary sarcoma: clinical characteristics and survival in a series of patients treated at a high-volume institution. *Int J Urol* 2017;24(6):425-431.
4. Miettinen M. Smooth muscle tumors of soft tissue and non-uterine viscera: biology and prognosis. *Mod Pathol* 2014;27(S1):S17-S29.
5. Samadi DB, Chughtai B, Akhavan A, Guru K, Rehman J. Incidental seminal vesicle smooth muscle neoplasm of unknown malignancy following robotic-assisted laparoscopic prostatectomy. *Can J Urol* 2008;15(3):4109-4111.
6. Vliet M, Kliffen M, Krestin GP, Dijke CF. Soft tissue sarcomas at a glance: clinical, histological, and MR imaging features of malignant extremity soft tissue tumors. *Eur Radiol* 2009;19(6):1499-1511.
7. Oliveira TS, Stamoulis DNJ, de Souza LRMF, Meneses ACO, Mori MM. Leiomyoma of the seminal vesicle. *Radiol Bras* 2018; 51(3):200-201.
8. Shaikh AS, Bakhshi GD, Khan AS, Jamadar NM, Nirmala AK, Raza AA. Leiomyoma of the seminal vesicle: a rare case. *Clin Pract* 2013;3(2):32.