A 26-year old female with leiomyosarcoma of the bladder and a serendipitous cystectomy – A success story from International Volunteers in Urology

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Introduction

Leiomyosarcoma is an extremely rare bladder malignancy, with fewer than 200 cases reported in the literature. The majority of reported cases have occurred in patients in the 6th and 7th decades of life. Five-year survival rates are reported between 47 and 62% and metastatic disease is common. Treatment has traditionally included radical cystectomy with lymph node dissection, though more recent series report that partial cystectomy is acceptable as long as a negative surgical margin is achieved. Here we report a case of leiomyosarcoma of the bladder in a 26-year-old female, treated with a gynecologic-tract and bladder neck sparing partial cystectomy with a “W” pouch neobladder.

Case presentation

A 26-year-old female presented with gross hematuria. She had no prior chemotherapy or significant exposure history. A CT scan was performed and a roughly 4.5 cm mass was seen within the bladder (Fig. 1). No additional lymphadenopathy or signs of metastatic disease were seen. She underwent transurethral resection of bladder tumor (TURBT) with pathology returning as leiomyosarcoma.

Shortly after the TURBT was performed, our team with International Volunteers in Urology (IVU) arrived. The case was presented to the team (as well as remotely to a number of Urologic Oncology faculty at The University of Iowa) and treatment options were discussed with the patient. It was quickly noted that obtaining ostomy supplies would effectively be impossible and that the patient desired to bear children later in life. On October 30, 2015, the IVU team, along with the local Urologists performed a gynecologic-tract and bladder neck sparing partial cystectomy with a “W” pouch neobladder. Intraoperatively, frozen sections and lymph nodes were taken and sent to pathology. A negative margin was achieved but was found to be close to the ureteral orifices. As such, the decision to perform a Wallace anastomosis was made. The decision to perform the bladder neck sparing cystectomy was based on tumor location and decided intraoperatively (Fig. 2). She remained in the hospital for 7 days and was discharged home with a suprapubic tube and catheter. The urinary catheter and ureteral stents were removed on POD 21. The remaining suprapubic catheter was removed one week later. Final pathology of the specimen was low grade leiomyosarcoma.

The patient is now 2 years post-surgery and remains cancer free based on her last CT scan in October of 2017. She is able to void voluntarily but does carry a significant residual. As such, she performs intermittent catheterization daily. She is very satisfied with the outcome and is enjoying a high quality of life.

Discussion

Leiomyosarcoma (LMS) is an exceptionally rare malignancy of the bladder. Sarcomas account less than 1% of all bladder malignancies with LMS reportedly representing less than 0.1% of all bladder cancers. The disease has a slight female predominance and most often presents exclusively, though reports of concurrent urothelial cell carcinoma exist.

Given the low number of cases, management is often challenging. Negative surgical margins have long been the goal with cystectomy as the mainstay of treatment. More recent reports have shown partial cystectomy as a viable option.

Despite, reports of non-surgical options, the primary treatment modality for bladder LMS remains radical or partial cystectomy. Regardless of treatment, given the aggressiveness of these tumors, prompt treatment is imperative. Margins status has been shown to be the main prognostic factor at final pathology. Local recurrence rates 16%, with the majority occurring within the first few years following...
treatment, demonstrating the need for close follow up. Survival differences between patients with low grade and high grade LMS are also vastly different. At a mean follow up of 47 months, Lee et al. reported disease related mortality of 0% vs. 50% for low vs. high grade LMS, respectively. The largest report on LMS to date included 183 patients and found that undifferentiated tumor grade was associated with worse outcomes and that most patients (63.2%) present with high grade tumors. Furthermore, many patients present with or develop metastatic disease, with up to 50% experiencing distant metastases.

Here, we report our case in which a 26-year-old female with no prior chemotherapy exposures or known risk factors presented with hematuria and was ultimately diagnoses with low grade LMS of the bladder. Our IVU team included a surgeon well versed in open cystectomy with neobladder formation. The Urologists at our Rwanda location were unfamiliar with these surgical techniques but were familiar with care of complex patients. The patient stressed the importance of maintaining the option for child bearing later in life. In addition, the patient and the local Urologists made it known that obtaining urostomy supplies was incredibly difficult. The decision to spare her bladder neck intraoperatively was made, in hopes that she would retain the ability to urinate voluntarily. The patient was seen at two years follow up and underwent a CT scan which demonstrated no evidence of disease (Fig. 3). She is able to void with Valsalva and performs intermittent catheterization 2 times per day given an elevated post void residual. She is very satisfied with her outcomes.

We believe this to be one of only a handful of bladder LMS treated with gynecological-tract sparing cystectomy with an ileal neobladder. It is likely the first in which the bladder neck was spared to preserve sphincteric function. She continues to perform intermittent catheterization, a practice not uncommon following traditional cystectomy with neobladder in women.

Conclusion

Leiomyosarcoma of the bladder is an ultra-rare entity and treatment should include prompt surgical removal with radical or partial cystectomy. Gynecological-tract sparing surgery appears appropriate for women of child bearing age, when local invasion is absent. Recurrence rates are high, especially for patients with high grade disease, leading to the need for continued long term monitoring. At 2 years of follow up, our patient is doing very well and is a prime example of the impact that international urologic volunteer work can have on patients.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eucr.2018.08.006.

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