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Robotic partial cystectomy for venous malformation of the bladder

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Introduction

Congenital vascular malformations of the urinary tract are uncommon. They may consist of venous, arterial, and/or lymphatic components. Hemangiomas, which have an arterial component, are the most common type of vascular malformation, yet still represent only 0.6% of all bladder lesions; venous malformations (VMs) are even rarer. Patients typically present with gross hematuria, although suprapubic pain and lower urinary tract symptoms have also been described[1]. Treatment of vascular malformations of the bladder ranges from endoscopic fulguration to excision depending on the extent of the disease. While open surgical treatment has been previously described, robotic partial cystectomy for VM of the bladder is a rare procedure. This report describes an unusual case in a teenager.

Case presentation

A 15-year-old female presented with severe gross hematuria for one week. Hemoglobin was 7.4 g/dL on initial presentation. She was noted to have a history of vascular malformations of the right leg and bladder. At age 12, she underwent arteriography for the bladder malformation which revealed no arterial inflow to the mass consistent with a VM, which was then treated through a combined laparoscopic plus cystoscopic injection of a sclerosing agent. She had also undergone multiple sclerotherapy and embolization procedures of her lower extremity lesion. Full body magnetic resonance imaging (MRI) was obtained; pelvic imaging (Fig. 1) demonstrated a 5 cm vascular bladder mass with intra- and extraluminal components, without any arterial enhancing component. Imaging also was notable for vascular lesions in her right lower extremity as well as right gluteal regions.

Given her persistent gross hematuria, the patient was taken to the operating room for cystoscopy and spot fulguration of the mass; bladder capacity was 400 mL. Her urine cleared, and she was discharged home. A genetics consultation suggested somatic mosaic mutations rather than a germline mutation in the genes involved in angiogenesis. Over the next several months she continued to have intermittent hematuria and urinary frequency, and the lesion grew slightly larger on repeat imaging. The decision was made to proceed with surgical resection of the mass via a robotic approach.

The video (video) highlights technical considerations of robotic partial cystectomy in the setting of vascular malformations of the bladder. The sigmoid colon was noted intraoperatively to have multiple small venous blebs without any apparent involvement with the bladder mass. Simultaneous visualization of the bladder mass both robotically and cystoscopically allowed for more precise planning and excision of the mass while also taking care to optimize postoperative bladder capacity. The mass was resected using a combination of the robotic Vessel Sealer, bipolar forceps, and curved monopolar scissors. The bladder was closed in 2 layers and noted to be leak-free at a volume of 240 mL. Pathologic evaluation (Fig. 2) confirmed a benign vascular malformation. The patient was discharged home on postoperative day two on oxybutynin. A cystogram performed 2 weeks postoperatively confirmed no leak and the catheter was removed. Two months postoperatively, she was voiding every 4–6 hours.

Supplementary video related to this article can be found at http://dx.doi.org/10.1016/j.eucr.2018.07.003.

She was subsequently admitted with bloody stools and a hemoglobin of 5.7 g/dL, and colonoscopy revealed a vascular malformation from 40 to 65 cm of the descending colon. MRI revealed mild enhancement of the wall of the sigmoid colon without any arterial enhancement. She was taken to the operating room for laparoscopic extended left hemicolectomy. Pathology revealed diffuse vascular malformations. Currently she is doing well.

Abbreviations: KTS, Klippel-Trenaunay Syndrome; MRI, magnetic resonance imaging; VM, venous malformation

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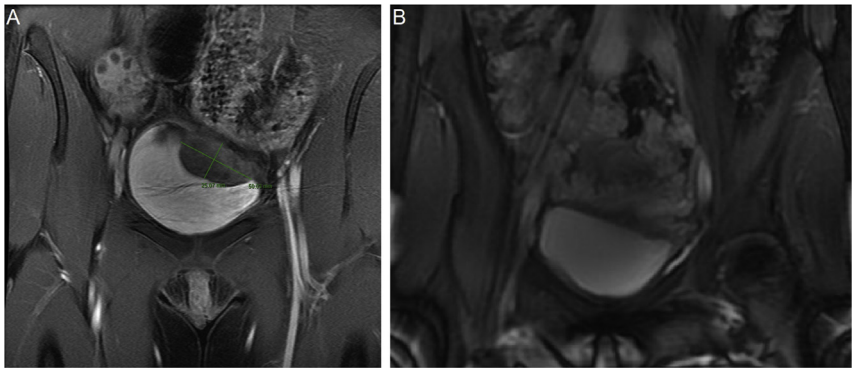

Fig. 1. MRI - Preoperative MRI (A) shows a 5 cm vascular mass with intra- and extraluminal components, without any arterial enhancing component. Postoperative MRI (B) obtained for gastrointestinal bleeding demonstrates smooth bladder wall without any lesions or filling defects.

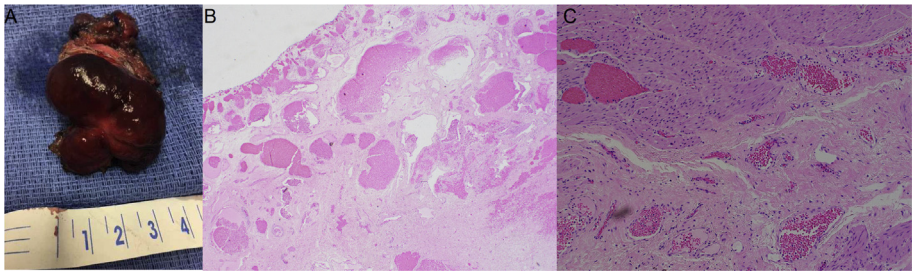

Fig. 2. Venous malformation – Gross specimen (A) and histopathological examination of the superficial (B) and deep (C) portions of the soft tissue mass as seen after H&E stain.

Discussion

VM of the bladder is a rare benign mesenchymal tumor. Hemangiomas are the most common vascular tumor of the urinary tract affecting the kidney and bladder. Patients typically present with gross hematuria. Most hemangiomas of the bladder are solitary and occur on the posterior wall or dome. Mean age at presentation is 58 years, and childhood disease is rare[2]. Bladder VMs are even less commonly described. Recognition of this subtype of vascular tumors is important, particularly in considering appropriate diagnostic tests and treatment options, as lesions will not be amenable to trans-arterial catheter embolization.

Cutaneous genital VM may predict involvement of internal structures, most commonly described in Klippel-Trenaunay Syndrome (KTS). This disease, caused by mutations in the PIK3CA gene, is characterized by port-wine stain dermatologic lesions, varicose veins, hypertrophy of bony and soft tissues including leg hypertrophy, and complex vascular anomalies with capillary, venous, and lymphatic components. One of the largest studies to date which evaluated 218 patients with KTS found that 23% of patients had cutaneous genital lesions, and up to 30% had genitourinary involvement; more than half of patients required interventional therapy in the form of endoscopic fulguration, angiographic embolization, or surgical excision[3]. In the present case which lacked characteristic lesions seen in KTS, genetic testing failed to identify a known germline mutation.

Different treatment options exist for genitourinary VM depending on location, size, and severity of symptoms. These include conservative management, cystoscopic fulguration using laser or electrocautery, sclerotherapy, and excision. Many tumors have extravesical extension making endoscopic management less suitable[4]. One case has been reported of a combined laparoscopic and cystoscopic approach with injection of sclerotherapy agent using 3% sodium tetradecyl sulphate in a child with a bladder VM whose lesion was not amenable to surgical excision[5]. Our patient had previously undergone a similar procedure years prior to presentation to our institution. We were unable to identify prior reports in the literature of robotic partial cystectomy for the treatment of VM of the bladder.

Conclusion

This case reinforces the typical presentation of vascular malformation of the bladder, highlighting the unique role of robotic approach to perform partial cystectomy in the treatment of bladder VM. We believe that a combined robotic and cystoscopic approach allows for clear visualization of the intravesical tumor margins to maximize postoperative bladder capacity.

Consent

Written/signed consent has been obtained by the patient's mother.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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