A rare case of a retroperitoneal lymphangioma causing chronic flank pain in an adult

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1. Introduction

Lymphangiomas are rare, non-malignant tumors that typically present before the age of two. These lymphatic malformations are commonly located in the neck and axilla (95%), and infrequently throughout the rest of the body (5%). Retroperitoneal lymphangiomas are exceedingly rare (<1.0%), and are usually asymptomatic and identified incidentally on cross-sectional imaging. These tumors are infrequently encountered by urologists and are usually managed by pediatric or general surgeons given the young age at presentation. Below, we present a unique case of an adult who underwent robotic-assisted, laparoscopic resection of a large retroperitoneal lymphangioma for the treatment of chronic flank pain.

2. Case presentation

A 27 year-old female with history of obesity, urolithiasis, bipolar disorder, and Ehlers-Danlos syndrome presented to the emergency department with 1-week of worsening, acute-on-chronic, right flank pain radiating towards her hip. The patient underwent a CT abdomen and pelvis with contrast that showed a 7.6 × 7.9 × 12.6 cm cystic mass located medial and inferior to the right kidney (Fig. 1). There was no evidence of right-sided hydronephrosis. The patient reported a 4 year history of chronic, intermittent right flank pain. She had previously seen an outside urologist who diagnosed her with a right, simple renal cyst based on retroperitoneal ultrasound. Her surgical history was notable for right-sided ureteroscopy for urolithiasis with prolonged ureteral stent placement due to a small caliber ureter. There was no personal or family history of genitourinary malignancy or congenital malformation.

Given the size and location of the mass it was thought to be potentially causing intermittent ureteral obstruction and/or mass effect on the right kidney. Therefore, the patient was extensively counseled on management options, including continued observation, needle aspiration with symptom reassessment, or definitive surgical excision. The patient elected for surgery. A pre-operative MRI with and without contrast to better define the architecture of the lesion showed an 8.0 × 6.5 × 12.3 cm multi-cystic lesion located medial and inferior to the right kidney with internal septations that were hyper-dense on T2-weighted imaging (Fig. 2).

The patient subsequently underwent an uncomplicated robotic-assisted, laparoscopic resection of the mass. A retrograde pyelogram was performed and right ureteral stent was placed at the beginning of the case to help delineate the ureter and assess whether the mass emanated from it. The mass was resected inferior to superior towards the renal hilum. The stalk of the mass was ligated at the posterior renal hilum using an endovascular stapler. The mass was resected inferior to superior towards the renal hilum. The stalk of the mass was ligated at the posterior renal hilum using an endovascular stapler. The mass was resected inferior to superior towards the renal hilum. The stalk of the mass was ligated at the posterior renal hilum using an endovascular stapler. The mass was resected inferior to superior towards the renal hilum. The stalk of the mass was ligated at the posterior renal hilum using an endovascular stapler. The mass was resected inferior to superior towards the renal hilum. 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Fig. 1. CT Abdomen and Pelvis with Contrast demonstrating a multilobulated 7.9 × 6.0 × 12.6 cm hypoattenuating cystic lesion with few punctate internal calcifications medial and inferior to right kidney.

Fig. 2. MRI of Abdomen and Pelvis demonstrating an 8.0 × 6.5 × 12.3 cm lesion with hyperintense signal on T2-weighted imaging. A few internal septations are noted which demonstrate mild enhancement on post-contrast sequences.
course was uncomplicated and she was discharged on post-operative day one. Her post-operative course was notable for a surgical site seroma that required percutaneous drainage. Of note, the patient reported resolution of her flank pain immediately postoperatively as well as at 4-weeks follow-up. Final pathology was consistent with a benign lymphangioma positive for D2-40 and CD34 on immunohistochemical staining (Fig. 3).

3. Discussion

Retroperitoneal lymphangiomas are rare, benign tumors that can be congenital or acquired. Primary or congenital lymphangiomas occur in children and are thought to arise from early lymphatic tissue that fails to form normal connections with the main lymphatic system, ultimately leading to lymphangiectasia. Secondary or acquired lymphangiomas tend to develop in adults and in areas of chronic lymphatic obstruction, chronic infection/inflammation, prior surgical resection, and radiation. These tumors are normally asymptomatic, but in rare circumstances can result in flank pain, lumbar pain, and vague abdominal symptoms when they are large and causing mass effect on surrounding organs.

In the case of our patient, it’s unclear whether she suffered from a congenital or acquired lymphangioma. It’s possible that a congenital lymphangioma grew as she aged and eventually caused her symptoms. However, Ehlers-Danlos syndrome is also known to be associated with lymphatic channel disruption. It’s possible that her underlying collagen disorder put her at greater risk for lymphangioma development, especially after complex ureteroscopy with prolonged ureteral stent placement on the ipsilateral side. The incidence of post-ureteroscopy lymphangioma development is unknown, but is likely also extremely rare.

Prior case reports have demonstrated long-term success with open and laparoscopic resection of symptomatic retroperitoneal lymphangiomas. In 2019, Stem and colleagues reported using a robotic-assisted, laparoscopic approach to resect a large retroperitoneal lymphangioma causing flank pain an elderly female. Similar to our experience, the robotic-assisted approach provided excellent visualization with minimal patient morbidity. In addition, our placement of a ureteral stent at the beginning of the case was advantageous in identifying and protecting the ureter throughout the procedure. Similar to prior reports, resection of the retroperitoneal lymphangioma resulted in relief of the patients chronic right flank pain.

To-date, only one case of a retroperitoneal lymphangioma resection has been reported in the urology literature. In 2019, Chaker and colleagues described an elderly female who presented with left flank pain and lower urinary tract symptoms who was found to have a retroperitoneal cystic mass. The patient underwent an exploratory lumbotomy with resection of the mass with final pathology showing a lymphangioma. The authors noted that their experience with retroperitoneal renal surgery was advantageous during their dissection.

Given the young age of presentation, pediatric and general surgeons most frequently encounter and manage patients with symptomatic lymphangiomas. However, given today’s urology residency program training, urologists are likely uniquely positioned to manage these lymphatic malformations when they are peri-renal and diagnosed in adult patients. Overall, our case demonstrates that these benign tumors can be successfully resected via a robotic-assisted, laparoscopic approach.

4. Conclusion

Lymphangiomas rarely develop in the retroperitoneum and are typically asymptomatic. This case demonstrates that retroperitoneal lymphangiomas can be atypical causes of chronic flank pain and can be successfully managed with robotic-assisted, laparoscopic resection.

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Declaration of competing interest

The author(s) declare(s) that there is no conflict of interest regarding the publication of this article.

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