En Bloc Robot-assisted Laparoscopic Partial Cystectomy, Urachal Resection, and Pelvic Lymphadenectomy for Urachal Adenocarcinoma

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Primary adenocarcinomas of the bladder and urachus are extremely rare, accounting for 0.5% to 2.0% of all bladder malignancies. During fetal development, the urachus develops into the median umbilical ligament that stretches from the umbilicus to the bladder. Adenocarcinoma accounts for 90% of all cases of urachal carcinoma. There is no consensus regarding the management of urachal carcinoma. Although the preferred treatment is wide local excision with partial or radical cystectomy, bladder-sparing management is increasing. We report a case of robot-assisted laparoscopic partial cystectomy with en bloc resection of the urachus and bilateral pelvic lymphadenectomy for urachal carcinoma. The robot-assisted laparoscopic approach allowed us to minimize surgical morbidity, postoperative pain, and convalescent time while maintaining the oncologic principle of wide local excision.

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KEY WORDS

Urachal adenocarcinoma • Robotic partial cystectomy • Pelvic lymphadenectomy

Primary adenocarcinomas of the bladder and urachus are extremely rare, accounting for 0.5% to 2.0% of all bladder malignancies.[1] The urachus is a musculofibrous band that extends from the dome of the bladder to the umbilicus. During fetal development, the urachus develops into the median umbilical ligament that stretches from the umbilicus to the bladder. Urachal carcinoma stems from the epithelium of the remnant of this structure, and adenocarcinoma accounts for 90% of all cases.[2] Historically, patients tend to have a poor prognosis, with 5-year survival rates of 11% to 61%.[3] Patients with urachal carcinoma most commonly present with dysuria, hematuria, abdominal pain, or umbilical discharge.

There is no consensus regarding the management of urachal carcinoma. Although the preferred treatment is wide local excision with partial or radical cystectomy,[4-6] bladder-sparing management is increasing because the published reports do not clearly support a survival advantage with more radical extirpation.[7,8]

We report a case of robot-assisted laparoscopic partial cystectomy with en bloc resection of the urachus and bilateral pelvic lymphadenectomy for urachal carcinoma. The robot-assisted laparoscopic approach allowed us to minimize surgical morbidity, postoperative pain, and convalescent time while maintaining the oncologic principle of wide local excision.

Case Report

A 20-year-old black woman presented to her primary care physician with abdominal pain, dysuria, and episodic gross hematuria for 1 month. Her medical history was unremarkable. Physical examination revealed a palpable suprapubic pelvic mass. The rest of her examination, including pelvic and complete lymph node examinations, was unremarkable. Initial laboratory test results were all normal. Computed tomography (CT) scan of her abdomen and pelvis revealed a perivesical mass. Simple enucleation of the perivesical mass was performed at an outside facility. Surgical pathology revealed an infiltrating urachal mucinous adenocarcinoma arising from a villous adenoma with a positive margin. The patient was subsequently referred to us for further evaluation and management. Subsequent positron emission tomography scanning showed no evidence of metastasis. Thus, robot-assisted laparoscopic partial cystectomy, excision of the urachus with bilateral pelvic lymphadenectomy, and excision of the umbilicus was planned.

Surgical pathology revealed an infiltrating urachal mucinous adenocarcinoma arising from a villous adenoma, with a positive margin.

A total of five laparoscopic/robotic ports were used. The camera port was placed in the supraumbilical position. Initial diagnostic laparoscopy revealed no evidence of abdominal metastasis. Extended bilateral pelvic lymphadenectomy was then performed. An en bloc specimen was resected with the dissection extending from just inferior to the umbilicus and encompassing the tissue between the right and left medial umbilical ligaments and the anterior dome of the bladder. The specimen remained attached to the umbilicus while the bladder was closed in two layers. A skin incision was then made around the umbilicus, and the umbilicus and prior dissected specimen were retrieved en bloc. Total surgery time was 300 minutes and intraoperative blood loss was 5 mL. There were no intraoperative or postoperative complications. The patient was discharged home on postoperative day 1 with a Foley catheter in place. Results of a cystogram done 10 days later were normal and the Foley catheter was removed. The final pathology revealed no evidence of residual tumor in the specimen and lymph nodes were free of cancer. No adjuvant therapy was proposed due to complete resection with negative margins and absence of a secondary location.

Discussion

The urachus is an embryonic remnant structure between the dome of the urinary bladder and umbilicus. Urachal carcinoma was first described in 1863 by Hue and Jacquin.[9] Urachal adenocarcinoma accounts for 10% of all primary bladder adenocarcinomas, which are frequently asymptomatic before dissemination.[10] Urachal adenocarcinomas have a predilection for early peritoneal spread and may also have bone, lung, and liver metastases.[11] This reported early tendency to disseminate may actually be a by-product of delayed diagnosis because the majority are asymptomatic.

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The diagnostic evaluation for urachal carcinoma should begin with a careful history and physical examination. Urinalysis with cytology may also be helpful. CT or magnetic resonance imaging scans

of the abdomen and pelvis may also provide information on local extent, lymph node involvement, and metastases. Further metastatic evaluation may be obtained via chest radiograph or bone scanning. Cystoscopy is necessary to evaluate whether the carcinoma has penetrated the bladder urothelium and determine the need for transurethral biopsy. The University of Texas MD Anderson Cancer Center (Houston, TX) has developed practical criteria for the diagnosis of urachal cancer, and Sheldon and colleagues[12] proposed a staging schema for urachal carcinomas.

Urachal masses may be cystic or solid. Mucinous urachal adenocarcinoma is a variant associated with mucin production. Clinically, it may be challenging to differentiate between cystic urachal mucinous adenocarcinoma and benign cystic tumors. Rarely, mucinous urachal adenocarcinomas may be associated with pseudomyxoma peritonei owing to urachal mucocele rupture into the peritoneal cavity.[13] The presence of calcification in the cyst or elevated carcinoembryonic antigen may be indicators of urachal adenocarcinoma.[14] The tumor marker carbohydrate antigen 19-9 has been described as elevated with some urachal carcinomas.[13] In symptomatic cases, presence of hematuria is a strong predictor of malignancy and bladder involvement. Patient age also should be considered in predicting malignancy in urachal masses, as the risk of malignancy increases remarkably after age 55.[8]

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Due to the low incidence of urachal adenocarcinoma, the establishment of standard therapy remains difficult.[11] In nonmetastatic cases, radical resection is the standard of care and the only curative treatment. En bloc resection of the urachus and its peritoneal surrounding, as well as partial cystectomy, are required to ensure clear surgical margins.[11,15] Some authors also recommend umbilectomy in all cases.[8,11] In a series from the Mayo Clinic (Rochester, MN), absence of umbilectomy did not correlate with prognosis and survival in multivariate analysis.[8] If skin umbilicus conservation is chosen, close follow-up is needed. No advantage to radical cystectomy compared with en bloc partial cystectomy with clear margins has been demonstrated.[8,15] The role of intrapelvic and iliac lymphadenectomy has not been clearly defined and has not been routinely recommended.[8] Although a minimally invasive surgical approach for benign urachal pathology has been well described,[16] rarely has laparoscopic resection of urachal adenocarcinoma been reported.[17] A robot-assisted laparoscopic approach for urachal adenocarcinoma of the bladder is feasible, even in challenging cases.[18,19] However, application of oncologic principles and experience with laparoscopic and robotic surgery is mandatory.

The role of adjuvant therapies, either radiotherapy or chemotherapy, has not been clearly evaluated.[8,15] Centers with extensive experience have not demonstrated a survival advantage with adjuvant radiotherapy, chemotherapy, or both.[8] However, some encouraging results have been obtained at the University of Texas MD Anderson Cancer Center with a 5-fluorouracil and cisplatin-based regimen in metastatic cases.[11,20] In cases with pseudomyxoma peritonei, cytoreductive surgery (peritonectomy) associated with perioperative intraperitoneal chemotherapy may be considered.[13]

Global 5-year survival is approximately 49% after urachal adenocarcinoma treatment.[8] Aside from tumor stage, negative surgical margins and low-grade tumors are the prognostic factors that have been shown to positively influence survival.[8,20] Survival rates between 65% and 100% at 5 years have been demonstrated for those with Sheldon stage I and II disease.[8] A Japanese study reported 88% to 100% survival rates without recurrence at 2 years after total cystectomy for patients with disease in Sheldon stage IIIA or less.[15] In contrast, the Mayo Clinic described survival rates near 60% and 0% for those with Sheldon stage III and IV disease (metastatic cases), respectively, at 5 years.[8]

Centers with extensive experience have not demonstrated a survival advantage with adjuvant radiotherapy, chemotherapy, or both. However, some encouraging results have been obtained at the University of Texas MD Anderson Cancer Center with a 5-fluorouracil and cisplatin-based regimen in metastatic cases.

Conclusions

The only curative treatment for urachal adenocarcinoma is surgical resection, which may achieve long-term disease-free survival when detected early. No adjuvant therapies have been demonstrated to be effective. Surgical resection should, at minimum, include complete urachectomy and partial cystectomy to include the bladder dome. This may be achieved via a robot-assisted laparoscopic approach, which minimizes surgical morbidity, postoperative pain, and convalescent time, while maintaining the oncologic principle of wide local excision. ■

References

1. el-Mekresh MM, el-Baz MA, Abol-Enein H, Ghoneim MA. Primary adenocarcinoma of the urinary bladder: a report of 185 cases. *Br J Urol*. 1998;82:206-212.
2. Petersen RO. *Urologic Pathology*. Philadelphia, PA: Lippincott Williams & Wilkins; 1986.
3. Dahm P, Gschwend JE. Malignant non-urothelial neoplasms of the urinary bladder: a review. *Eur Urol*. 2003;44:672-681.
4. Kakizoe T, Matsumoto K, Andoh M, et al. Adenocarcinoma of urachus. Report of 7 cases and review of literature. *Urology*. 1983;21:360-366.
5. Santucci RA, True LD, Lange PH. Is partial cystectomy the treatment of choice for mucinous adenocarcinoma of the urachus? *Urology*. 1997;49:536-540.
6. Burnett AL, Epstein JI, Marshall FF. Adenocarcinoma of urinary bladder: classification and management. *Urology*. 1991;37:315-321.
7. D'Addessi A, Racioppi M, Fanasca A, et al. Adenocarcinoma of the urachus: radical or conservative surgery? A report of a case and a review of the literature. *Eur J Surg Oncol*. 1998;24:131-133.
8. Ashley RA, Inman BA, Sebo TJ, et al. Urachal carcinoma: clinicopathologic features and long-term outcomes of an aggressive malignancy. *Cancer*. 2006;107:712-720.
9. Hue L, Jacquin M. Cancer colloide de la lombille et de paroi abdominale anterieure ayant envahi la vessie. *Union Med de la Siene-Inf Rouen*. 1863;6:418-420.
10. Wright JL, Porter MP, Li CI, et al. Differences in survival among patients with urachal and non-urachal adenocarcinomas of the bladder. *Cancer*. 2006;107:721-728.
11. Siefker-Radtke A. Urachal carcinoma: surgical and chemotherapeutic options. *Expert Rev Anticancer Ther*. 2006;6:1715-1721.
12. Sheldon CA, Clayman RV, Gonzalez R, et al. Malignant urachal lesions. *J Urol*. 1984;131:1-8.
13. Shinohara T, Misawa K, Sano H, et al. Pseudomyxoma peritonei due to mucinous cystadenocarcinoma in situ of the urachus presenting as an inguinal hernia. *Int J Clin Oncol*. 2006;11:416-419.
14. Carr NJ, McLean AD. A mucinous tumour of the urachus: adenoma or low grade mucinous cystic tumour of uncertain malignant potential? *Adv Clin Path*. 2001;5:93-97.
15. Asano K, Miki J, Yamada H, et al. Carcinoma of urachus: report of 15 cases and review of literature—is total cystectomy the treatment of choice for urachal carcinoma? [in Japanese]. *Nihon Hinyokika Gakkai Zasshi*. 2003;94:487-494.
16. Okegawa T, Odagane A, Nutahara K, Higashihara E. Laparoscopic management of urachal remnants in adulthood. *Int J Urol*. 2006;13:1466-1469.
17. Wadhwa P, Kolla SB, Hemal AK. Laparoscopic en bloc partial cystectomy with bilateral pelvic lymphadenectomy for urachal adenocarcinoma. *Urology*. 2006;67:837-843.
18. Spiess PE, Correa JJ. Robotic assisted laparoscopic partial cystectomy and urachal resection for urachal adenocarcinoma. *Int Braz J Urol*. 2009;35:609.
19. Madeb R, Knopf JK, Nicholson C, et al. The use of robotically assisted surgery for treating urachal anomalies. *BJU Int*. 2006;98:838-842.
20. Siefker-Radtke AO, Gee J, Shen Y, et al. Multimodality management of urachal carcinoma: the M.D. Anderson Cancer Center experience. *J Urol*. 2003;169:1295-1298.

MAIN POINTS

- The urachus is a musculofibrous band that extends from the dome of the bladder to the umbilicus. During fetal development, the urachus develops into the median umbilical ligament that stretches from the umbilicus to the bladder. Urachal carcinoma stems from the epithelium of the remnant of this structure, and adenocarcinoma accounts for 90% of all cases, although primary adenocarcinomas of the bladder and urachus are extremely rare—accounting for only 0.5% to 2.0% of all bladder malignancies.
- Due to the low incidence of urachal adenocarcinoma, the establishment of standard therapy remains difficult; although the preferred treatment is wide local excision with partial or radical cystectomy, bladder-sparing management is increasing because the published reports do not clearly support a survival advantage with more radical extirpation.
- Global 5-year survival is approximately 49% after urachal adenocarcinoma treatment. Aside from tumor stage, negative surgical margins and low-grade tumors are the prognostic factors that have been shown to positively influence survival. Survival rates between 65% and 100% at 5 years have been demonstrated for those with Sheldon stage I and II disease.
- The only curative treatment for urachal adenocarcinoma is surgical resection, which should, at minimum, include complete urachectomy and partial cystectomy to include the bladder dome. This may be achieved via a robot-assisted laparoscopic approach, which minimizes surgical morbidity, postoperative pain, and convalescent time, while maintaining the oncologic principle of wide local excision.