CASE REPORT

Urachal adenocarcinoma incidentally discovered in a young woman at elective caesarean section

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SUMMARY

Urachal adenocarcinoma is a rare non-urothelial malignancy that represents only 0.5% of all vesical cancers. Urachal adenocarcinoma most commonly occurs in the 5th to 7th decade. We describe a case of a 35-year-old woman (G3P1) who presented at 20 weeks pregnancy with two episodes of post-coital bleeding. Ultrasound demonstrated a uterine mass measuring 59×43×56 mm, presumed to be a leiomyoma. The decision was made to proceed to caesarean section at 38 weeks, given the progression of the presumed leiomyoma. A 2 cm bladder dome mass was incidentally discovered at the time of the caesarean section and an urgent intraoperative urology consult was sought. Resultantly, the bladder mass was dissected to the dome, and pathology returned urachal adenocarcinoma. This case report considers the treatment decisions involved when faced with urachal adenocarcinoma and reviews the literature surrounding this rare malignancy.

BACKGROUND

Urachal adenocarcinoma is a rare non-urothelial malignancy that represents only 0.5% of all vesical cancers.¹ Urachal adenocarcinoma most commonly occurs in the 5th to 7th decade.² It is an aggressive malignancy that is often diagnosed at an advanced stage and has both a high recurrence rate and poor prognosis.

The urachus is a vestigial tubular structure connecting the embryological allantois to the dome of the urinary bladder during early fetal life.¹ At 12 weeks in utero, the urachus regresses, either to become the median umbilical ligament or to persist as a cyst, sinus or diverticulum.¹ While the pathophysiology of urachal adenocarcinoma is poorly understood, a persistent urachus has a strong association with urachal malignancy in adulthood.³

To date, there have been no randomised control trials on urachal adenocarcinoma, and there are currently <300 cases published on an international scale.⁴

CASE PRESENTATION

We describe a case of a 35-year-old woman (G3P1) who presented in the 20th week of pregnancy with two episodes of post-coital bleeding. Ultrasound demonstrated a uterine mass measuring 59×43×56 mm, presumed to be a leiomyoma (figure 1). This was monitored with further ultrasound at 36 weeks, and showed growth to 75×54×73 mm (figure 2). The patient did not report previous umbilical discharge, dysuria, heavy uterine bleeding or abnormalities with her menstrual cycle.

The decision was made to proceed to caesarean section at 38 weeks, given the progression of the presumed leiomyoma.

A 2 cm bladder dome mass was incidentally discovered at the time of the caesarean section and an urgent intraoperative urology consult was sought. Resultantly, the bladder mass was dissected to the dome and flexible cystoscopy was performed to assess the intravesical component. The pathology returned urachal adenocarcinoma. The patient did not have any previous presentations suspicious for a persistent urachal remnant, the most common documented risk factor for urachal adenocarcinoma.¹

INVESTIGATIONS

Histopathology of the dissected mass demonstrated a sessile polypoid lesion and an attached 75×60×55 mm encapsulated mass of moderately differentiated adenocarcinoma that demonstrated a mucinous pattern with a mucin pool, characteristic of urachal adenocarcinoma (figure 3).

Histologically, the great majority of urachal malignancies are mucin secreting adenomas, comprising mucinous, enteric, mixed and signet ring cell subtypes.¹

There was no evidence of surrounding dysplasia or in situ carcinoma. The neoplastic cells were positive for CD15 and CEA, persuasive for urachal carcinoma. The lesion met multiple criteria for urachal carcinoma as established by Wheeler and Hill.⁵

Staging CT and positron emission tomography scan did not demonstrate evidence of metastatic disease, although note was made of anterior thickening of the urinary bladder wall, likely secondary to resection.

DIFFERENTIAL DIAGNOSIS

- ▶ Leiomyoma
- ▶ Urachal diverticulum

TREATMENT

The patient underwent umbilectomy, urachal remnant excision and pelvic lymph node dissection with bladder dome resection 8 weeks after caesarean section (figure 4). This is the current gold standard of practice for patients with low-risk disease, and is described in the current literature.^{6 7}

Cystectomy was successfully avoided in this patient, due to early resection of the bladder dome and absence of metastatic disease. The risks and benefits of a more conservative surgical approach

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Figure 1 Abdominal ultrasound at 32 weeks demonstrating presumed leiomyoma.

Figure 2 Abdominal ultrasound at 37 weeks demonstrating progression of presumed leiomyoma.

Figure 3 Operative resection of bladder dome mass.

Figure 4 Histopathology demonstrating moderately differentiated invasive adenocarcinoma with extracellular mucin.

were carefully considered given the patient's age and desire to bear further children.

The tissue and lymph nodes excised at this time were found to be benign on histopathological examination. The patient thus did not require adjunct chemotherapy or radiotherapy at the time of surgical resection.

OUTCOME AND FOLLOW-UP

The patient was followed up at 3 and 6 months. A repeat abdominal CT at 6 months demonstrated a poorly characterised liver lesion. This was further investigated with ultrasound, which demonstrated a circumscribed hypoechoic lesion measuring 19×14 mm in segment 4B. The lesion was described as hypoisoechoic and avascular.

For further evaluation, a liver MRI with Primovist was suggested by radiology, considering the history of adenocarcinoma. The lesion was reported as consistent with focal nodular hyperplasia, with no features suggestive of hepatic metastatic disease.

DISCUSSION

There have been no published cases of urachal adenocarcinoma discovered incidentally at the time of caesarean section, or during any elective gynaecological procedure.

Urachal adenocarcinoma usually presents in the fifth to seventh decade as aforementioned, and has been described as an aggressive malignancy with high recurrence rates.^{6,8}

An en bloc resection of the urachal ligament and umbilicus is recommended for patients who have surgically resectable disease, along with either a complete or partial cystectomy, to ensure negative margins.⁴ This form of operative treatment is preferred, as urachal tumours can occur anywhere along the urachus, including at the umbilicus in 7% of patients.⁷ Currently, an open surgical approach is favoured, as there is a paucity of research into laparoscopic or robotic urachal surgeries.

There has been investigation into the role of adjuvant chemotherapy for urachal adenocarcinoma, with uncertain benefits, although currently the combination of 5-fluorouracil and cisplatin is used for metastatic disease.² Urachal adenocarcinoma has a high recurrence rate and long-term survival is associated with negative surgical margins ($p=0.004$) and absence of nodal involvement ($p=0.01$).⁶ Research into chemotherapeutic regimens and other alternatives to surgery for urachal adenocarcinoma is limited by the rare incidence and poor prognosis

secondary to the aggressive nature of the tumour and variability of presenting symptoms.

In a clinical analysis of 14 cases of urachal adenocarcinoma,⁹ the predominant presenting symptoms were haematuria and lower urinary tract symptoms, including dysuria and frequency. Mucousuria and the development of a suprapubic abdominal mass have also been reported in the literature.² Cystoscopy largely revealed broad-based tumours located at the bladder dome, but provided an unsatisfactory assessment of extension and local spread. Eleven of the 14 patients were treated operatively with either partial or total cystectomy and three with advanced metastatic disease were treated with chemotherapy, two of whom achieved progression-free survival for 7 and 8 months, respectively.⁹

This demonstrates a need for further research into treatment of patients presenting with advanced disease who are not suitable for operative intervention.

Zong and Chen¹⁰ described treatment of recurrent urachal adenocarcinoma in a 41-year-old woman, using a chemotherapy regimen tailored to immunohistochemical testing. While tumour markers were responsive to the chemotherapeutic regimen, the patient ultimately died of the metastatic disease.² Personalised chemotherapy may be a viable option in those patients who do not respond to initial therapy.

Learning points

- Early urology consult for bladder dome masses found on imaging or at the time of operation is essential.
- Differential diagnoses of suprapubic masses on examination and imaging should be widely considered.
- Decisions for definitive treatment of urachal adenocarcinoma should be based on both, tumour grade and individual patient factors such as age, risks of surgical intervention and comorbidities.

In this case of a young, previously well woman with a 3-year-old as well as a new-born child after caesarean section, an urgent intraoperative urology consult resulted in early diagnosis and thus treatment of a rare and highly aggressive cancer that was otherwise largely asymptomatic.

This case is an interesting culmination of obstetric, gynaecological and urological presentations, and encourages clinicians to think carefully about differential diagnoses of masses discovered during neonatal ultrasound.

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REFERENCES

- 1 Paner GP, Barkan GA, Mehta V, *et al.* Urachal carcinomas of the nonglandular type: salient features and considerations in pathologic diagnosis. *Am J Surg Pathol* 2012;36:432–42.
- 2 Yu JS, Kim KW, Lee HJ, *et al.* Urachal remnant diseases: spectrum of CT and US findings. *Radiographics* 2008;21:451–61.
- 3 Ashley RA, Inman BA, Routh JC, *et al.* Urachal anomalies: a longitudinal study of urachal remnants in children and adults. *J Urol* 2007;178:1615–18.
- 4 Elser C, Sweet J, Cheran SK, *et al.* A case of metastatic urachal carcinoma treated with several different chemotherapeutic regimens. *Can Urol Assoc J* 2012;6:27–31.
- 5 Wheeler JD, Hill WT. Adenocarcinoma involving the urinary bladder. *Cancer* 1954;7:119–35.
- 6 Siefker-Radtke A. Urachal adenocarcinoma: a clinician's guide for treatment. *Semin Oncol* 2012;39:619–24.
- 7 Siefker-Radtke AO, Gee J, Shen Y, *et al.* Multimodality management of urachal carcinoma: the M.D Anderson Cancer Center experience. *J Urol* 2003;169:1295–8.
- 8 Tsiouris A, Ahmed HU, Kumar N, *et al.* Urachal tumour: clinical and radiological features of a poorly understood carcinoma. *Ann R Coll Surg Engl* 2007;89:17–18.
- 9 Chen ZF, Wang F, Qin ZK, *et al.* Clinical analysis of 14 cases of urachal carcinoma. *Ai Zheng* 2008;27:966–9.
- 10 Zong L, Chen P. Surgical and chemotherapeutic experience regarding a urachal adenocarcinoma with repeated relapse: case report and literature review. *World J Surg Oncol* 2013;1:170–6.

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