CASE REPORT

Urachal Mucinous Cystadenocarcinoma

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SUMMARY

A 57-year-old man presented with a 6-month history of pelvic fullness. He had no lower urinary tract symptoms or altered bowel habits. On examination, there was a non-tender pelvic mass which extended from the pubic symphysis to the level of the umbilicus. CT scan of the abdomen demonstrated a 22×11×11 cm cystic mass arising from the pelvis extending into the midline and superiorly to the umbilicus. Other than raised carcinoembryonic antigen of 7.6 ng/mL (<5.0), the remainder of his blood test were unremarkable. Flexible cystoscopy demonstrated a convex deformity of the bladder wall in keeping with the compression and displacement as seen on the CT. The patient underwent an open excision of the cystic structure (urachal remnant), partial cystectomy, partial excision of anterior abdominal wall and pelvic lymphadenectomy. A check cystogram performed 12 days following the initial operation was unremarkable.

22×11×11 cm cystic mass arising from the pelvis, extending into the midline and superiorly to the umbilicus. The lesion encased the anterior surface and dome of the bladder. The cystic lesion did not enhance on contrast administration. There were no other abdominal abnormalities on CT. Other than raised carcinoembryonic antigen (CEA) of 7.6 ng/mL (<5.0), the remainder of his blood tests were unremarkable; carbohydrate antigen (CA 19.9) of 23 U/mL (<35), creatinine: 1.03 mg/dL, eGFR: 80 mL/min. Three urine samples were sent for cytology, all of which were negative for malignant cells. A flexible cystoscopy was subsequently performed. The mucosa had a normal appearance and there was no involvement of the ureteral openings. No fistulas were visualised communicating with the cystic structure in the bladder dome or anterior wall. The right side of the bladder had a convex deformity of the bladder wall in keeping with the compression and displacement as seen on the CT.

BACKGROUND

The urachus, is a midline tubular structure that extends superiorly from the anterior dome of the bladder, to the umbilicus and runs within the umbilical cord.¹ During embryonic evolution, the urachus obliterates to form the median umbilical ligament. However, in approximately a third of adults, the urachus is persistent.^{1,2} A patent urachus may become the primary site of lesions, including cysts, fistulae and tumours, both benign and malignant.¹ We present the case of 57-year-old man with histologically confirmed massive urachal mucinous cystadenocarcinoma.

CASE PRESENTATION

A 57-year-old man presented with 6-month history of pelvic and abdominal fullness. He had no lower urinary symptoms, haematuria or altered bowel habits. His weight had been stable over the past 12 months and he denied any fevers or night sweats. Of note, he was a smoker of >30 pack per year and with history of chronic obstructive pulmonary disease, previous laryngeal cancer, atrial fibrillation, unprovoked deep vein thrombosis and pulmonary emboli. He was medicated with amiodarone and apixaban. On examination, there was a non-tender pelvic mass which extended from the pubic symphysis to the level of the umbilicus being widest inferiorly. When attempting to displace the mass laterally, it felt adherent to the anterior abdominal wall in the midline.

A contrast enhanced CT scan of the abdomen and pelvis was arranged. The CT scan demonstrated a

INVESTIGATIONS

Included in above case report.

TREATMENT

The patient underwent open excision of the urachal cyst, partial cystectomy, partial excision of anterior abdominal wall and pelvic lymphadenectomy. A midline incision extending from the xiphisternum to the umbilicus was performed. The urachal remnant was excised en bloc with the umbilicus and a 4-cm-wide portion of the anterior abdominal wall (figure 1). Within the pelvis, the cyst was densely adherent to the pubis and right superior pubic rami, requiring a challenging period of adhesiolysis and dissection. The cyst was also densely adherent to the anterior bladder. There was no gross involvement of the distal ureter's bilaterally. Cystotomy at the posterior bladder dome and en-bloc resection incorporating anterior bladder wall was performed, cautious to preserve as much bladder volume as oncologically safe (figure 2). The bladder defect was closed in two layers, with no leak seen on bladder distention. The external iliac lymph nodes on the right were palpable and an intraoperative decision was made to proceed with a bilateral extended pelvic lymphadenectomy, including the external iliac nodes extending to the aortic bifurcation, hypogastric nodes deep to the obturator nerve and obturator packet. Surgical specimens were sent for histology. The abdominal wall resection, due to the adherent mass, subsequently made abdominal wall closure tight despite closure with continuous and interrupted figure of eight sutures.

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Figure 1 Urachal remnant excised en bloc with umbilicus and a 4 cm wide portion of anterior abdominal wall.

Two days postoperatively, the patient reported a tearing sensation through the midline abdominal wound. On examination, the abdomen was expectedly distended however there was no midline hernia or defect in the closure palpable with cough impulse. An ultrasound scan of the abdomen demonstrated an apposed posterior rectus sheath. He was managed with bed rest. On day 4, the patient developed abdominal distention and vomiting, clinically consistent with small bowel obstruction. During an episode of vomiting, there was complete wound dehiscence and abdominal evisceration. The patient underwent laparotomy. Intraoperatively, it was evident some of the fascial sutures had cut through tissue compromising the whole length of the fascial closure.

A thorough inspection and copious warm saline wash of the bowel did not identify any injuries or areas of concern. Wide mobilisation of the fascia of the anterior abdominal wall was performed in order to facilitate large fascial suture bites on closure with multiple interrupted figure of eight 0-Polydioxanone sutures (PDS) along with continuous 0-loop PDS sutures. Day 4 following the relook laparotomy, the patient was tolerating a full diet and had opened his bowels. A check cystogram performed 12 days following the initial operation was unremarkable and the urethral catheter was subsequently removed. The patient was discharged home the following day on simple analgesia without further issues.

OUTCOME AND FOLLOW-UP

The en-bloc specimen, including the cystic contents, weighed 1.8 kg. The overall cyst with attached soft tissues measured 142 mm

Figure 2 Cystotomy at posterior bladder dome.

(superior-inferior) x 185 mm (anteroposterior) x 115 mm (right to left) (figure 3). The cyst itself had a wall thickness of 2–3 mm. No haemorrhage, papillae or solid components were identified macroscopically. Microscopically, features were consistent with urachal mucinous cystadenocarcinoma with 8 mm invasion of bladder muscularis propria. All surgical margins were clear and there was no evidence of metastatic disease to the iliac nodes. Reviewed 2 months postoperatively, the patient is well and has returned to full work duties.

Figure 3 En-bloc resected specimen.

DISCUSSION

Given the rarity of urachal neoplasms, the exact incidence is unclear but estimated to account for less than 1% of all bladder cancers.^{2,3} Interestingly, urachal carcinomas are predominantly adenocarcinomas, lined with mucus-secreting epithelial cells despite the normal urachus lining of transitional epithelium.²

In 1985, Johnson *et al* developed a modified the criteria for establishing the diagnosis urachal carcinoma: (1) tumour in the bladder dome or midline, (2) demarcation between the tumour and normal surface epithelium, and (3) absence of primary adenocarcinoma of another organ.⁴ Certainly, on imaging, and later with histological confirmation, the case we have presented satisfies this definition.

Consistent with other enteric-type adenocarcinomas, urachal adenocarcinoma may have detectable serum levels of CEA and carbohydrate antigen 19–9 (CA 19–9).^{3,5} Siefker-Radtke *et al* report 13 of 22 patients in their series with elevated CEA (range 7–326 ng/mL) and 6 of 10 patients with an elevated CA 19–9 (range 43–11 861 U/mL) when tested.³ The case presented had a mildly raised CEA of 7.6 ng/mL however normal CA of 19.9. In patients with raised values, tumour markers may provide valuable means of monitoring response to therapy and surveillance for disease recurrence.³

Since metachronous or synchronous carcinoma can occur along the urachal tract, urachectomy with en-bloc resection of the umbilicus, anterior abdominal wall, partial cystectomy and pelvic lymph nodes dissection where indicated is advised.² Although radical, this surgical approach is supported by the histological analysis of surgical specimens by Siefker-Radtke in which no patient had disease confined to the epithelium or urachal ligament; 27 of 32 patients had locally advanced disease that directly invaded either the bladder, peritoneum abdominal wall or local viscera with the remaining five cases having nodal involvement at surgery, supporting selective use of lymph node dissection.³ It is important to differentiate urachal adenocarcinoma from non-urachal adenocarcinoma of the bladder, as different treatment strategies need be employed. Non-urachal primary adenocarcinoma of the bladder responds poorly to partial cystectomy and this is best evaluated with a preoperative cystoscopy.⁶

No standard chemotherapy regimen for advanced urachal carcinoma has been established and The National Comprehensive Cancer Network states that conventional chemotherapy for urothelial carcinoma is not effective.⁷ They do however state the

use of chemotherapy or radiotherapy should be individualised and may be of potential benefit in select patients.⁷ It is our view that consideration should be given to those with node positive disease. As our patient had a lymph node dissection clear of any metastatic disease with clear surgical margins, we have not planned for adjuvant chemotherapy.

Learning points

- ▶ Consistent with other enteric-type adenocarcinomas, urachal adenocarcinoma may have detectable serum levels of carcinoembryonic antigen and carbohydrate antigen 19–9 (CA 19–9).
- ▶ Urachectomy with en-bloc resection of the umbilicus, anterior abdominal wall, partial cystectomy, and selective lymph nodes dissection is advised for patients with local disease when feasible.
- ▶ Adjuvant therapy should be considered for patients with node positive disease.

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