

An unusual case of cancer of the urachal remnant following repair of bladder exstrophy

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Abstract

Introduction We report the first case of cancer of the urachal remnant following repair of bladder exstrophy, in a renal transplant recipient.

Method A retrospective review of this clinical case and the associated literature were performed.

Conclusion This unusual case highlights two very rare entities. Bladder exstrophy has an incidence of 1 in 50,000 newborns, whereas urachal cancer accounts for less than 1% of all bladder tumours.

Keywords Urachal remnant · Bladder exstrophy

Case report

We report the case of a 49-year-old female, presenting with a 6-month history of vaginal discharge, intermittent per

vagina bleeding and a sensation of suprapubic pressure. She had a background of bladder exstrophy with associated diastasis of the rectus abdominis muscles and pubic symphysis, for which she underwent a cystectomy and ureterosigmoidostomy as an infant. Subsequently, she developed bilateral staghorn calculi, chronic renal impairment and a right-sided pyonephrosis, necessitating a right nephrectomy. At the age of 31, she was commenced on haemodialysis for end stage renal failure. After 2 years, in preparation for renal transplantation, she underwent resection of the ureterosigmoidostomy, ileal conduit formation and left nephrectomy. The same year she received a renal cadaveric transplant to the left iliac fossa, which was anastomosed to an ileal loop. There was immediate graft function. Medications on admission included cyclosporin, azathioprine, dexamethasone, folic acid and aspirin.

Bimanual pelvic examination revealed right adnexal and uterine masses. Graft function was maintained at baseline (urea 11, creatinine 130). Ultrasound of the transplant kidney showed preservation of the renal cortex with no evidence of hydronephrosis. MRI of the pelvis showed a right adnexal cystic mass and a second irregular mass extending from the left lateral aspect of the lower uterine wall (Fig. 1). Trans-vaginal ultrasound guided biopsies of the pelvic mass and the vaginal wall were taken. Histopathology revealed a papillary, villoglandular proliferation with colonic type metaplasia. The surface epithelial cells showed carcinoma in situ. Possible primary sites included an urachal remnant, the female genital tract or the colon. Carcinoembryonic antigen (CEA) measurement and full colonoscopy with random biopsies were normal. Dynamic CT of thorax and abdomen with oral contrast revealed no evidence of distant metastases.

An anterior pelvic exenteration was performed. Intraoperatively, a mass densely adherent to the left wall of the

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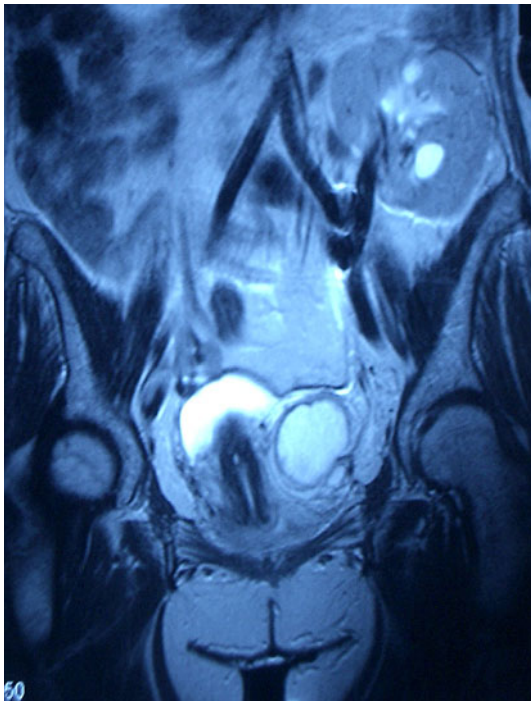


Fig. 1 MRI pelvis: 4 × 4 × 5 cm irregular mass extending from the left lateral aspect of the lower uterine wall towards pelvic sidewall. Left sided transplant kidney is evident

vagina, left iliac vessels and left sidewall of the pelvis was identified. Additionally, a large right ovarian cyst, adherent to the anus and rectum was seen. Following extensive resection, residual tumour remained along the iliac vessels and pelvic sidewall. A loop sigmoid colostomy was formed. The transplant kidney and ureter were preserved, including the ileal conduit.

Pathology of the right ovarian mass revealed a benign serous cystadenoma. An irregular fungating 5 × 5 × 5 cm mass was attached to the left inferior aspect of the uterus. Histological examination of the mass revealed the presence of a tubular structure with a muscular wall, lined by a well-differentiated adenocarcinoma with villoglandular papillary growth pattern. The tumour cells show enteric differentiation with goblet and paneth cell metaplasia (Fig. 2). Immunohistochemical staining showed the tumour to be positive for CK7, CK20 and CEA; and negative for oestrogen and progesterone receptors.

Features were consistent with a well-differentiated adenocarcinoma of the urachal remnant, present focally at the surgical resection margin. There was no tumour identified in the lymph nodes. The left ovary and fallopian tubes were unremarkable.

The patient received adjuvant chemoradiotherapy. Surveillance CT of the thorax, abdomen and pelvis at long-term follow up shows no evidence of recurrence or metastases.

Discussion

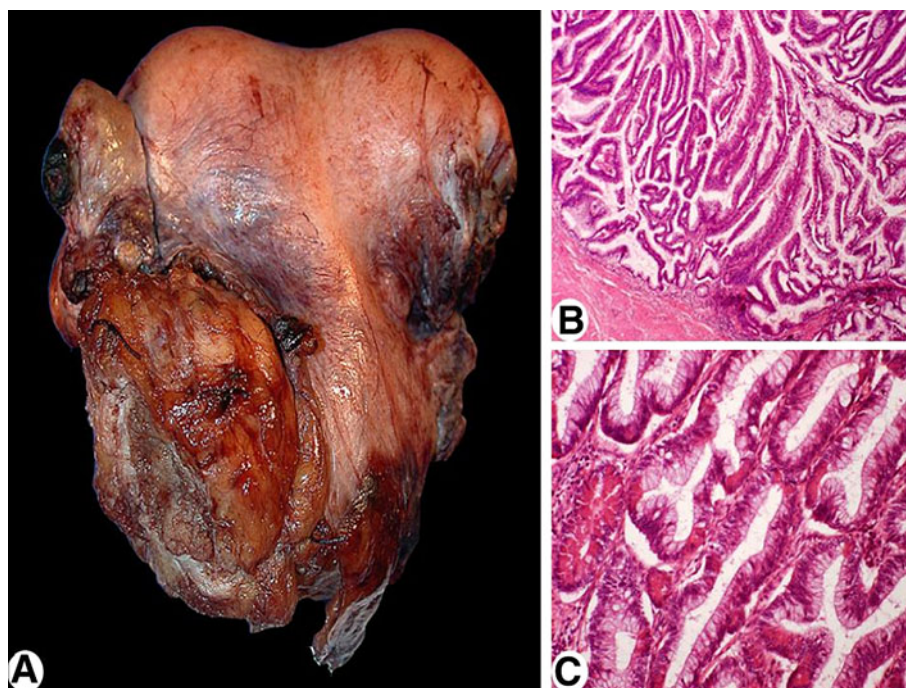
We report the first case of cancer of the urachal remnant following repair of bladder exstrophy in a renal transplant recipient. Our patient presented, aged 49 years, with a well-differentiated mucin-positive colonic type adenocarcinoma of the urachal remnant. She was 16 years post-cadaveric renal transplantation and on standard immunosuppressive regimen. Post-renal transplantation, cancer is a well-recognised complication with an overall incidence of malignancy being reported as approximately three times higher than the general population [1]. This unusual case highlights two very rare entities. Bladder exstrophy, also known as classic exstrophy or vesica ectopiae has an incidence of 1 in 50,000 newborns [2], whereas urachal cancer accounts for less than 1% of all bladder tumours [3].

Bladder exstrophy is a congenital anomaly confined to the foetal genitourinary tract. Embryologically, the bladder develops from the ventral cloaca as a three-layered structure: inner transitional layer, submucosal tissue layer and an outer smooth muscle layer. The foetal bladder descends caudally during gestation followed by a progressive narrowing. This continues until the urachus obliterates prior to birth as a fibromuscular band [4]. It is related to the umbilical arteries through the umbilicovesical fascia in a pyramidal arrangement. In the term foetus, it narrows to a fibrous band called the median umbilical ligament, extending from the umbilicus to the superior aspect of the dome of the bladder. The mechanism of bladder exstrophy formation is not well understood. It is proposed that an abnormally large cloacal membrane may account for the development of a wedge effect on the foetal abdominal wall. If following the caudal descent of the urorectal septum and fusion of the genital tubercles, the cloacal membrane ruptures classic bladder exstrophy results.

The male to female ratio for bladder exstrophy, derived from multiple studies is 2.3–1 [5]. Bladder exstrophy occurs more frequently in first born children and is more common in white infants [2]. Bladder exstrophy is associated with an increased incidence of primary adenocarcinoma of the bladder [6]. There are rare case reports showing squamous cell carcinoma of the bladder associated with exstrophy [6, 7]. Modern treatment of this condition requires a staged approach. The primary aim is to close the bladder/abdominal wall defect. Secondary aims include the establishment of urinary continence, preservation of renal function and provision of aesthetically acceptable external genitalia [5].

Urachal tumours were first described in 1863 by Hue and Jaequin, and account for 20–40% of bladder adenocarcinoma [3, 8]. Urachal cancers are primarily adenocarcinoma (89.7%); 63.6% are classified as mucin-positive, 29.7% as mucin-negative, 4.5% as containing

Fig. 2 Histology: **a** Bi-cornuate uterus with an attached urachal adenocarcinomatous mass. **b** Haematoxylin and eosin section of the urachal adenocarcinoma (original magnification $\times 200$). **c** The tumour cells show enteric differentiation with goblet and paneth cell metaplasia (original magnification $\times 400$)



elements of transitional cell carcinoma (TCC) and 2.2% as containing elements of TCC and small cell carcinoma [8]. Mucin-positive adenocarcinoma is sub-classified as colonic type (commonest), colloid type, signet-ring cell type and a mixed type. Adenocarcinoma of the urachal remnant is the most common malignancy associated with an urachal cyst and is often found at the apex of the bladder [9]. The standard treatment for localised urachal cancer is partial cystectomy, umbilicotomy and resection of the urachus. Chemotherapy is advocated for advanced or metastatic urachal cancers. Radiotherapy may be used for pelvic disease and palliation. The 5-year survival rate for localised disease which underwent surgery is 43% [8, 10]. Overall survival for distant metastatic disease is less than 1 year [8].

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