A Rare Case of Recurrent Urachal Adenocarcinoma of the Bladder

Abstract:

JF Sullivan, DM Fanning, I Cheema, T Creagh
Department of Urology and Transplantation, Beaumont Hospital, Beaumont, Dublin 9

Abstract

Urachal carcinoma is a rare, aggressive malignancy accounting for less than 1% of bladder neoplasms. These tumours are usually adenocarcinomas and occur at the dome or anterior wall of the bladder. They often escape early clinical detection, growing for prolonged periods prior to diagnosis, resulting in local invasion and systemic spread before therapeutic intervention is initiated. We present the case of a recurrent urachal carcinoma in a young female.

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Case Report
A 35 year old female presented complaining of 3 episodes of visible haematuria. She had a prior diagnosis of polycystic ovarian syndrome, no past surgical history and a 20-pack year history of smoking. Abdominal examination was unremarkable. Routine bloods tests were within normal limits. Urinalysis revealed micro-haematuria. Renal tract ultrasound was unremarkable. Cystoscopy revealed a solid lesion at the bladder dome, biopsies of which showed invasive moderately differentiated adenocarcinoma. Pathologically the neoplasm was seen to be coming up from beneath the urothelial surface with an abrupt change to an ulcerated region at the surface. Immunohistochemistry ruled out metastatic carcinoma of colonic origin. A contrast enhanced CT showed focal thickening of the anterior wall of the bladder (Figure 1).

Figure 1: pre-surgical contrast enhanced CT scan showing focal thickening of anterior wall of the bladder.

Figure 2: contrast enhanced CT scan showing local recurrence in anterior wall of the bladder post partial cystectomy

There was no lymphadenopathy or evidence of metastasis. A diagnosis of urachal cancer was made. She underwent a partial cystectomy and urachectomy. A 3cm solid lesion was evident at the bladder dome. Histology showed adenocarcinoma, with lymphovascular invasion and no evidence of disease at the margins (pT3b N0 M0 / IIIa Sheldon classification). She proceeded to receive 9 cycles of adjuvant chemotherapy (FOLFOX) and was followed up on a 3 monthly basis with surveillance cystoscopy and CT. Twenty months post-operatively a small area of local recurrence in the anterior abdominal wall was observed (Figure 2). Cystoscopy showed a recurrent bladder lesion. She proceeded to have an anterior pelvic extenteration with ileal conduit diversion. Pathology again reported a poorly differentiated, muscle invasive adenocarcinoma (pT3b N0 M0 / IIIa Sheldon classification). Four months post-operatively there are no signs of local recurrence or distant metastases.

Discussion
Urachal cancers are rare and aggressive malignancies[1] and due to the lack of prospective studies there is a paucity of information on best management. Patients often present with higher stage cancers at diagnosis because disease arises outside the bladder where it is asymptomatic. The criteria for diagnosis include a midline tumour; sharp demarcation between tumour and normal epithelium; enteric histology; absence of urothelial dysplasia/cystitis glandularis; and absence of a primary adenocarcinoma of another origin[2]. Currently the consensus is that en bloc resection of the urachal ligament and umbilicus is best practice with complete or partial cystectomy and bilateral lymphadenectomy[2]. Local recurrence has been reported to occur in 15-18% in the first 2 years[3,4]. The majority recur in the bladder and urachal remnant, as was the case in this patient, with a smaller proportion occurring elsewhere in the pelvis[5].

Tumour stage, grade and margin status are the most important factors predictive of recurrence and subsequent survival[3]. Currently there is no standard adjuvant chemotherapeutic regimen[1]. In this setting chemotherapy is usually offered to patients who wish to take an aggressive approach to their cancer, have a high likelihood of relapse (positive margins, node involvement) or when the umbilicus was not resected en bloc with the urachal ligament and bladder[6]. Histopathologically, urachal and gastrointestinal adenocarcinomas are similar and as such, colon cancer-specific FOLFOX- chemotherapy has been used in the adjuvant setting with moderate success[7,8].

Collected series of surgically treated urachal cancers show that nearly half of patients die from their disease[2-4]. Importantly however, not all had routinely undergone a complete urachectomy at time of surgery. Herr et al[3] retrospectively analysed a cohort of patients with localised urachal carcinoma. They reported en bloc resection of the tumour, urachus and an extended partial cystectomy cured 70% of patients with clinically localised disease and more than 80% with tumour confined to the surgical specimen. In conclusion, urachal carcinoma is a rare entity with a poor prognosis. Surgery remains the mainstay of therapy with the role of adjuvant therapies uncertain. The achievement of a complete urachectomy, umbilectomy and negative surgical margins, is critical to long-term survival. For recurrent non-metastatic disease, salvage resection is accepted as the most effective treatment and has been shown to result in prolonged survival4.

Correspondence: J Sullivan
Department of Urology and Transplantation, Beaumont Hospital, Beaumont, Dublin 9
Email: johnsullivan@rcsi.ie

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