Stage 0 mucinous adenocarcinoma in situ of the urachus

A B Paul, C R Hunt, J M Harney, J P R Jenkins, R F T McMahon

Abstract
Adenocarcinomas of the urinary bladder are rare (1–5% of bladder tumours) and of notoriously poor prognosis. About one third of such tumours arise in urachal remnants related to the bladder. This is believed to be the first report of in situ change in the urachal remnant. The patient presented with mucusuria and computed tomography showed a typical urachal cyst. After excision the cyst was found to contain mucinous adenomatous epithelium but without invasion of the basal lamina. Pathological stage is the best prognostic indicator in urachal tumours. Prompt investigation and management of mucusuria may allow the diagnosis of urachal tumours in this preinvasive stage.

Keywords: urachus; adenocarcinoma; carcinoma in situ

Adenocarcinoma of the urachal remnant accounts for 0.17–0.34% of all bladder malignancies and 20–39% of primary adenocarcinomas of the bladder.1 Adenomas of the urachus are even rarer.2 We describe a case of urachal mucinous adenocarcinoma in situ presenting at a preinvasive stage.

Case report
A 68 year old man presented with haematuria and mucusuria. He had no other abdominal symptoms. Abdominal and digital rectal examinations were unrevealing and sigmoidoscopy and barium enema showed an normal large bowel without evidence of colovesical fistula. At cystoscopy, a 1.5 cm diameter raised lesion of the dome of the bladder was seen. Mucus was discharging into the bladder lumen from a central punctum. Histopathological examination of biopsies from the lesion showed mucus secreting glandular epithelium, consistent with a patent urachus. The epithelium showed mild atypia. Subsequent computed tomography showed a 3 cm lesion anterior to the bladder dome. The lesion was indenting the bladder dome and contained low attenuation material consistent with mucus (fig 1). The lining of the cyst and the epithelium covering the papillary lesion consisted of mucus secreting cells showing nuclear pleomorphism and abundant mitoses. There was no evidence of invasion through the basal lamina (fig 3). The appearances amounted to those of a mucinous adenocarcinoma in situ. The remainder of the specimen was minutely examined for other areas of neoplastic change, but none were found and no other areas of epithelialised patent urachus were identified.

The patient made an uneventful recovery from surgery and is clinical and ultrasonographically free of recurrence after one year.

Discussion
Mucusuria occurs in around 25% of patients with urachal tumours. It is a rare urological symptom and always merits full investigation with cystoscopy and intravenous urogram, and commonly, as here, computed tomography. Computed tomography is the most sensitive imaging mode in the investigation of urachal tumours.3 As this case shows, the urgent investigation of the symptom of mucusuria may allow the diagnosis of tumour to be made when surgical cure is still possible.

Tumours arising within the urachus are divided into benign (adenoma) or malignant
lesions (adenocarcinoma). Most adenomas of the urachus are multiloculated cystic tumours which are lined by mucus secreting epithelium resembling that of the colon or rectum. Mucin producing adenocarcinomas represent 69% of malignant urachal neoplasms. This patient presented with a non-invasive stage 0 lesion with histological features intermediate between these two categories.

Grignon et al have reported their experience of 24 urachal neoplasms among a series of 72 primary vesical adenocarcinomas. They found a survival rate of 61% at five years and 46% at 10 years. In 41 patients with urachal adenocarcinomas, Nakanishi et al have examined the prognostic significance of ploidy, Ki-67 staining, and nucleolar organising region counts. They found in multivariate analysis that only differentiation and stage were useful prognostic factors. A urachal adenocarcinoma arising in a pre-existing adenoma has previously been reported.

As far as we are aware, this is the first report of a urachal tumour at the stage of carcinoma in situ. We raise the possibility that, as in colonic adenocarcinomas, there is a biological continuum of behaviour of glandular neoplasms in the urachus from benign to malignant. The lesion described here represents the premalignant phase of that continuum. The finding here of premalignant change in a urachal remnant stresses the importance of a wide excision of the urachus in its entirety, in the management of urachal tumours.

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