CASE REPORT

Complex mucinous cystadenoma of undetermined malignant potential of the urachus: a rare case with review of the literature

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Abstract

Urachal carcinoma is an uncommon neoplasm. Benign urachal tumours are extremely rare. All urachal mucinous tumours, regardless of tumour type, have the potential to behave in an aggressive clinical manner that includes the development of pseudomyxoma peritonei. We report a 58-year-old man who presented with lower abdominal pain. Ultrasound and CT imaging defined a large lobulated cystic mass superior to the urinary bladder. At laparotomy, the multiloculated cystic mass, 10 x 8 x 6 cm, could be completely separated from the bladder and was excised. It was smooth-walled and filled with thick mucinous fluid. Histology revealed a complex mucinous cystadenoma of undetermined malignant potential of the urachus. The cystic spaces were lined by mucin-secreting columnar epithelium that showed focal areas of mild atypia. The epithelium lacked architectural features of villous adenoma. There was no stromal invasion to support an invasive neoplastic process. No pseudomyxoma peritonei was present. The patient was well, with no tumour recurrence, at 6 months follow-up.

Key words: mucinous cystadenoma, urachal tumor, urachal carcinoma.

INTRODUCTION

The urachus, or the median umbilical ligament, is an embryological remnant of the allantois. Between the fourth and fifth months of development, the urachus narrows to become a fibromuscular strand, extending from the apex of the bladder to the umbilicus. The urachus lies in the extraperitoneal space of Retzius between the transversalis fascia anteriorly and the peritoneum posteriorly.1

Urachal carcinoma is an uncommon neoplasm. The incidence of urachal cancer ranges from 0.07% to 0.34% of all bladder tumours. However, urachal adenocarcinoma accounts for up to 36% of all reported bladder adenocarcinoma.2 Benign urachal tumors are extremely rare. Urachal villous adenomas, which may coexist with in-situ and invasive adenocarcinoma, is even more rare than urachal adenocarcinomas.3 All urachal mucinous tumours, regardless of tumor type, have the potential to behave in an aggressive clinical manner that includes the development of pseudomyxoma peritonei.4 However, very few cases of mucinous cystadenoma of the urachus have been reported in literature to date. We document a rare case of complex mucinous cystadenoma of the urachus in a 58-year-old male patient.

CASE REPORT

A 58-year-old man attended the urology outpatient department of KLES’s Dr. Prabhakar Kore Hospital & MRC with complaints of pain in the lower abdomen for 2 months. He gave no history of irritative urinary symptoms, gross hematuria, fever and umbilical soreness. He had no recent weight loss.

Clinical findings
On examination, a mass was palpable in the suprapubic region, measuring approximately 7x6 cm, oval in shape, smooth in surface, firm in consistency, non-tender, freely movable laterally but could not be pushed inferiorly.

Imaging findings
Ultrasound (USG) examination of the abdomen showed a thin-walled multicystic lesion in
the suprapubic region, measuring 8.3x7.8x6 cm with a volume of 208 ml. The wall of the lesion was not differentiable from the bladder wall dome. A computed tomography (CT) scan of the abdomen (Fig. 1) revealed a large lobulated cystic lesion with internal septations and calcifications in the lower abdomen superior to the bladder (causing mild indentation on the bladder), measuring approximately 7.7 x 7.2 x 6.4 cm, with minimal peripheral enhancement on contrast study. There was no clinical or radiological evidence of gastrointestinal, bladder or prostatic malignancy. A diagnosis of possible primary urachal carcinoma was put forth.

Operative findings
The patient underwent resection of the cystic mass. At laparotomy, the cystic mass could be completely separated from the bladder and excised. There was no evidence of pseudomyxoma peritonei.

Pathological findings
Gross examination revealed a multiloculated cystic mass measuring 10 x 8 x 6 cm, with smooth walled and filled with thick mucinous fluid (Fig. 2). Histological examination revealed a complex mucinous cystadenoma of the urachus. The cystic spaces were lined by mucin secreting columnar epithelia that showed focal areas of mild atypia, and filled with mucin. The mucin in the lumen of the cysts were devoid of any free floating aggregates of atypical epithelial cells, which is a feature of mucinous adenocarcinoma. There was no evidence of frank stromal invasion (Fig. 3).

There is no recurrence of the mass or any other complaints at 6 months of follow up. There was no evidence of pseudomyxoma peritonei.

DISCUSSION
The urachus, extending from the umbilicus to the urinary bladder apex, persists uncommonly in adults. Histologically, the urachus is a three layered structure, the innermost layer being lined with transitional epithelium in 70% of cases and with columnar epithelium in 30%, a middle submucosal layer of connective tissue, and an outermost muscular layer in continuum with the detrusor muscle.5

Both non-neoplastic and neoplastic conditions constitute the differential diagnosis of a urachal mass in an adult.6 Urachal tract remnants that abnormally remain patent are often subject to infection, resulting in an infected cyst, discharging sinus or fistula (umbilical-urachal sinus or vesico-urachal).5

Benign urachal neoplasms including adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas, are extremely rare. However, they are important in that they mimic urachal malignancy.5 Urachal mucinous tumour of uncertain malignant potential is extremely rare and is characterized by a multilocular cyst

FIG. 1: CT scan of the pelvis demonstrating a large lobulated cystic mass with internal septations and calcifications (arrow) in the lower abdomen superior to the bladder.

FIG. 2: Multiloculated cystic mass filled with thick mucinous fluid.

FIG. 3: Cysts lined by mucin secreting columnar epithelium. No stromal invasion was demonstrated. H&E X100. Inset: Higher magnification of columnar epithelium. H&E X 400.
showing the proliferation of atypical mucin-secreting cells, but without evidence of stromal invasion. As in ovarian and appendiceal borderline tumours, it represents a transitional stage of mucinous carcinogenesis in the urachus.\(^7\)

Literature search revealed only seven cases of urachal mucinous tumour of uncertain malignant potential,\(^6-12\) of which 3 cases were associated with pseudomyxoma peritonei. Males were affected in 5 cases while females in 2 cases.\(^6,7\) The patients were between 50-72 years of age, except in one case\(^7\) which occurred in a 29-year-old female. The present case would be the 8th case, occurring in a male aged 58 years.

Malignant urachal neoplasms are also rare and consist of mucin-producing adenocarcinoma in 69\%, mucin-negative adenocarcinoma in 15\%, sarcoma in 8\%, and transitional cell carcinoma in 3\% of cases.\(^2\) Metastatic disease from primary breast, colonic, ovarian, prostatic malignancies should be excluded clinically as they can mimic urachal mucinous tumours.\(^6\)

Urachal carcinoma should be distinguished from primary bladder cancers that occur in the dome of the urinary bladder. The criteria for this distinction were first proposed by Wheeler and Hill in 1954 and include, location in the dome of the bladder, absence of cystitis cystica or cystitis glandularis, predominant involvement of the muscularis rather than the submucosa, demonstration of a urachal remnant connected to the neoplasm and presence of a suprapubic mass. These criteria were modified later by Mostofi and colleagues to include the following additional elements i.e. sharp demarcation between tumour and normal surface epithelium, and tumour growth in the bladder wall branching into the space of Retzius.\(^2\)

Ultrasound allows localization of the tumour and detection of highly echogenic calcifications as well as of the solid components of the tumour in the anterior abdominal wall without any obscuration by intraabdominal organs including air-filled bowel. At CT, urachal carcinoma may be solid, cystic, or a combination of the two. Calcifications in a midline supravesical mass are considered nearly diagnostic for urachal carcinoma.\(^5\)

Ours is a rare case of complex mucinous cystadenoma of undetermined malignant potential of the urachus, that lacked architectural features of villous adenoma as well as any evidence of stromal invasion to support a invasive neoplastic process. Urachal mucinous cystadenoma does not microscopically have definite atypia as seen in adenocarcinoma, but seem to have a low-malignant potential because it can result in pseudomyxoma peritonei if ruptured, similar to appendiceal or mucinous cystic neoplasms.\(^11\) Although such lesions in the urachus have been called adenomas, considering their potential to cause pseudomyxoma peritonei, a different terminology, ‘mucinous tumour of uncertain malignant potential’ has been suggested.\(^8\)

The mucinous neoplasms of urachus must be extensively sampled histologically to exclude any foci of dysplasia or invasive malignancy and the patients should be followed clinically post-excision for evidence of local disease recurrence.

**REFERENCES**
