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Case Report

CASE REPORT

Malignant transformation of a urachal cyst—a case report and literature review

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Abstract

Urachal remnant diseases are very uncommon pathologies which are mostly benign. Rarely they can progress to a very aggressive form of Urachal cancer. The rarity of this condition has precluded large studies to help guide the diagnostic and therapeutic management of these potentially malignant lesions. In this case, a urachal cyst was discovered and conservative management was employed after a biopsy proved the lesion was benign. Unfortunately this patient represented several years later with a locally advanced urachal cancer. To date, this is the first clearly documented case of malignant transformation. The available literature surrounding these urachal cysts and cancers will be reviewed to determine if anything could have been done differently in this case and in the future should a similar case present.

INTRODUCTION

The urachus is a vestigial fibromuscular structure situated between the transversalis fascia anteriorly and the peritoneum posteriorly. It usually involutes in early childhood to form the median umbilical ligament. Partial involution can give rise to a number of pathologies.

Though most are benign, sinister pathologies do exist. Urachal cancer is a rare and aggressive form of cancer which arises from this remnant. A urachal cyst was discovered and conservative management was employed after a biopsy was negative for malignancy. Unfortunately this patient represented several years later with what appeared to be malignant transformation of this lesion. To date, this is the first clearly defined case of malignant transformation.

CASE REPORT

A 49-year-old man was evaluated for visible hematuria with a CT urogram (CTU) and a flexible cystoscopy. These revealed features of thin walled midline cystic structure. A transurethral biopsy was done and the histology confirmed a benign urachal cyst. The patient was reassured and discharged from follow up.

He represented four years later and a repeat CTU showed the lesion now contained thick calcified walls. He was referred to a tertiary center where a robotic assisted excision of the lesion and a partial cystectomy was performed. The histopathological analysis revealed a well-differentiated (G1 pT3a) adenocarcinoma, intestinal type, with evidence of positive margins. He was started on a regime of Folinic acid, 5-Flourouracil, Oxaloplatin (FOLFOX) regime of adjuvant chemotherapy. He tolerated his first two cycles of chemotherapy well and is currently under surveillance.

DISCUSSION

The urachus is derived from two embryologic structures: the caudally invaginating allantois from the yolk sac and the ventrally invaginating cloaca from the urogenital sinus. These two structures fuse and form a connection between the yolk sac

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and the urogenital sinus. As the bladder descends into the pelvis this structure usually becomes obliterated by the fifth month of gestation [1].

As many as a third of adults will have microscopically demonstrable urachal remnants (UR) present at autopsy [2]. The natural history and incidence of cancer formation is unknown. The largest meta-analysis to date contains just over 1000 cases of urachal cancer [3]. This suggests how rare and understudied this condition is.

Urachal cancer has a number of subtle presentations. Hematuria is the most common 54–48%, pain 40%, lower urinary tract symptoms 40% and a mass 17%, and mucinuria 17–25% [4]. In as many as 72% of patients abdominal imaging may demonstrate a midline calcification [5]. There is a significant overlap between the presentation and imaging findings of benign and malignant diseases thus rendering decision-making difficult. One of the larger databases from the Netherlands Cancer registry which included 150 cases over 20 years found that 30% of cases presented at a locally advanced stage or with metastatic disease [6]. This suggests the aggressive nature and the urgent need to diagnose and identify malignant potential in these lesions. Prognosis is poor with a 40–48% 5-year survival rate [6].

Meeks *et al.* attempted to analyze the preoperative accuracy of a number of diagnostic modalities (cytology, Imaging, EUA and TURBT) before definitive surgical excision. The highest sensitivity of 93% could be found with a TURBT [5]. It may be argued however that a TURBT may risk dissemination of a potentially aggressive tumor and should not be the diagnostic modality of choice. Unfortunately Imaging was only found to have a sensitivity of 61% [5]. Currently there are no studies on surveillance of these lesions and no risk stratification tools. Age over 55 years, hematuria and calcification are all somewhat associated with increased risk of malignancy [7].

There is still no consensus on the management of adult UR disease particularly if asymptomatic. Not enough data regarding the natural history exists. Surgical excision has been proven to be a safe procedure with minimal morbidity, whether open, laparoscopically or robotic assisted.

The current accepted management of resectable UC is wide surgical excision [8]. These excisions are now safely performed using minimally invasive strategies which make use of laparoscopic or robot assisted means. Results anecdotally may seem to be equivocal however no comparative studies have been done. Other surgical strategies have included partial cystectomies and even radical cystoprostatectomies [9].

Chemotherapeutic regimes match closely those used to treat adenocarcinoma of the bowel. The optimal systemic treatment is still unknown and is largely institution dependent. Various chemotherapy and immunotherapy combinations have been tried however the numbers are still too small to draw strong conclusions [10].

CONCLUSION

Given the malignant transformation in this case early excision of the UR may have prevented progression to malignancy. UR diseases such as urachal cysts are rare disorders with very little high level evidence to guide management. They do bear the potential to undergo malignant transformation. Urachal carcinomas are associated with a poor prognosis and often present in an advanced stage. With improvements in minimally invasive surgical techniques early surgical excision should be strongly considered to avoid malignant transformation to an aggressive cancer.

CONFLICT OF INTEREST STATEMENT

None declared.

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