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Urachal adenocarcinoma surgical management – Emphasis on enbloc umbilicectomy and urachal remnant excision with negative margins

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ABSTRACT

Urachal adenocarcinoma is a rare malignant epithelial cancer. It usually presents in advanced stages. We present a 35-year-old female with urachal adenocarcinoma presented with an anterior abdominal wall swelling and ulcer managed surgically with wide excision of anterior abdominal wall along with intra-abdominal tumour mass with umbilicectomy and contiguous urachal remnant excision with abdominal wall reconstruction with mesh repair who was doing well at one year follow up. We also present a brief review regarding pathologic criteria, evolution of various staging systems and surgical aspects of urachal adenocarcinoma.

Introduction

Urachal adenocarcinoma is rare constituting 0.17 to 0.34% of all bladder cancers and 20 to 39% of primary bladder adenocarcinomas.1 We report an urachal adenocarcinoma managed surgically emphasizing importance of enbloc umbilicectomy with urachal remnant excision and negative surgical margins as important prognostic markers.

Case report

A 35-year-old female presented with swelling over lower anterior abdominal wall since 3 months with associated ulceration and serous discharge since 10 days with occasional hematuria. Local examination revealed 10 × 8 cm swelling in hypogastrium with restricted mobility, firm to cystic consistency and overlying 5 × 4 cm ulcer (Fig. 1). Wedge biopsy has shown grade 1, adenocarcinoma. Levels of Ca-125, CEA and CA19.9 were normal. Urine analysis suggestive of microscopic hematuria with no growth in culture-sensitivity. Imaging (Fig. 1) was suggestive of heterogeneously enhancing lesion of 70 × 104*59 mm involving antero-superior end of urinary bladder, superiorly extending up to umbilicus; involving overlying abdominal wall and skin with few areas of ulceration; bilateral enlarged external iliac nodes largest 10 × 12 mm on right side. Cystoscopy shown erythematous mucosa at dome of bladder. Colonoscopy was normal.

After informed and written consent, intraoperatively two separate incisions; one circular incision around umbilicus and another elliptically around abdominal wall mass was planned. Flaps elevated and incision deepened. Umbilicus excised in contiguous with urachal remnant till the level of tumour mass. Tumour mass located at dome of bladder, infiltrating anterior abdominal wall and 4cm length of sigmoid colon. Explored solid organs and peritoneal cavity normal. Partial cystectomy along with en-mass excision of tumour mass + resection of involved sigmoid colon with bilateral pelvic lymphadenectomy; bladder and sigmoid colon repair was performed (Fig. 2). Prolene mesh used for abdominal wall reconstruction. Operative duration was 180 min with 300ml blood loss. One unit packed cell transfusion given post-operatively. Patient kept nil per oral for 5 days and IV antibiotics for 7 days. Drains removed on post op day 6. Patient discharged on post op day 8 and catheter removed on day 14.

Patient was doing well in follow up period with well healed scar (Fig. 2). Histopathology (Fig. 3) suggestive of urachal adenocarcinoma, grade II with areas of necrosis extending into skin of anterior abdominal wall; infiltrates into serosa, muscular layer of urinary bladder and extends up to mucosa projecting into bladder cavity; infiltrates into serosa and muscular layer of sigmoid colon with normal overlying mucosa; Resected margins of skin, sigmoid colon, bladder wall and inked soft tissue margins are free of tumour. All eleven (right) and thirteen (left) pelvic nodes were negative.

No adjuvant therapy was considered as surgical margins and nodes were negative. At 12 months follow up, patient was asymptomatic and with normal imaging.

Discussion

Mucin secreting or non-secreting adenocarcinomas arise from innermost epithelial layer of urachus, with lesser variants being
sarcomas, transitional cell and squamous cell carcinomas. Pelvic lymph nodes are regional nodes.

Most common symptoms are abdominal pain, hematuria, palpable suprapubic mass, irritative voiding, discharge from umbilicus. Nonspecific nature of most symptoms delays diagnosis leading to locally advanced presentation. Differential diagnosis includes primary bladder...
but identifiable only in 32% of cases; no primary adenocarcinoma other than cystitis glandularis; presence of urachal remnants is helpful for polypoid proliferation; no carcinoma in situ or glandular metaplasia of surface bladder urothelium; surface urothelium is free of glandular or muscularis or deeper tissues with sharp demarcation between tumor and bladder. 

Sheldon et al. based on observations from 117 cases first proposed surgico-pathological staging in 1983. Stage I-tumors confined to urachus and/or bladder; Stage II-tumors extending beyond muscular layer of urachus and/or bladder; Stage III-tumors infiltrating regional lymph nodes; Stage IV-tumors infiltrating non-regional lymph nodes or other distant sites. Both Mayo and Sheldon staging systems are highly correlated (p = .82; PSP<0.001) and compared favorably in their ability to predict survival. Sheldon staging with 8 stage categories was found to be over specified and unnecessarily complex, due to low number of patients in four of its staging strata (Stage I, IIIB, IIIC, IId) than the simpler four category Mayo staging as proved by Akaike Information Criterion (AIC).

Local control in form of surgical approach remains mainstay first line treatment and more effective than radiotherapy for resectable lesion. Surgery includes partial cystectomy, tumor debulking with en bloc umbillectomy and urachectomy with negative surgical margins. 7% of urachal cancers have umbilicus involvement and 81% of patients surviving at 5 years have urachal ligament excised. Umbillectomy and en bloc urachal remnant excision is justified to improve recurrence and survival rates. Survival benefit of radical cystectomy and en bloc umbillectomy and urachectomy with negative surgical margins or metastatic disease. Overall 5-year cancer specific survival ranges from 40% to 51% in published series, which is significantly better than those with non urachal adenocarcinomas and urothelial carcinomas of bladder.


doxorubicin and FOLFOX. Use of adjuvant therapy was not associated with significant improvement in cancer-specific survival (HR,1.6; 95% CI, 0.7–3.6) and response rates up to 48% documented in unresectable or metastatic disease. Overall 5-year cancer specific survival ranges from 40% to 51% in published series, which is significantly better than those with non urachal adenocarcinomas and urothelial carcinomas of bladder. 

Conclusion

Urachal adenocarcinomas are rare, presents in advanced stages, but better prognosis than other bladder tumors. Clinical diagnosis supported with imaging, initial radical surgical management to achieve negative margins with umbillectomy and urachal remnant excision, timely surveillance can help to improve outcomes and survival.

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Contribution details

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