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Case Report

A Rare Case of Metastatic Urachal Carcinoma

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Abstract

Urachal carcinoma is a rare tumor, it represents about 0.5-1% of all bladder malignancies. In the majority of cases it is an adenocarcinoma. For the treatment it is recommended for resectable tumors, a partial cystectomy with resection of the urachal ligament, the bladder dome and umbilicus. There is no standard neoadjuvant or adjuvant treatment, and the combinations of 5-fluoruracil with cisplatin are active in metastatic stage. A higher risk of relapse after surgery has been observed in some situations: positive margins, lymph node involvement, involvement of the peritoneal surface, or in the case of the umbilicus was not resected en-bloc.

Keywords: urachus; adenocarcinoma; immunotherapy

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Introduction

The urachus is a musculo-fibrous band that connects the dome of the bladder and the umbilicus [1]. Histologically, urachal tumors are frequently adenocarcinomas, and the histology of classic bladder cancer is typically transitional epithelium carcinoma or squamous carcinoma [2]. It often presents at metastatic stage and has a poor prognosis. Unlikely, there is no standard chemotherapy regimen for the treatment of urachal cancers, especially for the metastatic patients. We present a case of metastatic urachal cancer treated with different treatments.

Case presentation

A patient of 49 years old, has a familial history: small cell lung cancer of his mother and rectal cancer of his father, he presented abdominal pain and urinary tract infection.

He underwent a computed tomography scan (CT scan) in June 2011 which showed a left antero-lateral mass of the bladder of 14mm extending to the root of urachus.

A partial cystectomy was performed on June 2011 to remove the urachus, umbilicus and pelvic lymph nodes. Pathology showed a mucin producing adenocarcinoma, the tumor was extending to tissue peri vesical, classed pT3 pN0 R0 and a follow up was decided.

In February 2014 a CT scan showed pulmonary and pleural metastases (Figure 1).

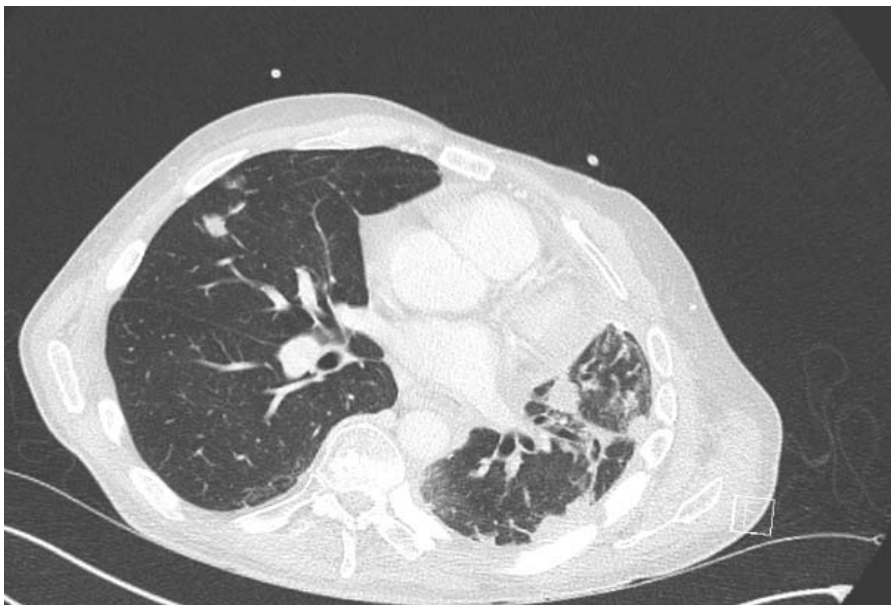

Figure 1 : CT scan showed pulmonary and pleural metastases

A resection of 2 nodules was performed which confirm a pulmonary metastasis of urachal adenocarcinoma and In April 2014 another resection of secondary lesion of pulmonary upper left lobe was done. The patient underwent a positron emission tomography (PET scan) which showed an increase in left pleural effusion thus the decision of multidisciplinary concertation meeting is chemotherapy type folfox-Bevacizumab, he received 10 cycles and in August 2016 he beneficed from

LV5FU2-Bevacizumab with stability as result. And from September to December 2016 LV5FU2 was replaced by Capecitabine. A CT scan showed new pulmonary lesion and bone metastases (Figure 2) .

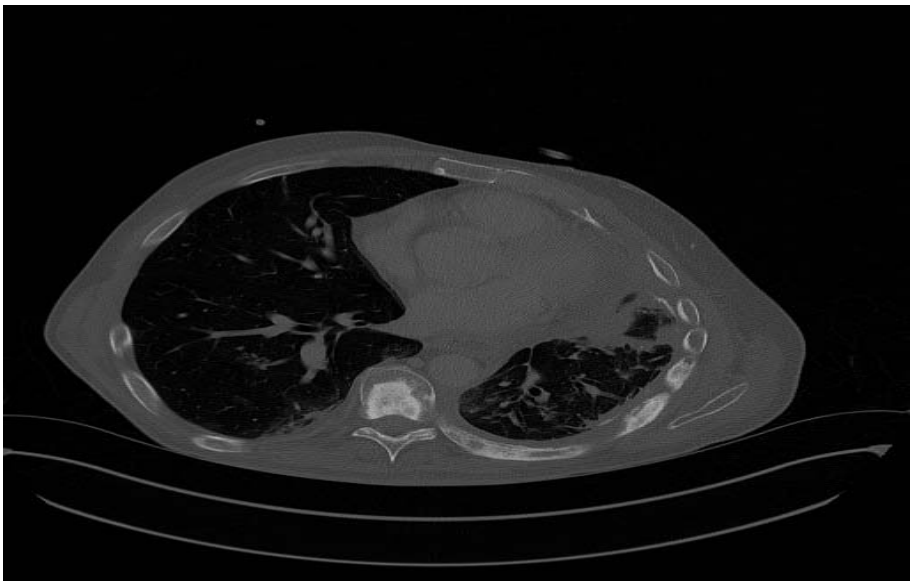

Figure 2 : CT scan showed bones metastases

The decision was a resection with talcage in February 2017. Four months after a clinical and biological progression of ACE was observed so the treatment was changed and the patient received 11 cycles of folfiri-Bevacizumab From July 2017 to January 2018 with switch to Capecitabine and Bevacizumab , he had also received a radiotherapy of dorsal rachis from December 2017 to January 2018 and as result a radiological stability was observed.

The patient was hospitalized in June 2018 because of deterioration of his general status and he died in July 2018.

Discussion

Urachal carcinoma (UrC) is rare tumor. The most frequent histological subtype is adenocarcinoma. They are asymptomatic for a long period of time and may be found incidentally on computed tomography scan (CT scan) or when symptoms of local invasion have developed [2].

The diagnosis of urachal cancer needs a multidisciplinary approach [3]. It is generally radio et chemo-resistant. The prognosis of metastatic urachal cancer is extremely poor [4].

Symptoms include irritative voiding symptoms and an important hematuria. The standard exams includes computed tomography scan(CT)or magnetic resonance imaging (MRI). Cystoscopy is also recommended [5].

The first system for staging urachal cancers was proposed by Sheldon and colleagues in 1984. A more simplified system has been recently proposed by the Mayo clinic [6]. The median survival for locally advanced or metastatic disease is between 12 and 24 months and the 5-year cancer specific survival rate for urachal cancers is only 50% [7].

Partial cystectomy is considered the gold standard for the treatment of urachal carcinoma when the disease is resectable. En bloc resection of the urachal ligament and umbilicus is recommended because tumors can be situated anywhere along the urachus [4].

There is no standard treatment in the both situations adjuvant and metastatic for urachal cancers. The choice of regimens has been based on case reports [8]. Chemotherapy regimens used in Canada to treat advanced bladder cancer, including gemcitabine+cisplatin or methotrexate+vinblastine+ doxorubicin+cisplatin (MVAC), have shown a few responses in urachal cancers [9].

And because of the enteric-type histology of urachal adenocarcinomas, chemotherapy regimens used to treat gastrointestinal malignancies may be efficace. Two case reports have described patients responding to irinotecan-based chemotherapy, which is used to treat colorectal cancers [10]. Two case reports using oxaliplatin-based chemotherapy have also shown benefit in urachal cancers [11]. There is one report of a triplet combination of 5-flurouracil, doxorubicin and mitomycin-C has shown activity in this disease [12]. And concerning clinical trials, there was one trial of a triplet combination of ifosfamide with paclitaxel and cisplatin in advanced non-transitional cell carcinoma that has reported a moderate activity in urachal cancers [13].

There is also a phase II study evaluating gemcitabine ,fluorouracil, leucovorin and cisplatin in metastatic urachal cancer and other adenocarcinomas of the bladder, and preliminary results from this study are encouraging .This regimen has been adopted as the adjuvant and front-line metastatic regimen at the MDACC.

In 2015, Le et al. evaluated the clinical activity of an anti-PD-1(pembrolizumab) in a cohort of metastatic carcinoma patients with or without MSI [11].The results of this phase 2 study, with results from another phase 2 study evaluating nivolumab (anti-PD-1) with or without ipilimumab (anti-CTLA-4) , showed that MSI status was able to predict clinical benefit from immunotherapy.

Conclusion

Urachal cancer is an aggressive tumor. Surgery is the gold standard in localized case and there is no standard treatment in the both situations adjuvant and metastatic for urachal carcinoma. In addition, agents that demonstrate efficacy in metastatic disease should be studied in trials to clarify the role of neoadjuvant or adjuvant chemotherapy in urachal adenocarcinoma.

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