Oncology

Multimodal therapy in urachal carcinoma with oligometastatic bone disease

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ABSTRACT

A 35-year-old male patient was admitted to our hospital for urachal carcinoma with oligometastatic bone disease. He received a surgical resection through the umbilical area, urachal ligament and bladder dome (partial cystectomy) and adjuvant chemotherapy based on cisplatin-gemcitabine regimens together zoledronic acid with a good tolerance, no toxicity. Nine months after surgery, our patient presented no symptoms and the metastasis had been brought under control with no apparent signs of recurrence as assessed in a follow-up CT. To our knowledge, is the first case report of a urachal carcinoma with oligometastatic bone disease who received multimodal therapy, including surgery resection.

Introduction

Urachal carcinoma is a rare and highly malignant form of bladder cancer (< 1% of all bladder carcinomas), which develops in the urachus. This aggressive cancer often evolves into distant metastasis. Because of its rarity and the small number of cases/series reported, there is little knowledge of this devastating pathology.

We report the case of a young male patient with urachal carcinoma that had already metastasized to the bones.

Case presentation

A 35-year-old male patient complaining of pain in the suprapubic region and hematuria underwent an ultrasound examination, which revealed a mass lesion localized in lower abdomen, and closely related to bladder dome.

The patient also underwent a computed tomography, a magnetic resonance imagining examination (Fig. 1) and a bone gammography. The overall diagnosis revealed evidence of a solid prevesical cystic mass intimately related to the bladder dome; two bone metastases were also detected in both the vertebral body D10 and right iliac fossa.

A flexible cystoscopy confirmed focal affectation into bladder.

Transurethral resection of the intravesical portion (Fig. 2) was performed and the anatopathological diagnosis showed invasive mucus-secreting adenocarcinoma poorly differentiated with a component of signet-ring cells with probable genesis in the urachus and extending into the tissue of the bladder.

After presenting this case to our Uro-Oncology Executive Committee, it was decided that a surgical resection and adjuvant chemotherapy would be the most suitable therapeutic approach. The resection was conducted through the umbilical area, urachal ligament and in the bladder dome (partial cystectomy: Fig. 2) as well as a bilateral pelvic lymphadenectomy. The anatopathological diagnosis of the surgical sample revealed an intestinal-type urachal mucinous cystadenocarcinoma that extended throughout the urachus. It was moderately demarcated, not invading the margins of the resected area, while the lymph nodes did not present tumorous affectation (Fig. 3).

One month after surgery, the patient received adjuvant chemotherapy with 145mg cisplatin and 2,450mg gemcitabine every three weeks “X” cycles and zoledronic acid every three months with good tolerance and no toxicity. Nine months after surgery, our patient presented no symptoms, and the metastasis had been brought under control with no apparent signs of recurrence as assessed in a follow-up CT.

Discussion

Urachal carcinoma is an infrequent malignancy with a predilection for male patients. When accompanied with symptoms, the most common may include hematuria, abdominal pain, mass lesions in the suprapubic region and umbilical secretion.1

While the urachal remnant is in close contact with the bladder urothelium, the majority of urachal cancers are mucus-secreting adenocarcinomas with signet-ring cells.2 Imaging exams (tumour localized in the bladder dome) and the cystoscopical findings (intact or ulcerated urothelium) are necessary in the differential diagnosis of a bladder adenocarcinoma, given the significant overlapping of histological and...
The primary treatment for localized urachal cancer includes complete surgical resection of the urachus, the umbilical region or the surrounding soft tissue combined with partial or radical cystectomy and bilateral pelvic lymphadenectomy. However, 20%-38% of patients experienced disease recurrence: pelvis (37%), bladder (34%), lungs (28%) and lymph nodes (18%). In most cases, local recurrence was observed two years after surgery.

After diagnosis approximately 20% of cases already present distant metastasis: liver (46%), bones (especially along the spine) and lungs (31%), peritoneal (15%) and nodular carcinomatosis (7%). Most of these patients will sadly die of this disease, with a mean survival rate of as little as one year.

Adjuvant therapy with chemotherapy and/or radiotherapy may minimize the recurrence risk, although the role of this therapeutic approach has not yet been established. The choice of regimens has been based largely on case reports and single institution experiences. The chemotherapeutic regimens in the treatment of advanced bladder cancer, including gemcitabine together with cisplatin or the combination of methotrexate, vinblastine, doxorubicin and cisplatin (MVAC), have shown a few anecdotal responses in urachal carcinomas, but have generally yielded rather disappointing results.

A retrospective series of 21 patients, including 14 with adenocarcinoma, had a 36% response rate to first-line cisplatin-based regimens.

The triplet combination chemotherapy regimen consisting of 5-fluorouracil, doxorubicin, and mitomycin-C has been reported to exhibit some activity against this carcinoma.

As far as clinical trials are concerned, a prospective study of ifosfamide, paclitaxel and cisplatin in male patients with advanced non-urothelial bladder cancer (11 in 20 with adenocarcinoma, including six of them with urachal adenocarcinoma) reported a 36% response rate and a mean survival rate of 25 months. There is also a phase II study whose preliminary results are quite encouraging after evaluating the quadruple combination of gemcitabine, fluorouracil, leucovorin and cisplatin (GemFLP) in metastatic urachal cancer and other bladder adenocarcinomas.

In our institution, we had been consistently administering regimens based on cisplatin and fluorouracil for bladder adenocarcinomas. Recently, we have been making use of combined regimens with cisplatin and gemcitabine and, much to our surprise, we have found this combination to exhibit not only greater tolerance but also greater effectiveness. Even though the significant reduction in the disease progression and the clinical improvement are common, the important and long-lasting responses have been few and far between. In the present case, we opted for a chemotherapeutic regimen of cisplatin and gemcitabine together with zoledronic acid in view of the oligometastatic bone disease and have managed to keep the disease under control. However, our follow-up period is insufficient to evaluate relapses.

Conclusion

Urachal carcinoma is an aggressive malignancy for which there is limited medical literature available. The initial silent behavioural pattern of this malignancy prevents an early detection. When diagnosis has been established in advanced stages of the disease, distant metastases are common.

Most medical groups vouch for surgery as the basis of treatment, although the role of chemotherapy and radiotherapy has not yet been established. Standard chemotherapy should include a regimen commonly used in gastrointestinal cancers.

The results of clinical trials currently underway might further define the standard treatment for this pathology. Further multicentre studies into the role of adjuvant therapies in the optimal management of this type of tumours seem truly justified.

Consent

The patient granted his verbal and written consent for the publication of this clinical case.
Conflict of interest statement

The authors declare that they have no conflicts of interest.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2019.100936.

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Fig. 3. The anatomopathological diagnosis of the surgical sample revealed an intestinal-type urachal mucinous cystadenocarcinoma that extended throughout the urachus. It was moderately demarcated, not invading the margins of the resected area, while the lymph nodes did not present tumorous affectionation.