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Case Report

Small Cell Cancer of the Genitourinary Tract: A Case Report and Review of the Literature

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Keywords
Small cell carcinoma · Genitourinary cancer · Extrapulmonary small cell carcinoma

Abstract
Small cell carcinoma of the urinary tract is an extremely rare disease with very few cases reported in the literature. Its clinical course is aggressive, and the prognosis is poor. Here, we present a case of metastatic extrapulmonary small cell carcinoma of the upper urinary tract in a 74-year-old African-American male. He initially presented with gross hematuria, 20-pound weight loss, and abdominal pain for 2 months. CT imaging showed a 14.0 × 7.0 × 16.0 cm retroperitoneal mass within the left renal fossa; biopsy revealed a carcinoma which was positive for synaptophysin and chromogranin. The patient also had detectable neuroendocrine cells in his urine cytology, confirming the diagnosis of small cell carcinoma. He was treated with carboplatin and etoposide as extrapolated from the treatment of its pulmonary counterpart. Due to the rarity of urinary tract small cell carcinoma, no randomized studies exist to guide therapy or management.

Background

Extrapulmonary small cell carcinoma (EPSCC) is extremely rare. It most commonly affects the gastrointestinal and the genitourinary systems [1]. Within the genitourinary tract,

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the bladder and prostate are the most frequently affected sites. The prognosis is poor, with a 5-year overall survival as low as 8.1% [2]. The incidence of EPSCC in North America is estimated to be between 0.1 and 0.4% [3]. EPSCC of the upper urinary tract, in particular, has had no more than 40 cases reported in the literature [4, 5]. The current knowledge of these rare tumors is limited, and treatment recommendations are mainly based on case reports, single-institution case series, or the extrapolation of results from pulmonary SCC [1, 3–8]. No consensus guideline exists, and further work is necessary to better our understanding of how to best manage these patients.

Case Presentation

A 74-year-old African-American male with a past medical history of severe chronic obstructive pulmonary disease, coronary artery disease, hyperlipidemia, and hypertension presented to our hospital in July 2016 with complaints of abdominal pain and gross hematuria for 1 week. His abdominal pain was sharp in nature and suprapubic in location. His hematuria had been worsening for the past week, and further history revealed that he had been having transient hematuria for the past year and a half. Additionally, for the past 2 months, he reported decreased appetite, fatigue, and a 20-pound weight loss. No other pertinent positives on review of systems were noted. His family history was unremarkable for cancers in any first- or second-degree relatives, but his social history did reveal that the patient was a heavy former smoker. Physical examination was significant for mild left lower quadrant abdominal tenderness and a large left supraclavicular mass that was firm and fixed.

CT imaging of the abdomen and pelvis revealed a 14.0 × 7.0 × 16.0 cm heterogeneously enhancing retroperitoneal mass within the left renal fossa extending into the midline (Fig. 1). It showed involvement of the surrounding vasculature, as well as liver and bone metastases of the L2 and L4 vertebral bodies. CT scan of the chest was positive for bilateral pleural effusions and an additional T3 lytic lesion, but no pulmonary lesion was found. A cystoscopy was performed which did not reveal any gross abnormality of the bladder; however, urine cytology showed malignant cells concerning for SCC. Biopsies of the left retroperitoneal mass and supraclavicular lymph node were performed, both of which showed immunohistochemistry positivity for synaptophysin and chromogranin and partial positivity for PanCK and CK7. They were negative for CD45, S100, CD3, CD20, and p63. These findings led to the diagnosis of SCC of the upper urinary tract with bone and liver metastases. MRI of the brain did not show any metastatic lesions.

He was started on chemotherapy with carboplatin and etoposide, along with growth factor support. He was also given monthly bisphosphonate therapy for the lytic vertebral lesions.

Repeat CT scans at 3 months, after 4 cycles of chemotherapy, showed marked volume reduction of the retroperitoneal mass (Fig. 1). In addition, the liver lesions were no longer evident. However, there was persistent left retroperitoneal tumor within the left renal sinus and left perinephric space, with extension into the surrounding vasculature. Furthermore, there was progression of bone metastases throughout the thoracolumbar spine, with a new pathologic fracture of the L2 vertebra. Physical exam was noteworthy for a significant decrease in the size of the supraclavicular lymph node.

Unfortunately, after 2 additional cycles of chemotherapy, re-staging scans showed multiple new hepatic metastases, the largest measuring 1.4 × 1.3 cm, and an increase in size of

the primary retroperitoneal mass. Because of this, he was offered salvage chemotherapy with weekly topotecan, but he did not tolerate the therapy well and expired of progressive disease 7 months from his initial diagnosis.

Discussion

EPSCC is a difficult disease to treat and challenging to study due to the small number of cases, which limits the development of randomized clinical trials. The most common treatment regimen used is a combination of a platinum agent with etoposide, extrapolated from studies for SCC of the lung. The response rate has been reported to be as high as 71% [9]. Ouzzane et al. [4] reviewed 39 cases and noted that a platinum and etoposide adjuvant regimen increased survival as compared to surgery alone in patients with EPSCC of the upper urinary tract, but this difference was not statistically significant. However, many case reports and single-institution experiences have shown a significant increase in median survival with chemotherapy [3, 4, 7–11]. Dakhil et al. [3] reviewed 35 cases from the University of Kansas tumor registry and found that a platinum-based agent with etoposide improved median survival to 36 months compared to 5 months without chemotherapy. Osaka et al. [10] reported a case of SCC of the ureter that was successfully treated by neoadjuvant cisplatin with irinotecan followed by nephroureterectomy with no recurrence or metastasis for 38 months postoperatively. Beddok et al. [5] reported similar success with an 80-year-old male with SCC of the pelvic ureter with carboplatin and etoposide.

Despite the support for the role of early chemotherapy, there is no head-to-head study that compared the efficacy of different chemotherapy regimens. To date, there is no consensus therapy for patients with EPSCC. More data are clearly needed to further our understanding of the optimum management of patients with this highly aggressive disease.

Statement of Ethics

The individual used for this case report has given his informed consent and is documented in our records.

Disclosure Statement

The authors declare that they have no conflicts of interest.

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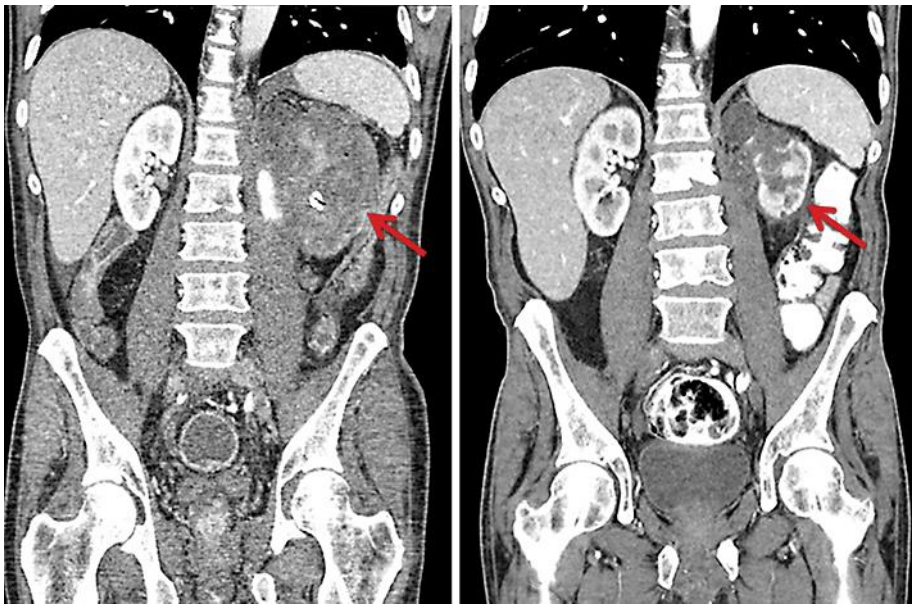

Fig. 1. Left retroperitoneal mass that was discovered at the time of presentation (left) compared to the reduced mass after 3 cycles of cisplatin and etoposide (right).