Urology Case Reports 6 (2016) 33–35

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Primary Seminal Vesicle Adenocarcinoma Presenting With Bilateral Orbital Metastasis: A Case Report

Matthew E. Sterling*, Robert C. Kovell, William I. Jaffe

Division of Urology, Department of Surgery, University of Pennsylvania Health System, Philadelphia, PA 19104, USA

ARTICLE INFO

Article history:
Received 6 February 2016
Received in revised form
15 February 2016
Accepted 17 February 2016

Keywords:
Hematospermia
Seminal vesicle adenocarcinoma

ABSTRACT

Seminal vesicle (SV) adenocarcinoma is a rare and poorly understood malignancy. Symptoms are non-specific and prognosis is extremely poor. Herein we present a case report of a primary SV clear cell adenocarcinoma with bilateral orbital metastases at the time of initial presentation treated with multimodal therapy including radiotherapy and multi-drug chemotherapy.

Introduction

Seminal vesicle (SV) adenocarcinoma is a rare and poorly understood malignancy. Symptoms are non-specific and patients often present with advanced or metastatic disease. Prognosis is poor with ~95% dying within 3 years of diagnosis.[1] We present a case report of a primary SV clear cell adenocarcinoma with bilateral orbital metastases at the time of presentation.

Case report

A 29-year-old man initially presented to his primary care physician (PCP) with blurriness and pain in his right eye, bloody discharge per urethra, and a weak urinary stream. For the bloody discharge and change in his voiding patterns he was prescribed antibiotics. For his eye pain and blurry vision he was referred to an Ophthalmologist. On examination a tumor was discovered in his right eye and a smaller tumor in his left eye. A computed tomography (CT) scan of the head showed no additional disease. Further staging with a CT of the chest, abdomen, and pelvis revealed an 11 × 7 cm partially cystic heterogeneous pelvic mass of uncertain origin posterior to the base of the bladder (Fig. 1). He was referred to urology due to his persistent bloody discharge and newly diagnosed pelvic mass. Digital rectal exam (DRE) revealed a large and firm mass indistinguishable from the prostate gland. Lab work revealed an elevated CA-125 at 40 U/mL and an undetectable prostate specific antigen (PSA) (<0.2 ng/mL).

He underwent a cystoscopic evaluation revealing a heterogeneous prostatic urethra with multiple abnormal protrusions and a mass effect on the bladder displacing the trigone off of the midline, but no mucosal lesions concerning for urothelial cancer. The abnormal prostatic protrusions were biopsied. We then performed a transrectal ultrasound-guided biopsy of the pelvic mass. Both biopsies revealed a poorly differentiated adenocarcinoma with papillary features, mucin production, and clear cell histology. Immunohistochemistry was performed and was positive for CK7 (Fig. 2), AE1-3, and Cam5.2. Tumor cells were negative for CK20 (Fig. 3), PSA, PSAP, Desmin, smooth muscle actin, CD30, PLAP, AFP, CD117, CD34, and S100. Based on all of these immunohistochemistry markers, the pathologic diagnosis was most consistent with a poorly differentiated carcinoma originating from the SV.

Given orbital and lymph node metastasis, upfront chemotherapy with carboplatin/paclitaxel was initiated and he completed 4 cycles. He underwent radiotherapy to his orbital lesions and large pelvic mass. He later developed a rectovesical fistula and underwent an ileostomy diversion. His disease burden was stable for 15 months until he developed local and distant progression. He continued to progress despite treatment and palliative care was provided. He expired 4.5 years after his initial diagnosis.

Discussion

Since primary SV adenocarcinoma was first described, only about 50 cases have been reported in the literature.[1,2] Despite

* Corresponding author. Tel.: +1 215 776 8955; fax: +1 215 662 7413.
E-mail address: Matthew.Sterling@uphs.upenn.edu (M.E. Sterling).

http://dx.doi.org/10.1016/j.eucr.2016.02.011

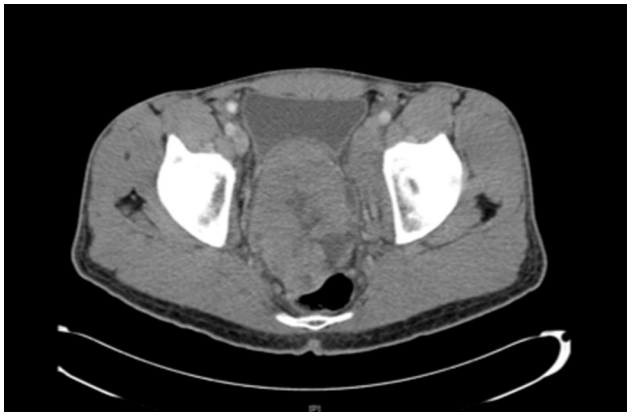

Figure 1. Contrast enhanced CT scan of the pelvis showing a large, heterogeneous mass.

improvements in imaging modalities, the diagnosis remains primarily one of exclusion.

Symptoms do not typically develop until late in the clinical course and are non-specific. The most commonly reported symptoms are hematospermia, hematuria, bladder outlet obstruction, and pelvic pain.[2] Because symptoms are initially non-specific or non-existent, diagnosis is delayed and most patients present with advanced or metastatic disease, often to locoregional lymph nodes. Our patient initially presented with bilateral orbital metastasis, which we believe is the first reported case of its kind. Physical exam typically reveals a non-tender mass on DRE, which may be indistinguishable from the prostate. It is thus of great importance to rule out other primary malignancies such as prostate, bladder, or colorectal cancers since SV carcinomas are often locally advanced at presentation and appear similar to these other malignancies.[2]

Imaging is critical in the diagnosis and helps rule out other potential malignancies. CT, magnetic resonance imaging (MRI), or transrectal ultrasound may all be useful in determining the diagnosis.[2] PET imaging can help exclude distant metastasis [3,4] or determine the response to treatment.[4] Cystoscopy and/or sigmoidoscopy, while non-specific, may be considered.

A laboratory evaluation should be obtained. Serum CA-125 levels are generally elevated for primary SV carcinomas, whereas PSA and CEA are typically normal.[2] Diagnosis is often confirmed via transrectal ultrasound-guided biopsies.[2]

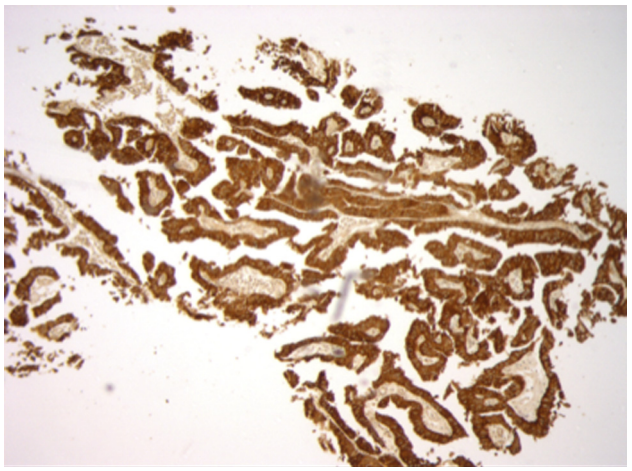

Figure 2. CK7 positive staining of transrectal biopsy of pelvic mass.

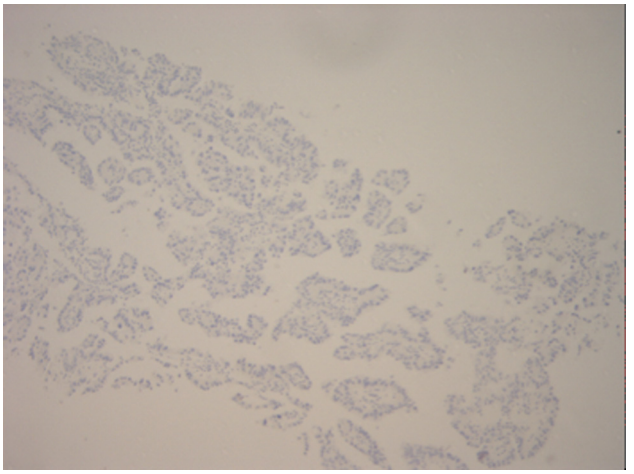

Figure 3. CK20 negative staining of transrectal biopsy of pelvic mass.

Even with improvements in radiologic studies, pathologic analysis remains the mainstay for diagnosis (Figs. 2 and 3). Adenocarcinoma is the most common malignant lesion of the SV whereas cystadenoma and mesenchymal tumors represent the most common benign lesions.[5] Immunohistochemistry is critical in differentiating SV carcinoma from cancers of the prostate, bladder, or rectum. SV carcinomas stain strongly positive for CK7 (Fig. 2) and CA 125, negative for CK20 (Fig. 3), and variable for CEA.[1–3] Further study will be needed to delineate which pathologic features correlate with prognosis.

There are no established guidelines for the treatment of primary SV adenocarcinoma. When the disease remains clinically localized, complete surgical resection is the mainstay of treatment. This can range from a simple excision to a complete pelvic exenteration depending on the location and size of the tumor.[5]

Multimodal therapy is the rule for more advanced cases of SV carcinoma. Radiation therapy is not effective for local disease control; however, it may be utilized as an adjunctive therapy in those with unresectable tumors or positive margins.[5] Androgen deprivation therapy has been used as an adjunct with reports suggesting improvements in survival.[1,5] For cases presenting with advanced or metastatic disease, chemotherapy can be tried, although its use is controversial and there exists no definitive recommendation on which regimen is best. Thyavihally et al, attempted 5-fluorouracil, leucovorin, and oxaliplatin in addition to androgen deprivation therapy with some success.[5]

In our patient, no surgical therapy was attempted given the widespread nature of his disease, but he responded well to targeted radiation therapy and multidrug chemotherapy with survival of 4.5 years from the time of diagnosis. Although his course was complicated, it shows that treating this rare and aggressive cancer with multiple treatment modalities may provide the best outcomes.

Conclusion

Given the rarity of the condition, non-specific symptomatology, and aggressive nature of the disease, SV cancers remain a challenging clinical entity for the practicing urologist.

Early diagnosis and aggressive extirpative therapy offer the best chance at a durable response.

Conflicts of interest

None.

Source of funding

None.

References

1. Benson Jr RC, Clark WR, Farrow GM. Carcinoma of the seminal vesicle. *J Urol*. 1984;132:483–485.
2. Thiel R, Effert P. Primary adenocarcinoma of the seminal vesicles. *J Urol*. 2002;168:1891–1896.
3. Navallas M, Vargas HA, Akin O, et al. Primary seminal vesicle adenocarcinoma. *Clin Imaging*. 2011;35:480–482.
4. Kreiner B, Denzinger S, Ganzer R, et al. Neuroendocrine carcinoma of the seminal vesicles presenting with Lambert Eaton syndrome: a case report. *J Med Case Rep*. 2010;4:320.
5. Thyavihally YB, Tongaonkar HB, Gupta S, Gujral S. Primary seminal vesicle adenocarcinoma presenting as isolated metastasis to penis responding to chemotherapy and hormonal therapy. *Urology*. 2007;69:778.e1–778.e3.