Angiosarcoma of the seminal vesicle: a case report of long-term survival following multimodality therapy

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Abstract

Angiosarcoma of the seminal vesicle is an extremely rare malignancy, with few published case reports in the literature. We present a case of primary angiosarcoma of the seminal vesicle in a 45-year-old male who was treated with multimodality therapy, consisting of neoadjuvant chemotherapy and chemoradiation followed by surgical resection and intraoperative radiation therapy. He has been free of cancer recurrence for more than six years after completion of therapy. To our knowledge, this represents the longest reported survival of a patient with this rare tumor, and one of the few cases reported using a multimodality therapy approach.

Case Report

A previously healthy 45-year-old male developed sharp left lower quadrant and groin pain. He was evaluated by his primary physician and his symptoms were thought to be related to diverticulitis. He was treated conservatively with an antibiotic, though there was no resolution of his symptoms.

A computed tomography (CT) scan of the pelvis revealed a 5.6×5.1 cm heterogeneously enhancing mass involving the left lobe of the seminal vesicle and minimally the right lobe across the midline. The mass was invading the left bladder base and ureterovesicular junction (UVJ), and a few cm of left distal ureter (Figure 1A). This was accompanied by left-sided hydrenephrosis. The mass also invaded the left obturator nerve and adjacent skeletal muscle, and extended into the left lateral rectal area without apparent pelvic sidewall involvement.

The tumor involvement was limited to the left seminal vesicle, the base of the prostate and the right bladder wall (Figure 1B-D). The mass extended laterally resultin effacement of the left obturator internus muscle. A fatty soft tissue plane separated the mass from the adjacent left anterior rectal wall. A chest CT showed a few small indeterminate pulmonary nodules and a small calcified granuloma within the left lower lobe. A colonoscopy showed no evidence of rectal invasion. Given the presence of locally advanced tumor and the high risk for micrometastatic disease, the multidisciplinary team consisting of urology, medical oncology, and radiation oncology physicians recommended multimodality therapy consisting of neoadjuvant chemotherapy and radiotherapy followed by potential surgical resection.

The patient received two cycles of neoadjuvant IMAP chemotherapy, consisting of ifosfamide (2500 mg/m² IV days 1-2), Adriamycin (40 mg/m² IV day 2), mitomycin (4 mg/m² IV day 2), cisplatin (60 mg/m² IV day 2), and mesna (1500 mg/m² IV and 2500 mg/m² per days 1-2). Repeat imaging following IMAP showed that the soft tissue mass in the left pelvis slightly decreased in size from 5.6×5.1 cm to 4.3×4.0 cm. The patient then received external beam radiation therapy (Figure 1E) consisting of 50 Gray in 25 fractions delivered daily over 5 weeks with 2 cycles of concomitant MAP chemotherapy, consisting of mitomycin (8 mg/m² IV), Adriamycin (40 mg/m² IV) and cisplatin (60 mg/m² IV).

Following a six-week recovery from preoperative chemoradiation, repeat imaging studies showed no significant change in the pelvic mass and no evidence of distant metastatic disease. He then proceeded to surgical resection with a radical prostatectomy, partial cystectomy, distal ureterectomy, bilateral pelvic lymph node dissection, and ureteroneocystotomy. Frozen section margins at the bladder and left pelvic sidewall were positive. Additional resection was performed and the margins were negative. An intraoperative electron radiation therapy boost (9 Gy) was given to the left pelvic sidewall as there was a concern for residual microscopic disease.

The tumor involved the left seminal vesicle, infiltrated adjacent skeletal muscle, and extensively involved regional nerves and ganglia, but tumor did not extend into prostatic parenchyma. The final surgical margins were negative for tumor. Multiple left and right pelvic lymph nodes (external iliac, internal iliac, common iliac and obturator regions) were negative for tumor involvement.

Post-operatively he had left thigh numbness that subsequently resolved. He developed a vesicourethral anastomotic leak which required prolonged (3 months) Foley catheter use; however, this completely resolved. He developed an abscess that was drained and resolved. The patient was then seen in regular follow up with serial imaging studies over the subsequent six years following surgery and he has not developed evidence of cancer recurrence.

Key words: angiosarcoma, seminal vesicle, prostate, neoadjuvant, surgery, chemoradiation, trimodality therapy.

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Discussion and Conclusions

Soft tissue sarcomas (STS) comprise 1-2% of all cancers and can appear in any type of connective tissue throughout the body. Angiosarcomas are a rare subtype of STS, and arise in endothelium of blood vessels and represent 2% of STS cases. Reported risk factors for development of angiosarcoma include prior exposure to ionizing radiation or toxic chemicals, longstanding lymphedema (Stewart-Treves syndrome), and venous stasis, although most patients diagnosed with angiosarcoma have no identifiable risk factors. Angiosarcomas are most commonly found in the scalp, face, neck, extremities, or breast. In contrast, angiosarcoma of the genitourinary system is exceedingly rare.

Very few cases of angiosarcoma of the prostate or seminal vesicle have been reported in the literature (Table 1). For angiosarcomas of the seminal vesicle, presenting symptoms may include testicular discomfort and...

Table 1. Clinical summaries of primary seminal vesicle and prostate angiosarcoma patients in literature, with one patient per row.

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Age</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>This study</td>
<td>2014</td>
<td>45</td>
<td>CT, RT, SR</td>
<td>NED, 6 years AT</td>
</tr>
<tr>
<td>Chiou et al.</td>
<td>1985</td>
<td>63</td>
<td>SR</td>
<td>Died, 1 month AT</td>
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<tr>
<td>Panageas et al.</td>
<td>1990</td>
<td>53</td>
<td>SR, RT</td>
<td>NED AT</td>
</tr>
<tr>
<td>Lamont et al.</td>
<td>1991</td>
<td>52</td>
<td>SR, RT, CT</td>
<td>Died, ~3 months AT</td>
</tr>
<tr>
<td>Matthew et al.</td>
<td>2006</td>
<td>55</td>
<td>SR, CT</td>
<td>NED, 2 years AT</td>
</tr>
<tr>
<td>Guo</td>
<td>2009</td>
<td>65</td>
<td>SR</td>
<td>Died, 2 months AT</td>
</tr>
<tr>
<td>Khaliq et al.</td>
<td>2012</td>
<td>73</td>
<td>SR, CT</td>
<td>Died, 2 weeks AT</td>
</tr>
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</table>

Angiosarcoma of the prostate

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Age</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Matthias et al.</td>
<td>1889</td>
<td>70</td>
<td>PC</td>
<td>Died, 6 months AT</td>
</tr>
<tr>
<td>Botesko et al.</td>
<td>1902</td>
<td>2</td>
<td>PC</td>
<td>Died, 1 day</td>
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<tr>
<td>Mogi et al.</td>
<td>1911</td>
<td>38</td>
<td>PC</td>
<td>Died, 4 months AT</td>
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<tr>
<td>Salleras et al.</td>
<td>1924</td>
<td>32</td>
<td>PC</td>
<td>Died, AD</td>
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<tr>
<td>Smith et al.</td>
<td>1986</td>
<td>60-42</td>
<td>SR, CT</td>
<td>Died, 6 months AD NED, 24 months AT</td>
</tr>
<tr>
<td>Chan et al.</td>
<td>1990</td>
<td>35</td>
<td>PC</td>
<td>Died, 5 weeks AD</td>
</tr>
<tr>
<td>Oliva et al.</td>
<td>2001</td>
<td>36</td>
<td>SR, CT</td>
<td>NED, 36 months AT</td>
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<tr>
<td>Chandan et al.</td>
<td>2003</td>
<td>77</td>
<td>SR</td>
<td>Died, 4 days AT</td>
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<tr>
<td>Lee et al.</td>
<td>2006</td>
<td>19</td>
<td>CT, SR</td>
<td>NED, 16 months AT</td>
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<tr>
<td>Guo et al.</td>
<td>2009</td>
<td>65</td>
<td>SR</td>
<td>Died, 2 months AT</td>
</tr>
<tr>
<td>Khaliq et al.</td>
<td>2012</td>
<td>73</td>
<td>SR, CT</td>
<td>Died, 2 weeks AT</td>
</tr>
</tbody>
</table>

AD, After diagnosis; AT, After treatment; CR, Chemoradiation; CT, Chemotherapy; NED, No evidence of disease; PC, Palliative care; RT, Radiotherapy; SR, Surgical resection.

Figure 2. Microscopic examination showed a poorly differentiated malignant tumor involving the left seminal vesicle soft tissue (A). The tumor showed large pleomorphic cells. New mitotic figures were also present. Immunostains were performed and the tumor cells were positive for FLI-1 (C) and CD31; and negative for Cam 5.2, cytokeratin 7, cytokeratin 20, actin, desmin, S100, CD117, synaptophysin, chromogranin and prostate specific antigen (PSA). These results supported the diagnosis of an angiosarcoma.

Figure 1. Imaging studies at the time of initial presentation show a mass emanating from the left seminal vesicle and invading the left posterior bladder wall and the left obturator internus muscle. The mass demonstrates contrast enhancement on axial computed tomography (A) and magnetic resonance imaging (B). The mass is hypointense on T2 weighted axial (C) and coronal (D) magnetic resonance imaging. E) Pre-operative external beam radiotherapy plan with a prescribed dose of 50 Gray delivered in 25 fractions. The gross tumor volume (GTV) is delineated in white outline and the planning target volume (PTV) is delineated in black outline. The GTV, representing the tumor as visualized on CT and MRI, was expanded by 1.5 cm to generate the clinical tumor volume (CTV). The CTV was expanded by 0.7 cm to generate the PTV.
perineal pain which may radiate to the lower back. Prostatic angiosarcoma can present with symptoms similar to prostatic adenocarcinoma, including dysuria, hematuria, or pelvic pain. Biopsy is required for definitive diagnosis. The defining histologic features of angiosarcoma are an abnormal appearance of the endothelial lining and cellular dedifferentiation. Generally, positive immunohistochemical staining for the normal endothelial lining and cellular dedifferentiation.

As angiosarcoma of the prostate or seminal vesicle is rare, no definitive treatment schemes have been established. Surgical resection is usually performed for lesions that are amenable to such an approach. However, clinical outcomes following surgical resection alone are generally poor. All 4 patients treated with this approach and reported in the literature developed distant metastases and death within one year (Table 1). In other reports, chemotherapy and/or radiotherapy have been used as an adjunct to surgery, with some patients experiencing freedom from disease recurrence, supporting the use of a multimodality treatment approach. Our patient had locally advanced angiosarcoma arising from the left seminal vesicle. He was felt to be at high risk for local and distant failure with surgical resection alone, thus we utilized multimodality therapy consisting of chemotherapy, chemoradiation, and surgical resection with IORT. He is alive and without evidence of disease recurrence for six years following completion of treatment, and we believe that this is the longest disease-free survival reported to date.

Recent phase II trials have demonstrated safety and modest efficacy of single agent paclitaxel or bevacizumab for unresectable angiosarcoma. Several case reports have demonstrated marked response of angiosarcoma to radiotherapy combined with paclitaxel or bevacizumab, with acceptable treatment-related toxicity. For patients with localized angiosarcoma of the prostate or seminal vesicle, multimodality treatment incorporating these regimens should be considered.

References