Case Report
Primary angiosarcoma of the prostate: a case report and literature review

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Abstract: Primary sarcomas of the prostate are rare, accounting for about 0.1% of all prostatic malignancies. Prostate angiosarcoma is exceedingly rare with only sixteen cases reported in the literature to date. Seven out of the sixteen cases were followed by radiotherapy for prostate adenocarcinoma. Despite the rarity, it usually presents with no detectable serum prostate-specific antigen levels and poor prognosis, representing a diagnosis and treatment challenge. The present study reports a new case of primary prostate angiosarcoma without radiation exposure history, reviewing the clinical symptoms, morphologic changes, prognosis, and treatment of angiosarcomas of the prostate.

Keywords: Prostate, angiosarcoma, primary

Introduction

Primary sarcomas of the prostate are rare, accounting for about 0.1% of all prostatic malignancies [1]. Rhabdomyosarcomas are the most frequent sarcomas of the prostate, accounting for more than 75% of all cases, typically seen in children and young adults. Angiosarcomas are malignant tumors of endothelial origin, frequently presenting in the deep soft tissue of the extremities, skin, breasts, and occasionally in different organs. It is exceedingly rare in the prostate with only sixteen cases reported in the literature. Angiosarcomas have been associated with exposure to radiation and chemical agents such as thorium, arsenic, and vinyl chloride. Seven of the cases of prostate angiosarcomas, mentioned above, were related to radiotherapy [2-4]. This study reports a new case of primary prostate angiosarcoma without history of radiation exposure, reviewing the literature of primary and secondary angiosarcomas of the prostate.

Case report

This case was a 41-year-old man with progressive dysuria, urinary bifurcation, frequent urination, and increased number of defecations. Symptoms were relieved after treatment with a urinary catheter but repeated after removal of the catheter. Since that time, the patient had experienced persistent abdominal discomfort for about one year. Serum prostate-specific antigen level of the patient was 0.08 ng/mL. Rectal palpation revealed a marked enlarged prostate. A large mass involving the left transitional zone, peripheral zone of the prostate, left seminal vesicle, and bladder was seen on magnetic resonance imaging (MRI) (Figure 1A). Computed tomography (CT) scans showed no obvious abnormalities of the brain, chest, and abdomen organs.

Pathologic diagnosis of angiosarcoma was made, according to the transrectal prostatic core biopsy. Considering the size of the tumor, the patient received adjuvant chemotherapy, with four cycles of gemcitabine and paclitaxel, before the operation. However, the tumor was not sensitive to chemotherapy. One month later, he received radical cystoprostatectomy, bricker operation, and underwent radiotherapy. The patient died six months after the operation with the tumor metastasizing to the brain and liver.
Figure 1. MRI revealed an ill-defined huge mass with low signal intensity of the prostatic origin on T2WI (A). Gross examination showed the tumor nearly replaced the whole prostate parenchyma and involved the bladder. The cut-off surface of the tumor was solid and gray with focally hemorrhage and necrosis (B).

Figure 2. Histological features of the areas of well-formed anastomosing vessels formed by atypical endothelial cells (A) and areas of solid sheets without clear vasoformation (B). The tumor cells were spindle-shaped or polygonal with hyperchromatic nuclei and partial conspicuous nucleoli.

Figure 3. Immunohistochemically, the tumor cells were diffusely or focally positive for CD31 (A), CD34 (B), ERG (C) and Fli-1 (D).

Immunohistochemistry

Routine 4-μm sections were prepared from formalin-fixed and paraffin-embedded (FFPE) tissue blocks on 3-aminopropyl-triethoxysilane-treated glass slides. Antigen retrieval was achieved by boiling the sections in 0.01 mol/L citrate buffer (pH 6.0) in a high-pressure cooker for 3 minutes. Envision kit (Dako Cytomation, Glostrup, Denmark) was used for immunohistochemical staining, according to manufacturer protocol.

Primary antibodies with the following dilutions were used for immunohistochemistry: CD31 (1:50; Dako Cytomation), CD34 (1:100; Dako Cytomation), CD99 (1:100; Dako Cytomation), CgA (1:100; Zymed, San Francisco, CA, USA), Desmin (1:100; Dako Cytomation), epithelial membrane antigen (EMA, 1:100; Dako Cytomation), ERG (1:400; Zymed), Fli1 (1:100; Zymed), myogenin (1:50; Zymed), pan-cytokeratin (PCK, 1:100; Zymed), P63 (1:50; Dako Cytomation), and S-100 (1:100; Zymed).

Results

Grossly, a huge lobulating prostate with the whole bladder was obtained from radical cystoprostatectomy. The prostate measured 7.5 cm × 6.0 cm × 6.0 cm, with a huge mass nearly completely replacing the whole prostate and invasion of the bladder. The cut-off surface of the tumor was solid and gray. Hemorrhaging and necrosis were seen focally (Figure 1B).

Microscopically, the tumor displayed a wide range of morphological appearances, ranging from areas of well-formed anastomosing vessels to solid sheets without clear vasoformation. Vasoformative areas consisted of ramifying channels lined by atypical endothel-
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## Table 1. Angiosarcomas of prostates reported in the literature

<table>
<thead>
<tr>
<th>Case</th>
<th>Publication year</th>
<th>Author</th>
<th>Patient age (years)</th>
<th>History of RT</th>
<th>Symptoms</th>
<th>Immunophenotype</th>
<th>Treatment</th>
<th>Clinical outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>1963</td>
<td>Shapira</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
</tr>
<tr>
<td>2.</td>
<td>1986</td>
<td>Smith</td>
<td>60</td>
<td>NM</td>
<td>LUTS</td>
<td>Factor VIII (+)</td>
<td>Radical cystoprostatectomy</td>
<td>Dead, 6 months</td>
</tr>
<tr>
<td>3.</td>
<td>1986</td>
<td>Smith</td>
<td>42</td>
<td>NM</td>
<td>Pain</td>
<td>Factor VIII (+)</td>
<td>Radical cystoprostatectomy + chemotherapy</td>
<td>Alive, 24 months</td>
</tr>
<tr>
<td>4.</td>
<td>1989</td>
<td>Wick</td>
<td>63</td>
<td>NM</td>
<td>NM</td>
<td>Vimentin (+), Keratin (-), EMA (-), PSA (-)</td>
<td>Radical prostatectomy + radiotherapy</td>
<td>Alive, 15 months</td>
</tr>
<tr>
<td>5.</td>
<td>1990</td>
<td>Chan</td>
<td>35</td>
<td>NM</td>
<td>Pain + Hematuria + LUTS</td>
<td>Factor VIII (+), vimentin (+)</td>
<td>NM</td>
<td>Dead, 5 weeks</td>
</tr>
<tr>
<td>6.</td>
<td>1992</td>
<td>Russo</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
</tr>
<tr>
<td>7.</td>
<td>2000</td>
<td>Luque Barona</td>
<td>78</td>
<td>NM</td>
<td>NM</td>
<td>NM</td>
<td>Surgery (no further details)</td>
<td>Dead, 9 months</td>
</tr>
<tr>
<td>8.</td>
<td>2001</td>
<td>Oliva Encina</td>
<td>31</td>
<td>NM</td>
<td>LUTS</td>
<td>Factor VIII (+), CD31 (+)</td>
<td>Radical prostatectomy + chemotherapy + radiation therapy</td>
<td>Alive, 36 months</td>
</tr>
<tr>
<td>9.</td>
<td>2003</td>
<td>Chan dan</td>
<td>77</td>
<td>Ext. RT</td>
<td>Hematuria + LUTS</td>
<td>Factor VIII (+), CD31 (+)</td>
<td>TURP</td>
<td>Dead, 4 Days after TURP</td>
</tr>
<tr>
<td>10.</td>
<td>2006</td>
<td>Lee</td>
<td>19</td>
<td>NM</td>
<td>Hematuria + LUTS</td>
<td>CD31 (+), CD34 (+)</td>
<td>Radical cystoprostatectomy</td>
<td>Alive, 16 months</td>
</tr>
<tr>
<td>11.</td>
<td>2009</td>
<td>Guo</td>
<td>NM</td>
<td>Ext. RT</td>
<td>Hematuria + perineal pain</td>
<td>CD31 (+), CD34 (+)</td>
<td>PE</td>
<td>Dead, 2 months</td>
</tr>
<tr>
<td>12.</td>
<td>2012</td>
<td>Humphrey</td>
<td>67</td>
<td>Ext. RT</td>
<td>LUTS</td>
<td>NM</td>
<td>TURP + PE</td>
<td>NM</td>
</tr>
<tr>
<td>13.</td>
<td>2012</td>
<td>Khaliq</td>
<td>73</td>
<td>Ext. RT + Brachytherapy</td>
<td>Hematuria + LUTS</td>
<td>VIII (+), CD31 (+)</td>
<td>PE + chemotherapy</td>
<td>Dead, Several months (no further details)</td>
</tr>
<tr>
<td>14.</td>
<td>2014</td>
<td>Campschoer</td>
<td>69</td>
<td>Brachytherapy</td>
<td>LUTS + perineal pain</td>
<td>CD31 (+), CD34 (+), factor VIII (+), PSA (-)</td>
<td>None</td>
<td>Dead, 4 months</td>
</tr>
<tr>
<td>15.</td>
<td>2015</td>
<td>Gupta</td>
<td>71</td>
<td>Brachytherapy</td>
<td>Hematuria + LUTS</td>
<td>Fli-1 (+), CD31 (+), CD34 (+)</td>
<td>Chemothery</td>
<td>NM</td>
</tr>
<tr>
<td>16.</td>
<td>2016</td>
<td>Wang</td>
<td>79</td>
<td>RT</td>
<td>Hematuria + LUTS</td>
<td>CD31 (+), CD34 (+), ERG (+), Fli-1 (+), c-Myc (+)</td>
<td>Radical cystoprostatectomy</td>
<td>Alive, 20 months</td>
</tr>
<tr>
<td>17.</td>
<td>The present study</td>
<td>Liu</td>
<td>41</td>
<td>No</td>
<td>LUTS</td>
<td>CD31 (+), CD34 (+), ERG (+), Fli-1 (+)</td>
<td>Radical cystoprostatectomy + chemotherapy</td>
<td>Dead, 6 months</td>
</tr>
</tbody>
</table>

Ext. RT, external radiotherapy; LUTS, lower urinary tract symptoms; NM, not mentioned; PE, pelvic exenteration; TURP, transurethral resection of the prostate.
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Disclosure of conflict of interest

None.

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References