Alveolar Rhabdomyosarcoma of the Prostate in a Young Adult Presenting with Acute Retention of Urine

Abstract

Prostate rhabdomyosarcoma is rare in adults. It is characterized by a high degree of aggressiveness, rapid local growth with the formation of large pelvic masses, often leading to urethral obstruction, and systemic spread, commonly to the lungs, liver, and bone. A 23-year-old male was referred to our department with symptoms of acute retention of urine, hematuria and constipation. He was earlier admitted in another hospital, wherein he was catheterized, evaluated and underwent both transurethral as well as transrectal biopsy of the prostate. Our case was clinically advanced, and chemotherapy treatment seemed adequate to control the disease, and reduce the symptoms and improve the quality of life.

Keywords: *Adults, alveolar, prostate cancer, retention of urine, rhabdomyosarcoma*

R. B. Nerli, Adarsh C. Sanikop[1], Vishal Kadeli, Shridhar C. Ghagane[2], Neeraj S. Dixit[2], Murigendra B. Hiremath[3]

Department of Urology, KLE Academy of Higher Education and Research (Deemed-to-be-University), KLES Kidney Foundation, KLES Dr. Prabhakar Kore Hospital and M.R.C, [1]Department of Pathology, JN Medical College, KLE Academy of Higher Education and Research (Deemed-to-be-University), KLES Dr. Prabhakar Kore Hospital and M.R.C, [2]Department of Urology, KLES Kidney Foundation, KLES Dr. Prabhakar Kore Hospital and M.R.C, Belagavi, [3]Department of Biotechnology and Microbiology, Karnatak University, Dharwad, Karnataka, India

Address for correspondence:
R. B. Nerli, Department of Urology, KLE Academy of Higher Education and Research (Deemed-to-be-University), KLES Kidney Foundation, KLES Dr. Prabhakar Kore Hospital and M.R.C, Belagavi - 590 010, Karnataka, India.
E-mail: rbnerli@gmail.com

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Introduction

Rhabdomyosarcomas, although relatively rare in adults, are the most common soft-tissue malignancy in children and adolescents, in whom they comprise approximately 60% of sarcomas reported per annum.[1] On histology, these neoplasms appear to be analogous to myogenesis as seen in the developing embryo. It would be more correct to define rhabdomyosarcoma as a tumor derived from primitive mesenchyme and exhibiting a profound tendency toward myogenesis than to define it as a cancer arising from skeletal muscle. Masson[2] referred these rhabdomyosarcomas as "rhabdopoietic sarcomas," as it better explained the observation that these tumors arose more from the viscera and axial soft tissues than from the extremities.[3] Historically, this appears to be a well-established phenomenon, considering Stafford's report in 1839 of a probable prostatic rhabdomyosarcoma in a 5-year-old child, and the first series of these tumors reported in 1894, in which the majority of cases involved the urogenital tract.[4,5]

Newton *et al*.[6] at the intergroup rhabdomyosarcoma study (IRS) undertook a huge task of testing various systems of classification and authored a new international classification. The ultimate goal of this group was to stratify rhabdomyosarcoma into prognostically significant and diagnostically consistent morphologic subgroups. The IRS Pathologic Review Committee (now part of the Children's Oncology Group) has continued to use this system to the present time.[7] Rhabdomyosarcoma has three recognized histologic variants: embryonal, alveolar, and pleomorphic. The embryonal subtype is the most common and with the most favorable outcome. The pleomorphic and the alveolar variants are less frequent and more aggressive. The international classification has retained the diagnostic features used for embryonal and alveolar variants. The category of alveolar rhabdomyosarcomas has been expanded to include solid variants that lack fibrous septa.

Rhabdomyosarcoma of the prostate commonly occurs in children and rarely is reported in patients above the age of 18 years[8] and commonly presents with symptoms of dysuria and obstruction to urinary flow. It is characterized by rapid growth leading to symptoms of bladder outlet obstruction and/or rectal compression. The lungs, liver, and bones are the common sites of metastases. The prostatic acid phosphatase and serum prostate-specific antigen (PSA) levels are usually normal. We report a case of rhabdomyosarcoma of the prostate in a young adult presenting with acute retention of urine.

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Case Report

A 23-year-old male was referred to our department with symptoms of acute retention of urine, hematuria, and constipation. He was earlier admitted to another hospital, wherein he was catheterized, evaluated, and underwent both transurethral as well as transrectal biopsy of the prostate. Following biopsy, the patient had gross hematuria and hence referred to our center. On examination, the patient had a suprapubic mass, firm, and nontender. Per-rectal examination revealed a large prostatic growth in continuation with the suprapubic mass. Prostate was firm and nontender.

Hemoglobin was 7.9 g %, serum creatinine was 0.79 mg%, serum PSA was 1.1 mg%, and liver function tests were within normal range. Computed tomography (CT) revealed a large heterogeneously enhancing mass lesion involving prostate gland measuring 9.6 cm × 7.6 cm × 9.4 cm with nonenhancing cystic/necrotic areas [Figure 1a and b]. Posteriorly, the lesion caused displacement of rectum with loss of fat planes. Superiorly, the lesion extended into the bladder base and laterally the lesion was abutting the right obturator internus muscle. A large necrotic left obturator lymph node measuring 3.4 cm × 3.2 cm was noted. An enlarged right periprostatic lymph node measuring 1.7 cm × 1.5 cm was also noted. Few well-defined soft-tissue density nodules were also visualized bilaterally in the lungs suggestive of metastases.

Magnetic resonance imaging (MRI) revealed a large dense heterogeneously enhancing soft-tissue mass lesion predominantly T1 isointense and T2 hyperintense, with central areas of hypointensity suggestive of necrosis. The prostate gland was not separately visualized from the lesion. Altered signal intensities noted in the right ala of sacrum suggestive of metastases. There were multiple pleural and pulmonary nodular lesions bilaterally suggestive of metastases.

Histopathological report of the biopsy specimens revealed a poorly differentiated tumor involving the parenchyma of the prostate [Figure 2a and b]. It comprised cellular nodules of round or oval immature appearing cells with negligible cytoplasm, indistinct cytoplasmic borders, and hyperchromatic nucleus. Few tumor cells showed dense, eosinophilic cytoplasm (rhabdomyoblasts). Necrosis was also seen. The tumor cells expressed Desmin and Myogenin and were immunonegative for cytokeratin, epithelial membrane antigen, CD 117, leukocyte common antigen, synaptophysin, chromogranin A, smooth muscle actin, and S-100 protein and CD 34 [Figure 3a-d]. A final diagnosis of rhabdomyosarcoma; solid alveolar subtype was made. In view of the metastatic disease, the patient was initiated on chemotherapy using three drugs, namely, cyclophosphamide, doxorubicin, and vincristine. The patient has received three cycles of the same (as of now), and the mass has markedly reduced in size.

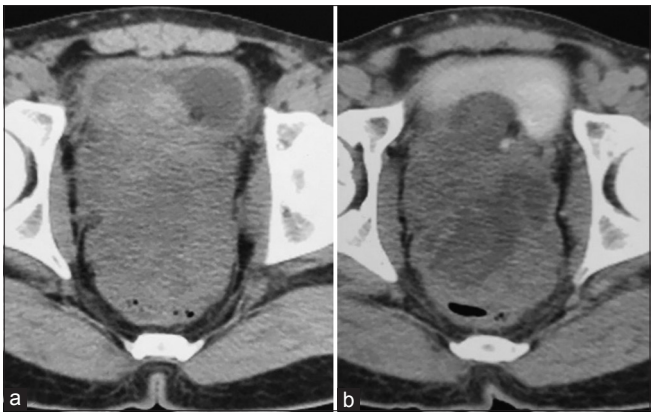

Figure 1: (a and b) Computed tomography kidneys, ureters, and bladder plain and contrast shows a large heterogeneously enhancing mass lesion involving the prostate 9.6 × 7.6 × 9.4. There is a nonenhancing cystic/necrotic area at the center

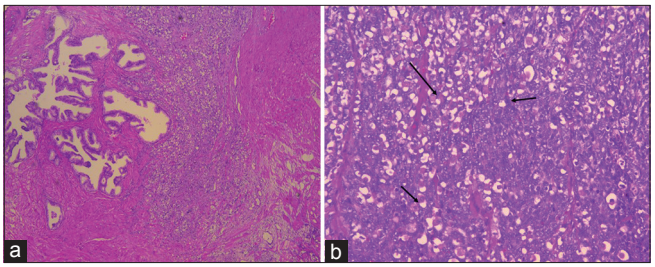

Figure 2: (a) H and E, ×40 sections show a focus of normal prostate on the left side and sheets of neoplastic cells infiltrating the stroma in the right side, (b) rhabdomyoblasts H and E, ×200 sections show sheets of rhabdomyoblasts. These cells have an eccentrically placed nuclei and abundant eosinophilic cytoplasm

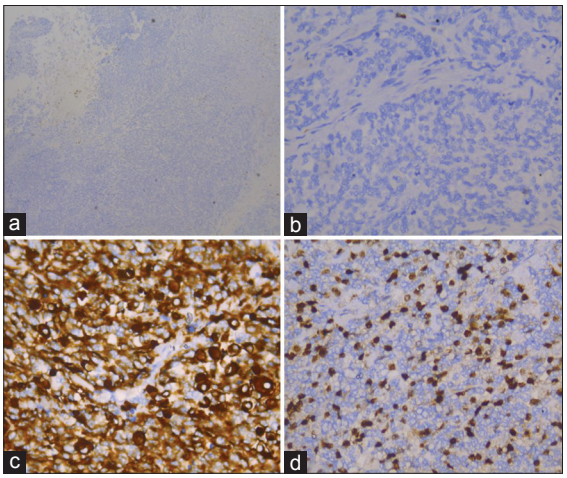

Figure 3: (a) Immunohistochemistry shows the neoplastic cells do not express cytokeratin, (b) immunohistochemistry shows the neoplastic cells do not express prostate-specific antigen, (c) Desmin ×400, the neoplastic cells express strong desmin positivity (cytoplasmic), (d) Myogenin ×400, the neoplastic cells express strong Myogenin (nuclear)

Discussion

In adults, rhabdomyosarcoma is very rare and clinical experience is largely limited to case reports.[9] Three histological subtypes have been identified: Embryonal with botryoid and alveolar histiotypes, generally affecting

young children and adolescents, and the pleomorphic histiotype that is usually encountered in adult patients.[10] The primary prostate rhabdomyosarcoma is rare in adults, and its clinical manifestations, natural history, prognosis, and treatment options can be found only in anecdotal descriptions. The presenting symptoms are often related to urethral obstruction, with frequency, hesitancy, and dysuria as predominant symptoms or, less often, with hematuria and acute urinary retention. In some patients, the compression of the rectum can cause constipation, rectal bleeding, and a sense of rectal fullness. It is usually characterized by extensive locoregional spread, with symptoms of urinary obstruction (due to bladder outlet obstruction), and a tendency to metastasize by bloodstream and regional lymphatics and to give early lung and bone involvement.

There are no pathognomonic radiological findings for prostate rhabdomyosarcoma. CT scan and MRI study usually reveal a large soft-tissue mass with areas of necrosis replacing the whole prostate, but the radiological differential diagnosis with prostate adenocarcinoma can be very difficult. The mass may be shown to invade periurethral and perivesical tissue, or it may even extend into the ischiorectal fossa. Calcifications are rare seen. MRI clearly shows the site of origin as the central prostate area, with compression of surrounding peripheral portion. The mass displays a well-defined low signal intensity pseudocapsule in T2-weighted images and a heterogeneous enhancement in postgadolinium T1-weighted images. The utilization of positron emission tomography-CT for rhabdomyosarcoma staging in adults is still to be consolidated in the literature; however, some reports have demonstrated the relevance of this method in the detection of the primary focus in metastatic disease or obscure metastases.

Due to the rarity of adult rhabdomyosarcoma, information regarding its clinical and biologic characteristics is very limited; large, multi-institutional trials have not been performed as yet, and only reports from single institutions are noted in the literature.[11] Moreover, a direct comparison between adult and pediatric patients, critical to understanding factors responsible for the different outcomes, has not been performed. In pediatric patients, older age has been associated with poorer outcome,[12] and available information suggests that unfavorable prognostic variables (e.g., alveolar subtype and regional and distant spread) are more frequent in adults than in children.[12] Sultan *et al.*[13] analyzed data from 1071 adults (age >19 years) and 1529 children (age ≤19 years) reported in the public access database as having rhabdomyosarcoma, diagnosed from 1973 to 2005. Adults with rhabdomyosarcoma had significantly worse outcome than children (5-year overall survival rates, 27%, 1.4% and 61%, 1.4%, respectively; $P < 0.0001$). Tumors in adults were more likely to be at an unfavorable site (65% vs. 55%; $P < 0.0001$) and to have histologies that are unusual during childhood, particularly the pleomorphic subtype (19%) and not otherwise specified (43%). Regional and distant spread was not more frequent in adults. Adults had significantly worse outcome than children with similar tumors. The most significant difference was in localized disease; 5-year survival estimates were 82% and 2.0% for children and 47% and 2.9% for adults ($P < 0.001$). Multivariate analysis showed that age, histologic subtype, primary site location, stage, and local control with surgery and/or radiation were significant predictors of survival. However, alveolar subtype and unfavorable primary site lost significance when analysis was restricted to adults.

The diagnosis is usually based on biopsy and histopathological evaluation of the same, which can reveal variable grades of differentiation along the myogenesis pathway. The treatment largely depends on the disease stage at the time of presentation. A localized prostate disease can be treated with radical surgery.[14] Unfortunately, prostate rhabdomyosarcomas are often caught late in the disease process, and the tumor size is huge and extensive, as in our patient's case. Patients with nonmetastatic large prostate rhabdomyosarcoma, which is initially not resectable, can be treated with neoadjuvant chemoradiation therapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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