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Rhabdomyosarcoma of the urinary bladder in an adult: Case report and review of the literature

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A B S T R A C T

Rhabdomyosarcomas with bladder involvement in adults is an extremely rare tumor with approximately 35 cases reported. Because of its rarity in adults the exact treatment modalities are not entirely clear. Treatment is based on Children's Oncology Group in RMS. There is no significant difference in stage, lymph node status, gender, and site between adults and children. The 5 year survival for pediatric RMS is 66% and for adults 22%. It is clear that our understanding and treatment of pediatric RMS is much greater than that of adult patients and the possible reason for these differences are discussed.

1. Introduction

Transitional cell carcinoma is the fourth most common malignancy in the United States. Risk factors include age, male sex, smoking and environmental toxins. The overall 5 year survival rate is 77%. Only 35 cases of adult rhabdomyosarcoma of the bladder have been reported with a survival rate of 22%. In children the 5 year survival is 66%. The diagnosis, treatment and possible reasons for the differences in the above outcomes are discussed.

2. Case report

A seventy-year-old male presented to the urologist with gross hematuria. CT Urogram demonstrated an ill-defined mass in the anterior bladder wall measuring approximately 11.1 × 6.2 cm with associated bilateral hydronephrosis (Fig. 1). A TURBT was attempted but the tumor was deemed unresectable via a transurethral approach. Workup prior to surgery did not demonstrate any metastatic disease. The patient subsequently underwent radical cystoprostatectomy with creation of an ileal conduit. 3 months after discharge, an MRI revealed a 15 × 8.4 × 8.3 cm pelvic mass with direct invasion of the rectum and extension to the base of the penis and metastatic disease to the lungs. The patient recieved a regimen of GTA (gemcitabine/taxol/adriamycin) which he completed. He was switched to a conjugate of IgG and monomethyl Auristatin. He is currently living with metastatic disease.

The surgical specimen revealed a 15 cm bladder mass with a high-grade malignant neoplasm. The hematoxylin and eosin stain (Fig. 2) demonstrates a small blue cell tumor and the initial differential included sarcomatoid carcinoma, urothelial carcinoma with neuroendocrine features, neuroendocrine tumors, and lymphomas.[1] IHC is essential in differentiating these tumors and IHC ruled them out. The positive CD 56 (Fig. 3) marks several different neoplasms including rhabdomyosarcoma (RMS). Because of the positivity for CD 56, IHC for myogenin and desmin was performed and both were positive, so the diagnosis of rhabdomyosarcoma was made. The surgical margins were negative but there was focal extravesical extension and foci of vascular invasion. All resected lymph nodes were negative for RMS.

3. Discussion

Rhabdomyosarcoma is the most common childhood soft tissue malignancy and accounts for 6% of all childhood malignancies.[2] It is rare in adults, accounting for only 2–5% of adult soft tissue tumors.[3] Bladder involvement by RMS is extremely rare with approximately 35 cases reported. Because of its rarity, exact treatment modalities are unknown. The Children's Oncology Group Soft Tissue Sarcoma Committee utilized a combination of RT, chemotherapy, and surgery, and the 5-year survival rates have improved success rates from 55% to 71%[2] but treatment regimens used in children have not been as successful in adults. The overall 5-year survival for adult RMS in the kidney, bladder, and prostate is 17%, 22%, and 33% respectively.[3] In contrast, pediatric RMS involving the bladder has a 5-year survival of 66% percent and 22% in

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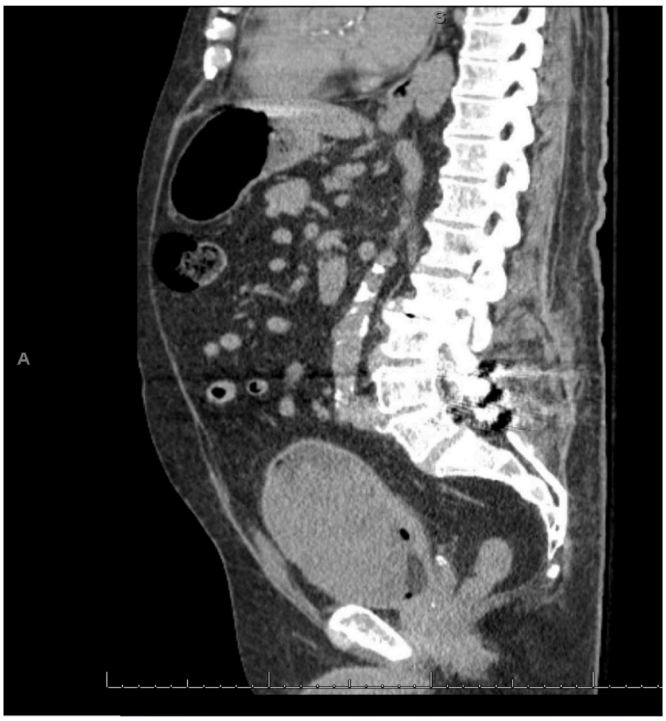

Fig. 1. CT urogram, sagittal view, demonstrating large intravesical tumor.

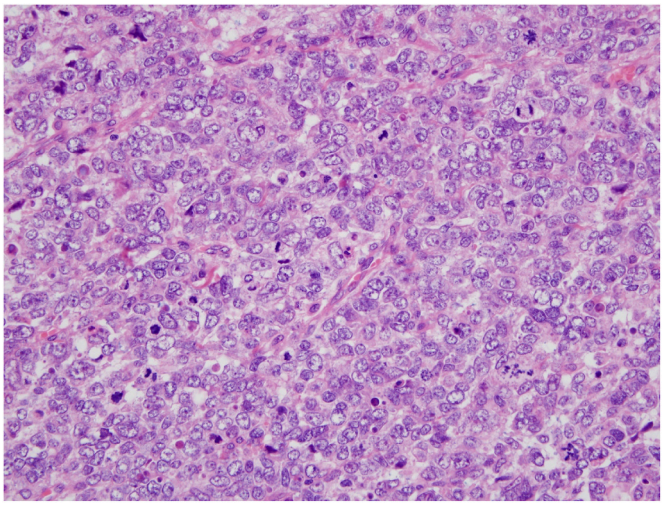

Fig. 2. H&E slide demonstrating small blue cell tumor with numerous mitotic figures. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

adults.[4]

Currently most treatment regimens in adults consist of surgery, chemotherapy, and radiotherapy in combination or as single/dual modalities, but as Nguyen noted there is no standardized treatment regimen for adults as there is in children.[4] Our patient demonstrated a 15 cm recurrent pelvic mass that involved the rectum; based on the recommendations of our multimodality tumor board, he was treated with gemcitabine/taxol/adriamycin with Neulasta x 6 cycles and then was started on PADCEV. Liu et al. found that patients receiving greater than 19 weeks of chemotherapy had a 90% OS compared to patients who received less than 19 weeks who had an OS of 0%. According to the SEER study,[3] adult patients who underwent surgery had a significantly better prognosis. There is no difference in the stage at the time of presentation,[3] nor is there a difference in sex, histology, stage, or lymph node status between children and adults.

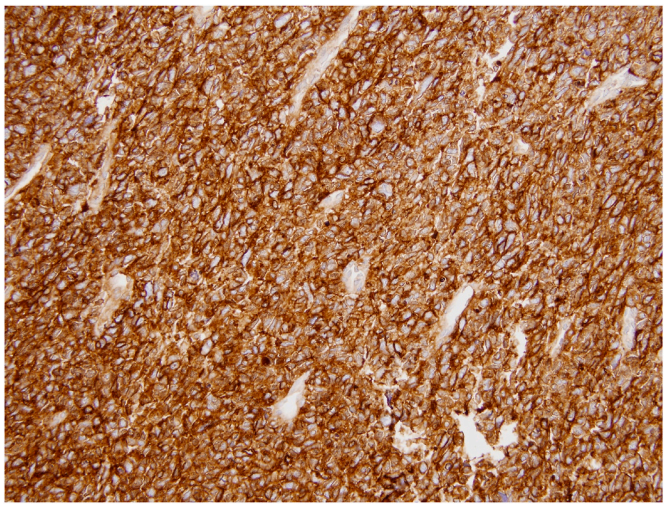

Fig. 3. IHC for CD 56 demonstrating diffuse strong positivity.

Several researchers have found that histologic subtype is a factor in prognosis. Liu found that the prognosis for patients with the pleomorphic subtype was much worse and that the proportion of the pleomorphic subtype increases with age. Another possibility raised by Gordon[5] is that pleomorphic RMS may represent a different type of tumor based on chromosomal analysis. This would not be surprising as many neoplasms have been reclassified based on chromosomal analysis.

4. Conclusions

Rhabdomyosarcoma in adults is rare and because of its rarity the exact treatment modalities are not entirely clear. Adults have about one third the survival rate of children with RMS. There is no significant difference between adults and children regarding stage, site, gender, and lymph node status, so these clinical parameters may not be the reasons for the differences in outcomes between the two populations. Some possible reasons include: 1) There is no standardized universally accepted treatment protocol for adults 2) The dosage of chemotherapy in adults varies greatly 3) The histologic classification of the tumor needs to be standardized 4) Some cases of RMS in adults may be reclassified if chromosomal analysis were performed. We need more thorough, standardized data because there is a significant difference between our understanding and treatment of pediatric RMS compared to adult RMS.

Declaration of competing interest

None.

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