Embryonal Rhabdomyosarcoma of the Prostate in a Young Adult

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ABSTRACT

Embryonal rhabdomyosarcoma (ERMS) is a primitive and aggressive soft tissue tumor that arises from premature mesenchymal cells and accounts for less than 1% of prostate malignancies. We present the case of a 26-year-old Filipino male who initially sought consultation and work-up for acute urinary retention. Although his prostate-specific antigen (PSA) level was normal, a kidney-ureter-bladder (KUB) ultrasound revealed an incidental finding of an enlarged prostate gland. Subsequent imaging tests (MRI and CT) identified a large, partially exophytic mass on the prostate with intravesical extension. The patient underwent a biopsy via transurethral resection at another institution, which yielded findings of a poorly differentiated carcinoma, primarily suggesting prostatic adenocarcinoma with a Gleason score of 10 (5+5). A request for slide review was submitted to our institution, and subsequent immunohistochemistry (IHC) studies—highlighting the tumor's immunoreactivity to myogenin and desmin—confirmed the diagnosis of ERMS. In this report, we discuss the clinical features, pathogenesis, treatment, diagnosis, and prognosis of this rare prostate tumor.

***Keywords**: Embryonal rhabdomyosarcoma (ERMS), Immunohistochemistry (IHC), Prostate, Young adult*

INTRODUCTION

Embryonal rhabdomyosarcoma (ERMS) is a rare mesenchymal tumor that originates from undifferentiated mesenchymal cells and accounts for less than 1% of all prostate malignancies.[1] This tumor is predominantly found in the pediatric population, typically occurring in paratesticular sites, and exhibits a wide spectrum of morphologic features, including round and spindled cell morphology. In adults, it is rare and most commonly arises in the head and neck region and tends to display a more aggressive behavior with high recurrence and metastasis rates.[2]

Tumor occurrence in the prostate of adults is even rarer and highly unusual, with only a few cases published worldwide. To date, only eight cases of prostatic ERMS in young adults (ages 18 to 35) have been reported. The prostate is considered an exceptionally uncommon and unfavorable site for ERMS, and the definitive diagnosis requires confirmatory immunohistochemical (IHC) studies, making early histopathologic diagnosis particularly challenging.[2-3] To our knowledge, this is the first reported case of prostatic ERMS diagnosed in a young adult in the Philippines.

CASE ILLUSTRATION

This is a case of a 26-year-old Filipino male who presented with sudden onset, severe suprapubic pain associated with urinary retention. The retention was characterized by frequent episodes of incomplete and painful voiding.

Clinical history revealed that the patient had begun experiencing intermittent episodes of urinary difficulty five months prior and had already been prescribed Tamsulosin, which provided partial symptom relief. The abrupt onset of severe suprapubic pain prompted the patient to seek emergency consultation at a local hospital in their province, Nueva Ecija. Initial management was directed toward acute urinary retention and complicated urinary tract infection; however, the patient demonstrated only minimal clinical improvement despite completion of antibiotic treatment and Foley catheter insertion. A KUB ultrasound revealed an incidental finding of an enlarged prostate gland, although the patient had a normal PSA level (1.82 ng/mL). Subsequent multiparametric MRI with contrast identified a large, circumscribed, partially exophytic mass (5.4 × 4.6 × 4.4 cm) on the right side of the prostate, demonstrating internal areas of pre-existing high T1 signal, likely representing blood products, and heterogeneous contrast enhancement with central necrosis. A low T2 signal rim was also noted, likely representing a pseudocapsule. There was marked leftward compression of the adjacent low rectum, but with no definite tumor infiltration. The seminal vesicles were mildly displaced superiorly (right more than the left). There were no pathologically enlarged lymph nodes, ascites, or focal enhancing bone lesions, as confirmed by bone scintigraphy. The kidneys, ureters, and urinary bladder were normal. A radiologic impression of granulomatous prostatitis versus a primary malignancy was given. The patient later underwent TURP in the same provincial hospital, where findings revealed a poorly differentiated carcinoma, primarily considered prostatic adenocarcinoma with a Gleason score of 10 (5+5). Given the rarity of prostate cancer in this age group,[4-5,] a second opinion and slide review were requested and directed at our tertiary institution by the attending clinician.

Three months post-TURP, during the patient's transit to Metro Manila, his symptoms progressed, now accompanied by constipation and rectal pain. Upon arrival at our hospital, further worsening of his suprapubic pain and

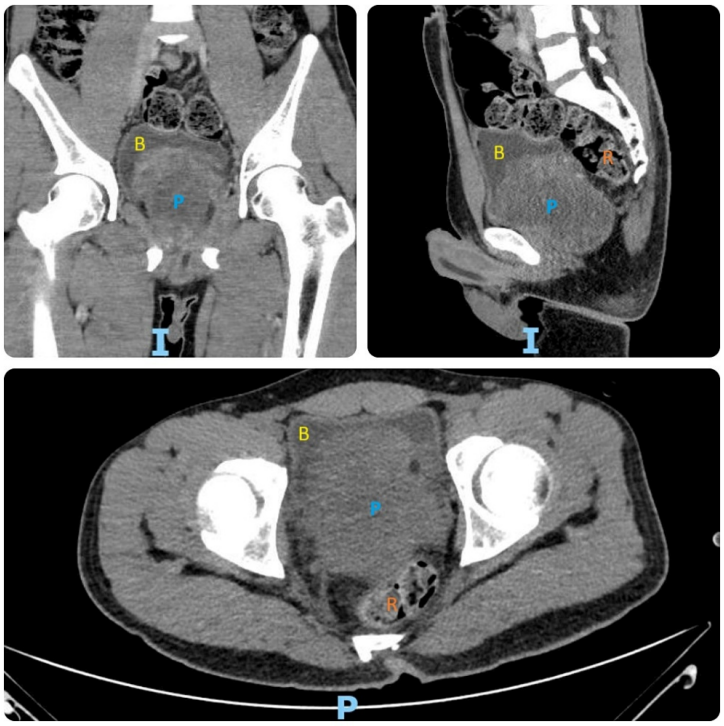

Figure 1. CT scan of the whole abdomen: Coronal view (top-left), Sagittal view (top-right), and Axial view (bottom). Revealing a markedly enlarged prostate with irregular borders and areas of hypodensity, measuring 8.0x6.7x11.1 cm (AP x T x CC). It exhibits intravesical extension through the posteroinferior urinary bladder wall and compresses the distal rectum posteriorly, causing severe luminal narrowing. P – prostate, B – bladder, R – rectum.

gastrointestinal complaints led to an assessment of a partial bowel obstruction, suspected to be secondary to his prostate malignancy. Non-contrast and contrast-enhanced axial CT scan of the whole abdomen was done, revealing a markedly enlarged prostate with irregular borders and areas of hypodensity, measuring 8.0x6.7x11.1 cm (AP x T x CC). It exhibits intravesical extension through the posteroinferior urinary bladder wall and compresses the distal rectum posteriorly, causing severe luminal narrowing (**Figure 1**). While awaiting the pathology results from the slide review of the patient's previous prostate biopsy—the one originally performed in the province and now being re-evaluated at our institution to confirm whether the lesion really represents an early-onset carcinoma or an alternative tumor type—a diverting colostomy was scheduled as part of interim management.

DISCUSSION

On slide review, histology revealed a well-delineated tumor border, tumoral hyper- and hypocellular areas, along with perivascular condensation in a background of loose, fibromyxoid stroma (**Figure 2A-C**). The individual cells were poorly differentiated, exhibiting enlarged, round, hyperchromatic nuclei (**Figure 2C**). Tumor classes considered in the differential diagnosis for a young adult prostate included hematolymphoid (e.g., DLBCL), mesenchymal (e.g., ERMS), and epithelial (e.g., adenocarcinoma) tumors.[6] Prioritizing the ruling-in of these differentials before considering other non-epithelial tumors formed the basis for selecting specific IHC stains: CD20, Myogenin, Desmin, and CK. Immunoreactivity to Desmin and Myogenin confirmed a rhabdomyoblastic differentiation pattern (focal or less diffuse staining pattern) characteristic of ERMS (**Figure 2D-F**). This contrasts with alveolar RMS, which exhibits a different morphology and a more diffuse staining pattern. Testing for gene fusion (e.g., PAX3-FOXO1A) is reserved for prognostication and for challenging cases where excluding a mixed embryonal-alveolar subtype is necessary. In this present case, our patient tolerated the diverting colostomy procedure well with good pain control and with significant improvement of his symptoms. Plans for metastatic evaluation and initiation of chemotherapy (VAC-IE regimen) were promptly made following the confirmation of ERMS diagnosis. However, the patient chose to pursue these interventions at a medical facility in his home province, resulting in loss to follow-up.

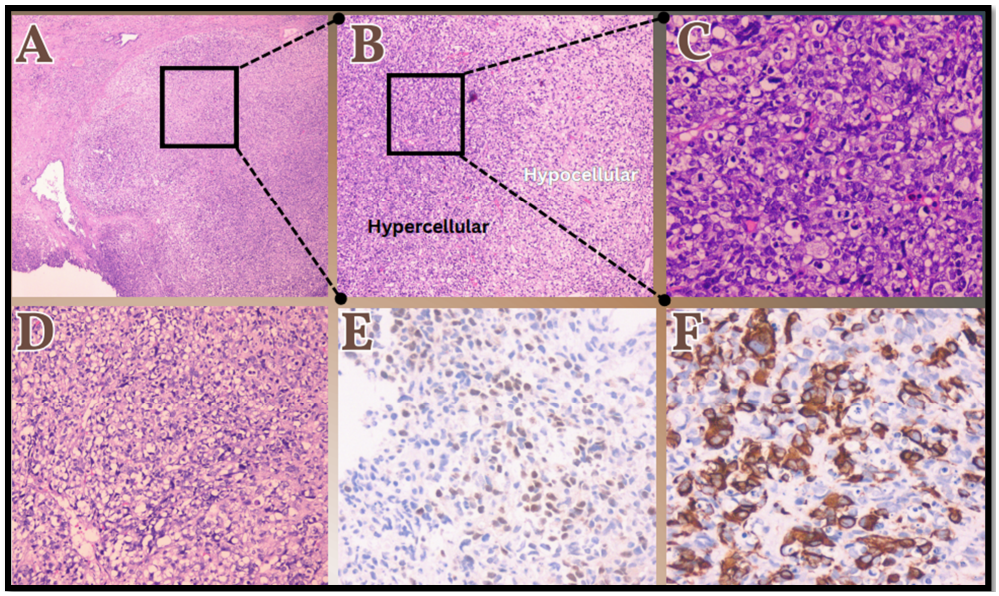

Figure 2. Scanner (A), low-power (B), and high-power (C and D) views of the tumor showing alternating hyper- to hypocellular areas with fibromyxoid stroma. Immunoreactivity to Myogenin (E) and Desmin (F) confirming rhabdomyoblastic differentiation.

Adult ERMS occurs only sporadically and is a very rare event compared to pediatric ERMS, which has few reported syndromic associations (e.g., Beckwith-Wiedemann (11p15.5), Li-Fraumeni (p53), Costello (RAS), Neurofibromatosis (NF1), Noonan (PTPN11), Dicer1, and Gorlin (PTCH1)).[7-8] Adults also differ in site predilection and prognosis (**Figure 3**). Children respond better to treatment than adults since established protocols for staging and management exist for them (e.g., Children's Oncology Group,[8] intergroup rhabdomyosarcoma study group, and International Society of Pediatric Oncology Malignant Mesenchymal Tumor Group). Treatment includes chemotherapy, with or without surgery, and/or radiotherapy. The usual adapted chemotherapy regimen includes vincristine, dactinomycin, and cyclophosphamide (VAC), though one study had reported better applicability of ifosfamide-based chemotherapy in young adults due to improved tolerance, hence highlighting the need for more clinical trials.[7]

Nearly all ERMS display loss of heterozygosity in region 11p15.5, which includes genes for IGF2, H19, and CDKN1C. Other molecular defects include alterations in p53, RAS, CDKN2A, Rb1, MYCN, MET, and ALK genes.[2] Conversely, the characteristic chromosomal translocations seen in alveolar RMS (causing chimeric transcription factors PAX3-FOXO1 or PAX7-FOXO1) are not observed in ERMS.[3,9]

Among the eight case reports of prostatic ERMS in young adults (ages 18-35), only one had conveyed cured disease for 49 months after chemotherapy completion, one was lost to follow-up, and the remaining had died in <1.5 years after diagnosis (survival range is 5-17 months, median: 11).[10] This is considerably lower than the median survival of 29 months among adult patients with ERMS not specifically located in the prostate.[11] Poor prognosis in adults is linked to factors like unfavorable location, aggressive subtype (mixed & spindle cell), advanced disease (high recurrence & metastasis), & lack of standardized treatment.[12]

Treatment varies by stage and extent of the disease. For localized disease, surgery (radical prostatectomy with pelvic lymph node dissection) may be recommended. Radiation is often added for high-risk cases, such as those with extraprostatic extension and positive margins. Chemotherapy is crucial since 25% of embryonal RMS cases have spread at diagnosis (metastatic disease constitutes half of the cases in young adults). Treatment side effects can include infertility, so young adults should be pre-informed and provided opportunities

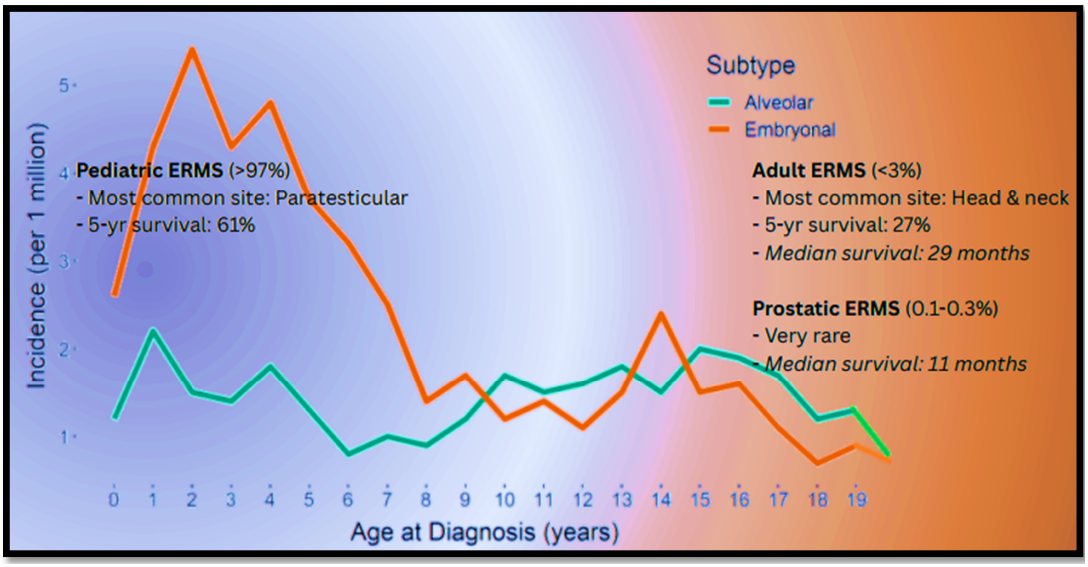

Figure 3. Incidence of ERMS by Age (Peak: 0-5). Adult ERMS (which commonly occurs in the head & neck) is rare and has a poorer prognosis and survival compared to pediatric ERMS (which commonly occurs in paratesticular sites).[13] The prostate is considered an even more uncommon and unfavorable site.

like sperm preservation.[18] Other side effects include hematologic, cosmetic, and neurologic toxicity, growth delays, bowel obstruction, and cardiomyopathy.

CONCLUSION

ERMS can rarely occur in the adult prostate, which carries a very poor prognosis with high recurrence and metastasis. Given the rarity and aggressiveness of this tumor, clinicians should maintain a high index of suspicion in young adults presenting with a prostatic mass to ensure timely diagnosis, avoid delay in appropriate management, and prolong patient survival.

CONFLICT OF INTERESTS

The authors have no conflict of interest to declare related to this study.

ACKNOWLEDGMENTS

The authors acknowledge Dr. Marcelo Severino Imasa for his contribution to this report.

REFERENCES

1. Luo H, Weng X, Zhong Z, et al. Spindle cell embryonal rhabdomyosarcoma of the prostate in an adult patient: a case report and review of the literature. J Int Med Res. 2024;52(3):03000605241234755. doi: 10.1177/03000605241234755
2. Schildhaus HU, Lokka S, Fenner W, et al. Spindle cell embryonal rhabdomyosarcoma of the prostate in an adult patient: a case report and review of the literature. Diagnostic Pathology. 2016;11(1):56. https://doi.org/10.1186/s13000-016-0507-1
3. Morgenstern DA, Rees H, Sebire NJ, Shipley J, Anderson J. Rhabdomyosarcoma subtyping by immunohistochemical assessment of myogenin: tissue array study and review of the literature. Pathol Oncol Res. 2008 Sep;14(3):233-8. doi: 10.1007/s12253-008-9012-5. Epub 2008 May 21. PMID: 18493875.
4. Bleyer A, Spreafico F, Barr R. Prostate cancer in young men: An emerging young adult and older adolescent challenge. Cancer. 2020 Jan 1;126(1):46-57. doi: 10.1002/cncr.32498. Epub 2019 Sep 25. PMID: 31553489.
5. Gupta S, Gupta A, Saini AK, Majumder K, Sinha K, Chahal A. Prostate cancer: How young is too young? Curr Urol. 2017;9(4):212-215. doi: 10.1159/000447143. Epub 2016 Dec 26. PMID: 28413383; PMCID: PMC5385860.
6. Chang, JM, Lee HJ, Lee SE, Byun S-S, Choe GY, Kim SH, Seong CK, Unusual tumours involving the prostate: radiological–pathological findings, British Journal of Radiology. 2008;81(971):907–15. https://doi.org/10.1259/bjr/68294775
7. Singh A, Phulware RH, Kumar A, Kishore S. Embryonal rhabdomyosarcoma of the prostate: Clinico-Pathological hHighlights with review of literature. J West Afr Coll Surg. 2024;14(2):225-8. doi: 10.4103/jwas.jwas_36_23. Epub 2024 Feb 22. PMID: 38562397; PMCID: PMC10980313.
8. Egas-Bejar D, Huh WW. Rhabdomyosarcoma in adolescent and young adult patients: Current perspectives. Adolesc Health Med Ther. 2014;15:115-25.
9. Treetipsatit J, Kittikowit W, Zielanska M, Chaipipat M, Thorner PS, Shuanshoti S. Mixed embryonal/alveolar rhabdomyosarcoma of the prostate: Report of a case with molecular genetic studies and literature review. Pediatr Dev Pathol. 2009;12:383-9.
10. Konno M, Mitsuzuka K, Yamada S, et al. A case of adult metastatic rhabdomyosarcoma of the prostate cured by long-term chemotherapy with local radiation. Urol Int. 2019;102:118-21.
11. Johnson J, Brant A, Sze C, Burgos AM, Sheuka N, Scherr D. Embryonal rhabdomyosarcoma of the adult prostate: case report and review. Urol Case Rep. 2021;40:101953. doi: 10.1016/j.eucr.2021.101953. PMID: 34900598; PMCID: PMC8640110.
12. Alenezi S, Zahir M, Alenezi J. Embryonal rhabdomyosarcoma of the prostate in a young male: A rare case report. Int J Surg Case Rep. 2023;106:108228. doi: 10.1016/j.ijscr.2023.108228. Epub 2023 Apr 18. PMID: 37086503; PMCID: PMC10154731.
13. Sultan I, Qaddoumi I, Yaser S, Rodriguez-Galindo C, Ferrari A. Comparing adult and pediatric rhabdomyosarcoma in the Surveillance, Epidemiology, and End Results program, 1973 to 2005: An analysis of 2,600 patients. J Clin Oncol. 2009;27:3391-7.
14. Niimi K, Hashimoto Y, Kurokawa S, Okada A, Tozawa K, Kohri K. Embryonal rhabdomyosarcoma of the prostate. Int J Clin Oncol. 2010;15:93-6.
15. Latz S, Ellinger J, Goltz D, et al. Spindle cell rhabdomyosarcoma of the prostate. Int J Urol. 2013;20:935-7.
16. Andhika DP, Hardjowijoto S. Embryonal rhabdomyosarcoma of the prostate in an adult: A rare case report. Folia Medica Indonesiana. 2021;57(2): 177–85. doi: 10.20473/fmi.v57i2.21594
17. Mao K, Li P, Yuan Y, et al. Embryonal rhabdomyosarcoma of prostate combined with prostatic abscess in an adult patient: a case report and literature review. Transl Androl Urol. 2024 Sep 30;13(9):2146-52. doi: 10.21037/tau-24-117. Epub 2024 Sep 23. PMID: 39434757; PMCID: PMC11491222.

18. Uy TG, Alcala GR, Bocobo J, et al. Case report: Alveolar rhabdomyosarcoma of the prostate presenting as lower urinary tract symptoms in a Young Adult. Authorea. May 03, 2023. doi: 10.22541/au.168310018.85956384/v1