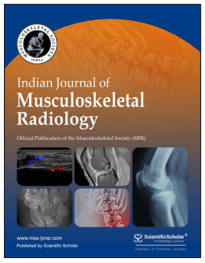
ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.mss-ijmsr.com

Indian Journal of Musculoskeletal Radiology



Case Report

Rare site and age of presentation: Prostatic sarcoma of the Ewing family in an adolescent

Khalid Mehmood[1], Asad Shehzad[2]

Departments of [1]Radiology and [2]Transplat and Urology, Sindh Institute of Urology and Transplantation, Karachi, Pakistan.

***Corresponding author:**
Khalid Mehmood,
Department of Radiology
Sindh Institute of Urology
and Transplantation, Karachi,
Pakistan.

nazilakhalid@gmail.com

Received: 08 July 2021
Accepted: 09 October 2022
EPub Ahead of Print: 19 November 2022
Published: 21 December 2022

DOI
10.25259/IJMSR_40_2021

Quick Response Code:

ABSTRACT

Ewing sarcoma/primitive neuroectodermal tumor (PNET) is the second most primary malignant bone tumor in children and adolescents between 10 and 25 years. The extraosseous sarcomas are rare and often cause diagnostic confusion for clinicians. The cell of origin is unclear, but exhibits rapid growth and recurrence, leading to a poor survival rate of 10% only. This imposes an increased risk of chronic health conditions, financial burden, and impaired quality of life. We report the uncommon presentation in age and site of the tumor in the peri-prostatic tissue of a 14-year-old boy and discuss our treatment approach. An exploratory observational case study was conducted. The ultrasound of an enlarged pelvis mass showed a large heterogeneously hyperechoic lesion with central necrosis and increased vascularity in the mass. The magnetic resonance imaging scan and histopathology of the transrectal ultrasound-guided biopsy specimen confirmed the prostate tumor. The transrectal biopsy proved to be a feasible and effective diagnostic tool for such an unclear pelvic mass. The extraskeletal manifestation of Ewing's sarcoma imposes diagnostic confusion, but the case fulfilled the extraosseous Ewing sarcoma's diagnostic characteristic. The Ewing's sarcoma/PNET of the prostate being rare and posing a poor prognosis needs vigilant differential diagnosis and a multidisciplinary treatment approach for optimal and cost-effective treatments.

Keywords: primitive neuroectodermal tumor, adolescents, transrectal biopsy, extraskeletal ewing's tumor, chemotherapy

INTRODUCTION

Ewing sarcoma/primitive neuroectodermal tumors (PNETs) are the second-most primary malignant bone tumor in children and adolescents, but the extraosseous sarcomas rarely manifest in the prostate.[1] The worldwide reported number of prostate Ewing's sarcoma is <10. These have a very poor survival rate of less than a year due to rapid growth and recurrence with a survival rate of 10% only. It imposes an increased risk of chronic health conditions which not only adds to the financial burden but also impairs the functioning of life.[2] The case discussed was a minor with prostate sarcoma.

CASE REPORT

A 14-year-old patient, an Afghanistan immigrant, was referred from another city to the emergency department with 3 months history of right flank pain, burning micturition, and poor stream. The patient's past medical history was unremarkable.

On initial assessment and suspicion of a pelvic mass, the ultrasound pelvis revealed a large heterogeneously hyperechoic lesion with central necrosis and increased blood flow on Doppler ultrasound.

The MRI pelvis with and without contrast on Axial T1WI [Figure 1], T2WI, SAGITAL and T2WI coronal, and diffusion-weighted image (DWI) axial images showed large abnormal signal intensity mass, measuring 7.3 × 6.4 × 6.7 cm, isointense to the muscles on T1WI and T2WI with significant post-contrast enhancement and central area of necrosis seen. The mass was encasing the right-sided iliac vessels and ureter. Posteriorly, it was infiltrating the seminal vesicles and prostate gland [Figure 2]. The fat planes were also focally indistinct with the rectum. The DWI [Figure 3] showed high signal intensity in the large prostatic tumor due to restricted diffusion, while the apparent diffusion coefficient (ADC) map derived from DWI images showed a low ADC value in the prostate tumor.

Transrectal ultrasound-guided biopsy under general anesthesia was done to establish the histological evaluation and complete tumor staging of the mass. It showed characteristic cores of neoplastic lesions exhibiting sheets of distinguished small round blue cells with scanty cytoplasm and monomorphic hypochromatic nuclei. The panel of IHC markers showed leukocyte common antigen, chromogranin, and myogenin negative, whereas CD99 showed diffuse membranous positivity and Fli-1 diffuse nuclear positivity [Figure 4]. Both morphological and IHC features were characteristic of Ewing's sarcoma/PNET.

The contrast computerized tomography (CT scan) of the chest, abdomen, and pelvis showed heterogeneous enhancing locally infiltrating lesions in the pelvis [Figure 5]. The perilesional, inguinal, and para-aortic lymphadenopathy was suggestive of PNET, while the CT chest appeared normal. The bone marrow biopsy of the iliac crest and bone scintigraphy was negative for metastatic involvement of the skeleton. The prostate-specific antigen was 0.10 ng/ml. Initially, blood urea and creatinine were normally raised and fluctuated from 6.7 to 1.2 mg/dl, but bilateral percutaneous nephrostomy intervention addressed the concern effectively.

The treatment strategy for Ewing's family of carcinoma including PNET is multimodal. This includes chemotherapy followed by surgery and radiotherapy. Chemotherapy often is a combination therapy.

DISCUSSION

In 1921, James Ewing described the fourth most frequent primary malignant tumor of bone accounting for approximately 3% of all pediatric cancers.[1,3] Ewing sarcoma is most frequent in the first three decades of life.[3] The extraskeletal Ewing sarcoma is rare having a prevalence of 15%. Moreover, Ewing sarcoma of bone is 20%.[3] It usually manifests in young patients.[2,4]

The various reported sites of extraskeletal Ewing sarcoma include the paravertebral region (32%), lower extremities (26%), chest wall (18%), retroperitoneum (11%), pelvis and hip (11%), and upper extremities (3%).[2,4] The genitourinary system and other visceral organs are exceptionally rare sites of occurrence. Other rare reported sites are the urinary bladder, kidneys, adrenal gland, and penis.[5,6] Until now, the literature review revealed that the worldwide reported number of prostate Ewing's sarcoma is < 10 including our case. Almost all the patients were young adults (median age 27, range: 20–31). This reflects it's very rare, exhibits poor survival, and occurrence.[4-6]

The diagnostic confusion occurred regarding the extraskeletal manifestation of Ewing's sarcoma, but the case fulfilled the diagnostic criteria of extraosseous Ewing's sarcoma by having the following characteristics:

(a) MRI showed no osseous involvement, (b) bone scintigraphy revealed no increased uptake of bone or periosteum adjacent to the tumor in static images, (c) light microscopy manifested lesions consisting of small round blue cells with no differentiation features, and (d) exhibited cytoplasmic glycogen.[7]

The poor prognosis of pelvic disease owes to large masses imposing difficulty in full resection.[4,8] Lung metastasis is the most common (80%). The MRI proved to be vital to

S. No.	Title	Site of sarcoma	Age range
Case 1–6	Primitive neuroectodermal tumor/Ewing's sarcoma in adult uro-oncology: A case series from a developing country[1]	Kidney, prostate and adrenal gland.	20–34 years
Case 7	Ewing's sarcoma/primitive neuroectodermal tumor of the prostate: A case report and literature review[2]	Prostate	29 year
Case 8	Primitive neuroectodermal tumor/Ewing's sarcoma of the prostate in a young Indian adult – an extremely rare case report[3]	Prostate	40 year

1. Mohsin R, Hashmi A, Mubarak M, Sultan G, Shehzad A, Qayum A, *et al*. Primitive neuroectodermal tumor/Ewing's sarcoma in adult uro. Can Urol Assoc J 2013;7(5-6):e458-9. https://dx.doi.org/10.5489/cuaj.1393
2. Published online June 12, 2013.Oncology: A case series from a developing country. Urol Ann 2011;3:103-7.
3. Rastog N, Soni N, Mandawat A, Singh P, Tanwar R. Ewing's sarcoma/primitive neuroectodermal tumor of uterine cervix: A rare case report. Asian Pac J Health Sci 2016;3(1):165-7.

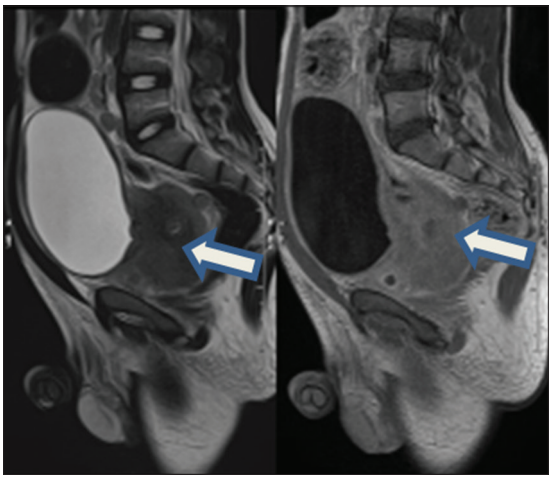


Figure 1: A sagittal T1WI and T2W2 MRI image of the prostate: The prostate is almost entirely replaced by the tumor mass. The mass is indenting the bladder base. The central necrotic area is noted (white arrow).

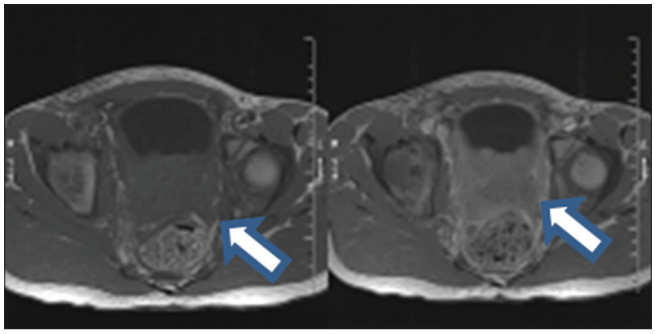


Figure 2: A pre- and post-contrast axial T1WI MRI pelvis image shows enhanced prostate mass with the central area of necrosis (white arrow).

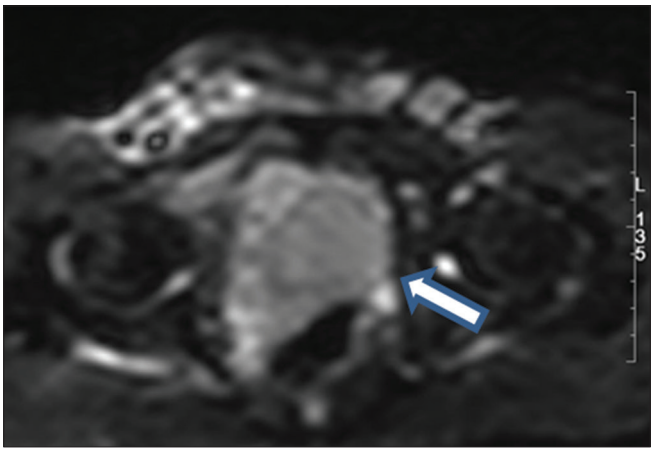


Figure 3: Diffusion-weighted image shows high signal intensity in large prostatic mass due to restricted diffusion (white arrow).

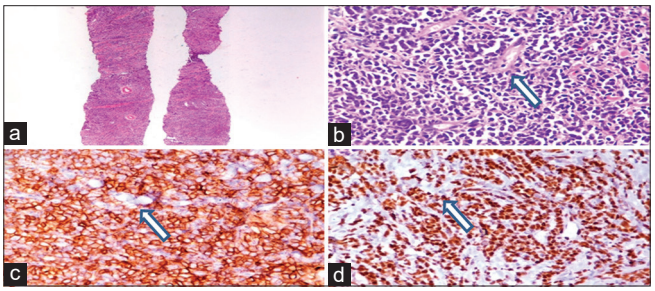


Figure 4: Histopathology of the lesion. (a) Low-power view showing two cores of lesion composed entirely of tumor tissue (HE, ×50). (b). High-power view showing small round blue cell tumor (blue arrow) (HE, ×400). (c) CD 99 immunostaining showing diffuse membranous positivity in tumor cells (blue arrow) (CD, ×400). (d). FLI 1 immunostaining showing diffuse nuclear positivity in tumor cells (blue arrow) (FLI 1, ×400).

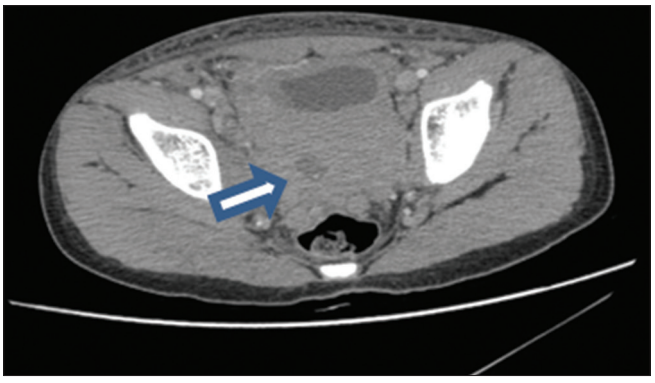


Figure 5: An axial section of CT scan pelvis on portovenous phase shows mild enhanced prostatic mass. A single area of necrosis is noted in the mass (white arrow).

evaluate response to neoadjuvant therapy (chemotherapy, radiotherapy, or both) and to detect local recurrence or metastatic disease.[9] Almost 1–2% of survivors are at risk of developing a secondary malignancy along with 20% of second bone sarcoma.[10]

Multidisciplinary management is the mainstay of treatment initiated by a biopsy to confirm the diagnosis, followed by neoadjuvant chemotherapy (before local control) and adjuvant (after local control) therapy. The size of the primary tumor shall be reduced by surgical resection, radiation therapy (RT), or both. This provides not only local control but also evaluates the degree of tumor necrosis and aids in preventing the recurrence emerging from small islands of chemotherapy-resistant cells.[10] The combination therapeutic approach with aggressive therapies such as surgery and chemotherapy (with or without RT) shall improve 5-year survival.

The case under discussion is about the prostate. Once multi-agent chemotherapy is completed and tumor size reduced by surgical resection, the patient would need prostatectomy which will be followed by radiotherapy to improve survival duration.

The strength of this case report is the availability of the data from the management team. The patient is under treatment for the final cycle of chemotherapy. Following chemotherapy, the size of the tumor regression shall be assessed and monitored for radical prostatectomy. Being an inhabitant of a distant city the future compliance and adherence to treatment are uncertain. In developing countries, the impact of socioeconomic barriers and poor health services is detrimental factors and may impede future compliance.

CONCLUSION

Sarcomas are a rare, but clinically relevant differential diagnosis in a young adult presenting with a large heterogeneous mass in the trunk, extremities, or soft tissues with internal hemorrhage and cystic necrosis, along with the lack of calcification and nodal metastases, which may help to establish the diagnosis and manage accordingly.

The rare occurrence of Ewing's sarcoma of prostate and poor prognosis garners enough attention to the differential diagnosis, histological confirmation, and definitive classification to establish multimodal, interdisciplinary, and collaborative treatment approaches for optimal and cost-effective therapeutic measures.

Acknowledgment

The authors are very grateful to the histopathology department for the detailed reporting of the biopsy specimen.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Dickman P. Ewing's sarcoma/primitive neuroectodermal tumor. Pathol Case Rev 2000;5:60-70.
2. Ross K, Smyth N, Murawski C, Kennedy J. The biology of Ewing sarcoma. ISRN Oncol 2013;2013:759725.
3. Angervall L, Enzinger FM. Extraskeletal neoplasm resembling Ewing's sarcoma. Cancer 1975;36:240-51.
4. Javery O, Krajewski K, O'Regan K, Kis B, Giardino A, Jagannathan J, *et al*. A to Z of extraskeletal Ewing sarcoma family of tumors in adults: Imaging features of primary disease, metastatic patterns, and treatment responses. AJR Am J Roentgenol 2011;197:W101522.
5. Mohsin R, Hashmi A, Mubarak M, Sultan G, Shehzad A, Qayum A, *et al*. Primitive neuroectodermal tumor/Ewing's sarcoma in adult uro-oncology: A case series from a developing country. Urol Ann 2011;3:103-7.
6. Okada Y, Kamata S, Akashi T, Kurata M, Nakamura T, Kihara K. Primitive neuroectodermal tumor/Ewing's sarcoma of the urinary bladder: A case report and its molecular diagnosis. Int J Clin Oncol 2010;16:435-8.
7. StuartHarris R, Wills EJ, Philips J, Langlands AO, Fox RM, Tattersall MH, *et al*. Extraskeletal Ewing's sarcoma: A clinical, morphological and ultrastructural analysis of five cases with a review of the literature. Eur J Cancer Clin Oncol 1986;22:393400.
8. Song HC, Sun N, Zhang WP, Huang CR. Primary Ewing's sarcoma/primitive neuroectodermal tumor of the urogenital tract in children. Chin Med J 2012;125:932-6.
9. Fuchs B, Valenzuela RG, Petersen IA, Arndt CA, Sim FH. Ewing's sarcoma and the development of secondary malignancies. Clin Orthop Relat Res 2003;415:82-9.
10. Ma Z, Brimo F, Zeizafoun N. Primary Ewing's sarcoma/primitive neuroectodermal tumor (ES/PNET) of the penis: A report of an extraordinarily unusual site and a literature review of extraskeletal Ewing's sarcoma. Int J Surg Pathol 2013;21:63-7.

How to cite this article: Mehmood K, Shehzad A. Rare site and age of presentation: Prostatic sarcoma of the Ewing family in an adolescent. Indian J Musculoskelet Radiol 2022;4:115-8.