Case Report

# Diagnostic challenges in reporting a case of extraskeletal Ewing's sarcoma of the prostate in a young Indian adult

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## ABSTRACT

Extraosseous prostatic Ewing's sarcoma (ES) in young adults is extremely rare. We present a case of locally advanced prostate ES/primitive neuroectodermal tumor (PNET) in a 28-year-old male. He presented with difficulty in micturition and defecation for 6 months. On magnetic resonance imaging, a mass lesion was found involving the central and peripheral zones of the prostate, and it was provisionally diagnosed as a neoplastic lesion with a differential diagnosis of lymphoma or a neuroendocrine tumor. A transrectal ultrasound-guided fine needle aspiration cytology and needle biopsy, followed by histopathological examination, and immunohistochemistry confirmed the diagnosis of Extraosseous ES. The patient underwent a cystoprostatectomy followed by multi-agent chemotherapy. Presently, the patient is alive and is being followed up regularly. Nevertheless, this tumor has a very poor prognosis, with the longest reported survival of 24 months.

**Key words:** Ewing's sarcoma, NK2 homeobox 2, primitive neuroectodermal tumor, Prostate.

Ewing's sarcoma (ES) and primitive neuroectodermal tumor (PNET) are primarily small round cell tumors of soft tissue and bones [1]. ES is reported in various organs, and the prostate gland is infrequently involved by metastatic tumors [2]. Due to the absence of unique clinical features, histopathological examination is essential in making an accurate diagnosis. Moreover, overlapping morphological features with small cell carcinoma, lymphoma, alveolar rhabdomyosarcoma, and melanoma made the diagnosis challenging. Immunohistochemistry aided in further corroboration.

We would like to document the first case report of Estraskeletal ES manifesting in the prostate from North-East India.

## CASE REPORT

A 28-year-old male presented to the urology Outpatient Department of Gauhati Medical College and Hospital with difficulty in micturition and defecation for the past 6 months.

The patient was cachectic, and on digital rectal examination, an irregular, hard mass was felt in the prostate gland; no obvious lymph node swelling was noted.

Biochemical investigations were advised and revealed a normal white blood cell count. C-reactive protein, prostate-specific antigen (0.5 ng/mL) levels, and urine routine examination were within normal limits. On magnetic resonance imaging scan (Fig. 1), we found an extensive intraprostatic tumor (measuring 13.5 cm × 9.8 cm × 90 cm) involving the central and peripheral zones with sparing of the transitional and anterior fibromuscular zones. It was provisionally diagnosed as a neoplastic lesion with a differential diagnosis of lymphoma or a neuroendocrine tumor. A transrectal ultrasound-guided fine needle aspiration cytology (FNAC) and needle biopsy were done.

Cytomorphology showed atypical small round cells arranged predominantly in clusters, having fine chromatin and scant cytoplasm in a hemorrhagic background (Fig. 2). It was reported as a small round cell tumor. Histomorphology sections from the ultrasound-guided biopsy showed solid sheets of uniform small round cells having fine chromatin and scant eosinophilic cytoplasm. Mitosis was also noted. No epithelioid cells or spindle cells were seen.

Background stroma was not sclerotic or myxoid (Fig. 3a and b). Hence, the differential diagnosis of

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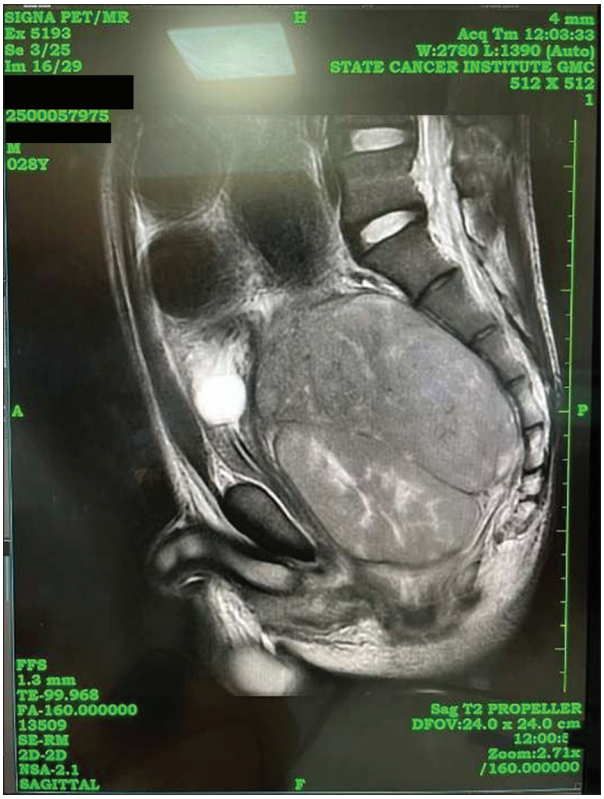


**Figure 1: Magnetic resonance imaging showing an extensive intraprostatic tumor involving the central and peripheral zones**

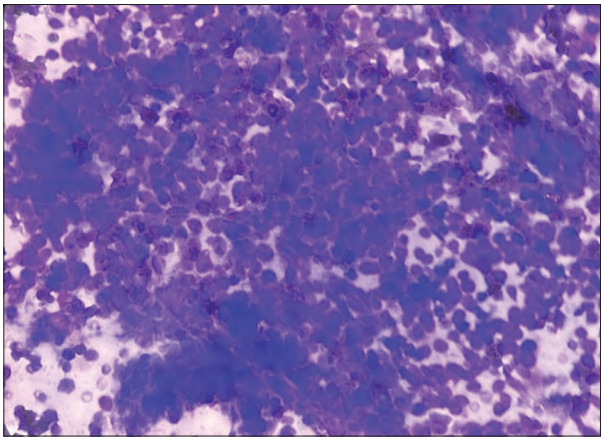

**Figure 2: MGG-stained smears show sheets of small, round cells having hyperchromatic nuclei and scant cytoplasm**

neuroendocrine tumor, small round cell sarcomas, lymphoma, alveolar rhabdomyosarcoma, and melanoma was made. A provisional panel of IHC markers, including PanCK, CD45, S100, Myogenin, Synaptophysin, and Chromogranin was negative. But CD99 showed diffuse positivity (Fig. 3c). In the second panel of IHC, NKX2.2 showed strong diffuse nuclear positivity (Fig. 3d). CD45 negativity ruled out lymphoma. Capicua transcriptional repressor (CIC) rearranged sarcomas have diffuse sheets of round cells with uniform nuclear morphology and light eosinophilic cytoplasm. Some cases display predominantly spindled, epithelioid, or plasmacytoid morphology.

On H&E, these features were not appreciated in our biopsy [3]. Further, NKX2.2 positivity in the present case ruled out the possibility of CIC rearranged sarcomas.

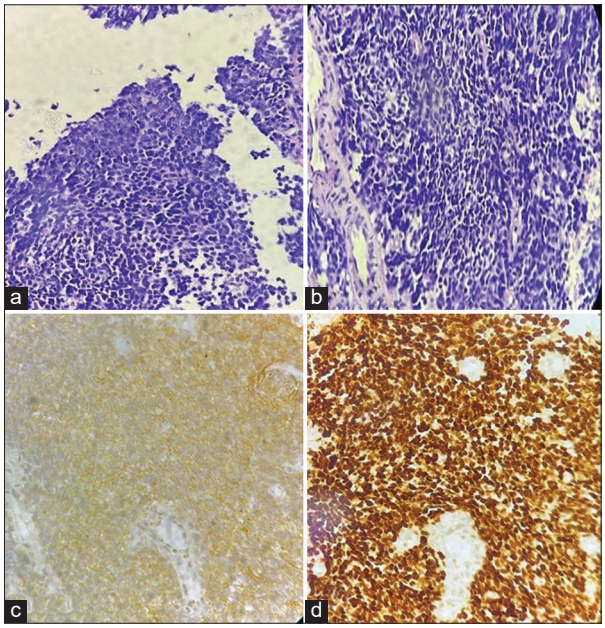


**Figure 3: (a and b) H&E-stained section shows sheets of uniform small round cells having fine chromatin and scant cytoplasm; (c) Tumor cells show diffuse membranous positivity for CD99 (×40); (d) Tumor cells show strong nuclear positivity for NKX2.2 (×40)**

Tumors with EWSR1-non-ETS fusion have tumor cells arranged in cords, nests, trabeculae, and pseudoacinar pattern. Cells have hyperchromatic or vesicular chromatin and prominent nucleoli with eosinophilic to clear cytoplasm. Hemangiopericytic vascular network is present. Cells may have cartilaginous or osteoid-like differentiation. Stroma is fibrohyaline or myxohyaline. These features were not seen in our prostate biopsy specimen [3]. On IHC, negativity for myogenin and S100 ruled out round cell sarcoma with EWSR1-non-ETS fusion.

Sarcoma with BCOR genetic alteration has solid sheets and fascicles of small round blue cells along with short plump spindled cells separated by a delicate capillary network, in a myxoid stroma [3]. The present case did not show spindle cells or myxoid stroma on H&E. On immunohistochemistry, CD99 was diffusely positive in more than 90% of the tumor cells, and NKX2.2 was strongly diffusely positive, which ruled out BCOR rearranged sarcomas. The final diagnosis of Extraskeletal ES was established. Molecular studies for EWSR1-FLI1 and EWSR1-ERG were advised.

## DISCUSSION

ES is more commonly encountered in bone and soft tissue. Extraosseous ES, formerly known as Peripheral PNET, shows consistent EWSR1-ETS family gene fusion, most commonly EWSR1-FLI1, as a consequence of reciprocal t(11;12)(q24;q12) translocation [3]. In addition, a rare subtype of round cell tumors harbors EWSR1-non-ETS fusion, and these lesions are known as Ewing-like sarcomas [3]. For extraosseous primary, the most common sites are: trunk (32%), extremity (26%), head and neck (18%), retroperitoneum (16%), and other

sites (9%) [4]. As for the field of urology, individual cases of extraosseous ES have also been reported in the Urinary bladder, kidney, adrenal glands, and even penis [5-8]. To the best of our knowledge, fewer than 50 cases of prostatic ES are known worldwide, being first described in 2003 by Coleacchia *et al*. [4,9,10] and the latest by Chakravarthi *et al*. in 2025 [11].

In the study done by Chakravarthi *et al*. [11], Funahashi *et al*. [12], Wu *et al*. [2], and Esch *et al*. [4], the age group of presentation was 20–35 years, which was similar to our study. On the other hand, only one patient presented at the age of 64years in the study done by Sharma *et al*. [10]. The majority of the cases presented with lower urinary tract symptoms, except for the case reported by Charkravarthi *et al*., which presented with progressive lower back pain and abdominal discomfort along with systemic metastases resulting in a guarded prognosis [11]. Similar to the present case, most of the patients had early diagnosed localized disease with regular follow-up and disease-free survival.

## CONCLUSION

Extraosseous ES of the prostate is extremely rare. Its prognosis is very poor. Hence, extra attention should be paid to the differential diagnosis of this entity. Histological categorization is paramount for definitive treatment. Presently, the treatment of choice is surgery followed by chemotherapy. New targeted therapy needs to be developed for clinical application in the future. Early diagnosis is curtail, given the aggressive nature of the tumor for systemic metastasis.

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