Atypical Presentation of Ewing Sarcoma in the Spermatic Cord of a Young Adult

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ABSTRACT

This case report describes a rare case of an 18-year male from rural Balochistan, Pakistan. The presentation was unusual with weight loss and slow-growing, mildly tender, mobile swelling, in the left inguinal region suggesting neoplastic aetiology, misdiagnosed as an enlarged inguinal lymph node. Tumour markers were negative and there was no evidence of nodal involvement. An excision biopsy of the inguinal mass showed a reactive lymph node. The patient underwent uncomplicated radical orchidectomy after a month, and on histopathology, it turned out to be Ewing sarcoma. Later, contrast-enhanced CT (CECT) chest, abdomen, and pelvis (CAP) confirmed widespread pulmonary metastasis while the bone scan was normal. Adjuvant chemotherapy was introduced as per the National Comprehensive Cancer Network (NCCN) guidelines. Despite all efforts, it recurred as an aggressive metastatic disease with a recurrence of mass larger than the previous one at the surgical site with involvement of local and distant organs, taking its toll on health and leading to the demise of the patient within one year.

Key Words: *Ewing sarcoma, Spermatic cord, Paratesticular lesion*.

How to cite this article: Ali A, Waheed A. Atypical Presentation of Ewing Sarcoma in the Spermatic Cord of a Young Adult. *JCPSP Case Rep* 2025; **3**:232-234.

INTRODUCTION

Ewing sarcoma (ES) is a rare, highly malignant neoplasm, accounting for approximately 1% of all childhood cancers, and primarily manifests in the soft tissues, long bones, pelvis, and chest walls of paediatric and adolescent patients. Only 15-20% of cases are extraskeletal in origin.[1] Its occurrence in the spermatic cord is exceedingly rare, making diagnosis even more challenging, and delaying the initiation of appropriate treatment, worsening the prognosis. The typical presentation of ES involves pain and swelling. Histologically, ES features small round blue cells, positive for CD99 and FLI1 on immunohistochemical staining. Cytogenetically, there is a translocation between chromosomes 11 and 22, t(11;22) (q24;q12), forming the EWS-FLI1 fusion gene. The management generally involves a multimodal approach; surgical resection, chemotherapy, and sometimes radiotherapy.[2,3]

Despite advances in treatment, the prognosis for ES remains guarded, particularly with metastatic disease.[4] Regular follow-up and imaging are essential for monitoring disease recurrence and progression.

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Received: August 16, 2024; Revised: November 02, 2024; Accepted: November 09, 2024
DOI: https://doi.org/10.29271/jcpspcr.2025.232

CASE REPORT

An 18-year male patient from rural Balochistan, Pakistan, presented with a 2-months history of a slowly growing, painless, and left inguinal mass. He had no significant medical history and denied any trauma; however, he had undocumented weight loss with a preserved appetite.

Physical examination revealed a painless, non-tender, mobile mass of the size of a walnut (approximately 4×5 cm) in the left inguinal region a few centimetres from the scrotal margin. On ultrasound of the abdomen and the left inguinoscrotal region, a mobile mass was seen in the para-testicular location measuring 47×54 mm with both solid and cystic components showing neoplastic aetiology with suspicion of an enlarged lymph node (Figure 1).

CT scan of the abdomen and pelvis showed a soft tissue density lesion with internal necrosis confluent with a lymph node in the left inguinal region. Further evaluation with an MRI pelvis suggested no evidence of visceral metastasis.

As radiological investigations failed to find a primary source, the patient was referred to a tertiary care oncology centre. An excisional biopsy of the lymph node showed a lymph node with reactive changes with immunohistochemical (IHC) staining showing CD20 positivity in B-cells, CD3 in T-cells, while CD30 was negative. In the following days, the condition of the patient did not improve and he became more cachectic and anorexic. He was counselled and underwent an uncomplicated radical orchidectomy for the suspicion of a scrotal primary. The histopathology of the specimen showed a round blue cell tumour consistent with the ES of the spermatic cord. The tumour was 2 mm away

from the nearest resection margin (R0). Unremarkable testicular parenchyma was seen in the background. The IHC staining showed OCT3/4, negative; SALL4, focal positive; MyoD1, negative; desmin, negative; NKX2.2, positive; CD99, positive; LCA, negative; and TdT, negative. Periodic acid-pschiff (PAS) stain was positive for glycogen.

Keeping in view the aggressive natural history of ES, the patient was started on adjuvant chemotherapy as per the NCCN guidelines i.e. initially with VAC/IE (vincristine, doxorubicin [Adriamycin], and cyclophosphamide alternating with ifosfamide and etoposide) and then with VAC (vincristine, doxorubicin, and cyclophosphamide).[3]

Follow-up bone scan revealed no osteoblastic metastasis; however, contrast-enhanced CT (CECT) chest, abdomen, and pelvis (CAP) revealed multiple lesions in bilateral lung parenchyma and mass in the left hemipelvis along with multiple inguinal and para-aortic node involvement suggesting widespread metastatic disease despite the removal of primary source and chemotherapy. The patient was kept on palliative treatment afterwards till the last of his days.

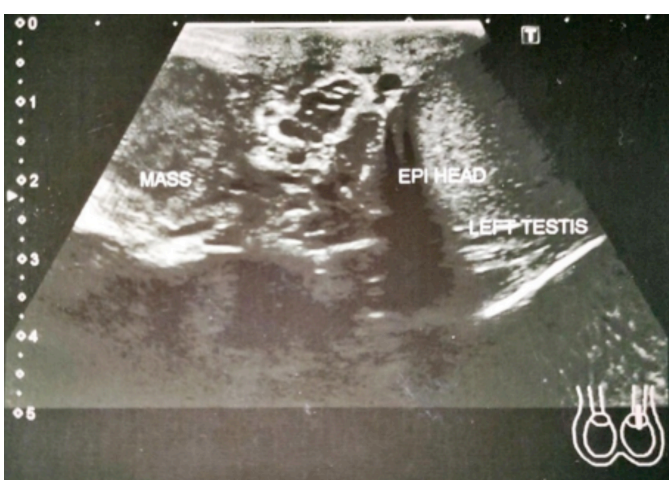

Figure 1: Ultrasonography of left inguinoscrotal region.

DISCUSSION

ES is a rare, aggressive, and malignant neuroectodermal tumour most commonly affecting the long bones, pelvis, and chest wall, with rare extra-skeletal occurrences. It requires prompt diagnosis and management. ES of the spermatic cord is exceedingly rare, leading to potential misdiagnosis as more common scrotal pathologies, thus delaying treatment.[2] One such case has been documented by Yashaswi *et al*., where a right inguinal mass in a 49-year man, who was misdiagnosed as an extra-abdominal gastrointestinal stromal tumour (GIST), turned out to be an extraskeletal ES.[5]

This case involved a painless, progressively enlarging inguinal mass without significant systemic symptoms except for undocumented weight loss, typical of ES presentations. Imaging studies such as ultrasonography and CT scans are essential for initial assessment. Magnetic resonance (MR) and fluorodeoxyglucose-positron emission tomography (FDG-PET) imaging are used for initial diagnosis and detection of metastasis,[6] but definitive diagnosis requires histopathological and molecular analysis, particularly identifying the EWSR1-FLI1 fusion gene, characteristic of ES regardless of the location.[3] In contrast to the ES of bones that show a characteristic periosteal reaction and aggressive osteolytic activity in surrounding tissue, ES of the spermatic cord appears as a localised mass complicating the diagnosis.

For ES of the spermatic cord, radical inguinal orchiectomy is the surgical treatment of choice, followed by systemic chemotherapy. Chemotherapy targets micrometastatic disease, which is commonly present at diagnosis but not detectable by imaging. Definitive treatments for localised disease include chemotherapy and surgery. Radiation therapy is effective in unresectable diseases.[6] Despite radical orchiectomy and adjuvant chemotherapy, this patient developed widespread metastasis, a poor prognostic indicator. The prognosis depends on tumour location, size, metastatic spread, and treatment response. Metastatic disease at diagnosis significantly worsens the prognosis, with a five-year survival rate of 20-30% compared to 70-80% for localised disease.[4]

This case highlights the challenges in diagnosing and managing rare ES presentations. The unusual spermatic cord location underscores the need for a thorough differential diagnosis. Rapid disease progression, despite aggressive treatment, highlights ES's aggressive nature and the necessity for early, prompt multimodal treatment. Continuous research and clinical trials are vital for improving outcomes for ES patients.

PATIENT'S CONSENT:
Informed consent is obtained from patients to publish the data concerning this case.

COMPETING INTEREST:
The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:
AA: Conception and design of the study, acquisition of data, analysis, and interpretation of data.
AW: Drafting of the article and revising the manuscript critically.
Both authors approved the final version of the manuscript to be published.

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