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EXTRASKELETAL EWING SARCOMA OF SCAPULAR SOFT TISSUE MIMICKING LIPOSARCOMA: A DIAGNOSTIC CHALLENGE IN A YOUNG FEMALE

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ABSTRACT

Antimicrobial resistance (AMR) has become one of the most serious global public health threats, driven by the rapid
Ewing sarcoma is the second most common frequent malignant bone tumor in children and adolescents, with neuroectodermal characteristics. Extra skeletal Ewing sarcoma (EES) is a rare, aggressive small round-cell tumor that develops in soft tissues without primary bone involvement, with a slight male predominance. Due to its rarity and overlapping features with other soft tissue tumors, EES poses a significant diagnostic challenge, especially at atypical anatomical sites. We report a case of 21-year-old female who came with complaints of swelling over the right scapular region. Radiological examination suggestive of liposarcoma, while histopathological examination showed features of malignant small round cell tumor, without underlying scapular bone involvement. Immunohistochemistry showed CD99 positivity and Ki- 67 labeling index of approximately 25%. Molecular analysis couldn't be done due to resource limitation, However the diagnosis was established by integrating histopathological morphology, immunohistochemistry (CD99-positive), and clinicoradiological correlation.

EES arises from the long bones of the extremities more commonly, however this case highlights a rare occurrence in the soft tissue overlying a flat bone (scapula) without bony involvement. This unsual anatomical location, particularly in a young female, and the imaging findings mimicked liposarcoma, creating a significant diagnostic challenge. Although molecular confirmation is considered the diagnostic gold standard, it was not feasible in our institute due to resource limitations. Nevertheless, the diagnosis of EES was reliably established through a comprehensive and evidence-based approach integrating characteristic histomorphology, strong membranous CD99 immunopositivity, and clinic radiological correlation. This multimodal diagnostic strategy is well-recognized as an acceptable alternative in resource-limited settings. This case highlights the importance of considering Ewing sarcoma in the differential diagnosis of soft tissue masses at atypical sites for accurate diagnosis and early management.

KEYWORDS: Extraskeletal Ewing sarcoma, Scapular region, Small round cell tumor, CD99, Immunohistochemistry.

INTRODUCTION

Ewing sarcoma is the second most common frequent malignant bone tumor in children and adolescents, with a slight male predominance following osteosarcoma. EES is a malignant neoplasm arising from undifferentiated small round cells in soft tissues rather than bone. It belongs to the Ewing sarcoma family of tumors (ESFT), which also includes osseous Ewing sarcoma and primitive neuroectodermal tumor (PNET) [1]. Its incidence is lower than that of osseous Ewing sarcoma.

It is characterized by specific chromosomal translocations, most often t (11;22) (q24; q12), which results in EWSR1–FLI1 fusion gene [2]. This fusion protein plays a critical role in tumorigenesis by promoting abnormal cellular proliferation and survival. EES commonly involves the chest wall, paravertebral region, lower extremities, head and neck, and pelvis, with clinical presentation varying according to tumor location [3].

Despite its rarity, EES poses significant diagnostic and therapeutic challenges. Accurate diagnosis requires a multimodal approach, including imaging studies, histopathological examination, immunohistochemistry, and molecular genetic analysis to identify characteristic translocations. Molecular confirmation is not absolutely required when morphology and immunohistochemistry are typical, provided alternative diagnoses are excluded. Management typically involves multimodal therapy, including wide local excision or radiotherapy, followed by systemic chemotherapy.

Case Report:

A 21-year-old woman who presented with swelling over the right scapular region for six months. The swelling was insidious onset and gradually increased in size. On clinical examination, a solitary swelling measuring 10×8 cm was noted.

Radiological evaluation suggested a soft tissue neoplasm, with a provisional diagnosis of liposarcoma, located in the intramuscular plane of the right posterior axillary fold extending into the scapular region.

Fig 1: A soft tissue mass in the right posterior axillary fold extending into the scapular region.

A trucut biopsy specimen was received in the histopathology laboratory. Microscopic examination revealed cores of a neoplasm composed of sheets of small round cells with scant cytoplasm and hyperchromatic nuclei. Intervening hyalinized blood vessels and increased mitotic activity was observed, while necrosis was absent. Based on the histomorphological features, a diagnosis of a malignant small round cell tumor was considered, suggestive of extra skeletal Ewing sarcoma. The patient subsequently underwent wide local excision with right scapulectomy. Frozen section analysis of all surgical margins was negative for tumor involvement. Routine Histopathological examination was done. On gross examination, a unifocal tumor measuring 12.5 x 8.5 x 5.3 cm was identified in the right shoulder region. Microscopic examination confirmed the diagnosis of extra-skeletal Ewing sarcoma. The FNCLCC grading system assigned the tumour a Grade 2 (score 5). There was no evidence of lymph vascular infiltration or tumour necrosis and all surgical margins were free of tumor, with the closest margin being the superior margin (<0.5 cm).

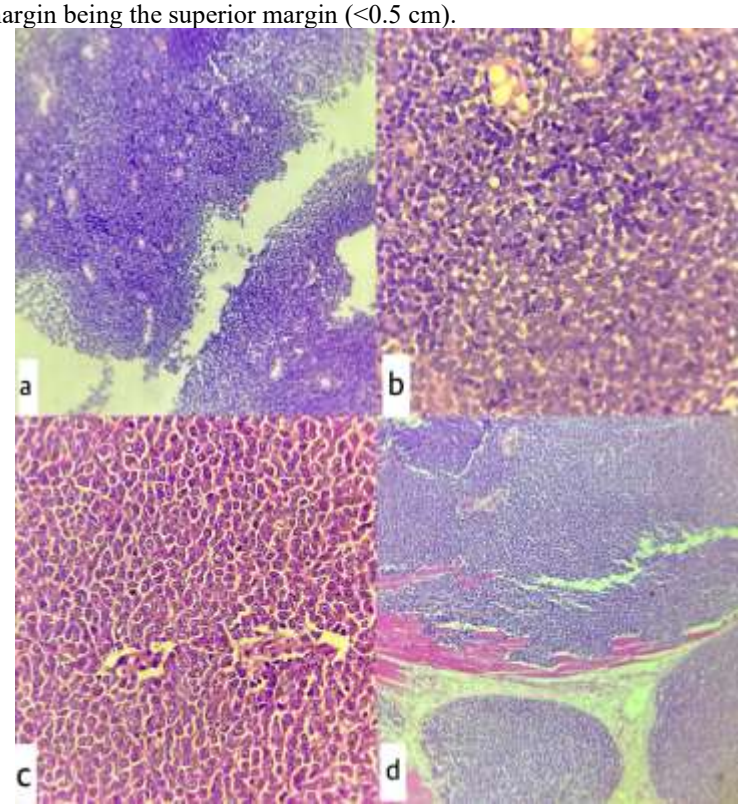

Fig 2: (a) Trucut biopsy, Swelling Right scapula, sheets of small round cells with hyperchromatic nucleus and scanty cytoplasm in a diffuse sheet pattern. (b, c. d) wide local excision with right scapulectomy., Features of Malignant small round cell tumor (H&E, 10x &40x)

Immunohistochemical analysis demonstrated strong membranous positivity for CD99 in approximately 90% of tumor cells. Ki-67 showed a labeling index of approximately 25%, indicating a high proliferative activity.

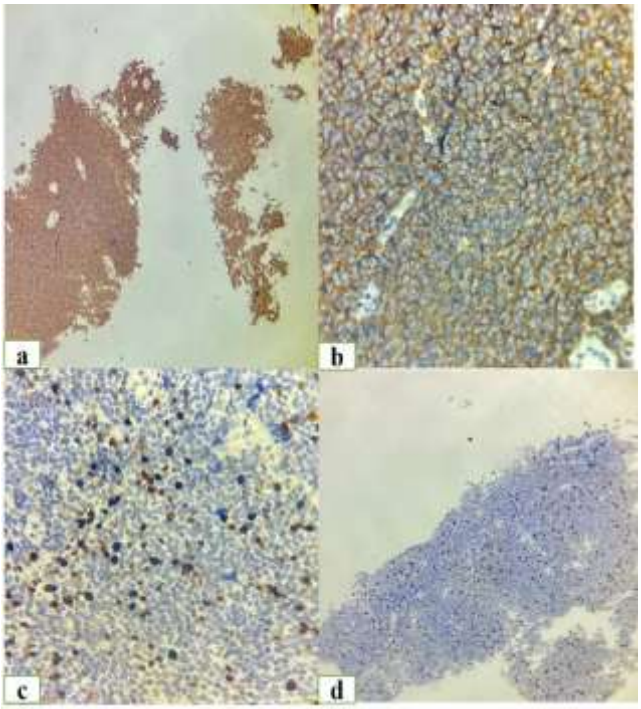

Fig 3: (a,b) Tumor cells showing strong membranous positivity for CD99, (c, d) Ki-67 immunostaining showing approximately 25% nuclear positivity indicating high proliferative index

Molecular confirmation with EWSR1 rearrangement is considered the gold standard, but its unavailable in our institute. Hence, the diagnosis was made in keeping with WHO-based criteria for Ewing sarcoma family tumors, based on characteristic histopathological morphology and strong membranous CD99 expression, exclusion of other small round - cell tumors and clinico-radiological findings.
Pathological staging according to AJCC 8th edition was pT3 pNx, based on tumor size (10–15cm) and absence of nodal evaluation. Importantly, there was no involvement of the underlying scapular bone, confirming the Extraskeletal Ewing sarcoma of the scapular soft tissue.

DISCUSSION

Extraskeletal Ewing Sarcoma (EES) is an uncommon, aggressive, malignant small round cell tumor that arises from soft tissues without primary bone involvement [4]. It is a member of the Ewing sarcoma family of tumors as it shares the histopathology, immunohistochemistry, and molecular features [5,6]. Ewing sarcoma typically arises from the diaphysis of long bones; however, extraskeletal involvement at atypical sites has been increasingly recognized. However, involvement of the scapular region, particularly confined to the overlying soft tissue without underlying bone involvement, is exceedingly rare, especially in a young female patient [7,8].
The present case underscores a significant diagnostic challenge, as it highlights an unusual anatomical presentation of EES due to its radiological mimicry of liposarcoma, which is consistent with previous reports indicating that EES often presents as a non-specific soft tissue mass and may mimic other sarcomas on imaging. Studies by Abboud A et al. [5] have reported that EES most frequently involves the paravertebral region, lower extremities and chest wall, with relatively fewer cases in the scapular region, particularly in a young female, emphasizing the rarity of this presentation.
Microscopically, EES is characterized by sheets of homogeneous, small, round cells with scanty cytoplasm and hyperchromatic nuclei. These features overlap with other small round cell tumors, necessitating IHC evaluation. In this case, strong membranous positivity for CD99 supports the diagnosis. CD99 expression has been reported in more than 90% of Ewing sarcoma cases in the literature, reinforcing the diagnostic utility. The Ki-67 labeling index of approximately 25% indicates a high proliferative activity of the tumor, consistent with the aggressive behavior of EES. The differential diagnosis of small round cell tumors in soft tissue includes lymphoma, rhabdomyosarcoma, synovial sarcoma, and desmoplastic small round cell tumor. These were excluded based on histomorphological features and immunohistochemical profile. Thus, the diagnosis of EES was strongly supported.
Molecular confirmation with detection of EWSR1 gene rearrangement remains the gold standard for diagnosis, although it was not performed in this case due to its unavailability in our setting. Studies by Denis R et al. [9] have demonstrated that a reliable diagnosis can be established using a characteristic histopathology and IHC finding sin resource -limited settings. Accordingly, this case fulfills the WHO-diagnostic framework of EES, including sheets of small round cells, strong CD 99 membranous positivity, elevated Ki-67 index, exclusion of other small round cell tumors and absence of underlying bone involvement.
The management of EES requires a multimodal approach. Complete surgical excision with negative margins is a key prognostic factor [8]. In the present case, wide local excision with right scapsulectomy achieved tumor-free margins, with no involvement of the underlying scapular bone, confirming its extra skeletal origin. The patient subsequently received adjuvant combination chemotherapy with vincristine, actinomycin D, and cyclophosphamide (VAC regimen) to address

potential micro metastatic disease [9]. At 6-month follow-up, the patient remained clinically stable with no evidence of local recurrence or distant metastasis on surveillance imaging. Radiotherapy may also be considered in selected cases. Prognosis in EES depends on several factors, including tumor size, site, presence of metastasis, and response to therapy [5,10]. Larger tumor size, as seen in this case (>10 cm; pT3), is generally associated with a less favorable prognosis, highlighting the importance of early diagnosis and comprehensive treatment.

CONCLUSION

Extraskeletal Ewing sarcoma involving the scapular soft tissue is an exceptionally rare entity, particularly in a female patient. Only a limited number of such cases have been reported in literature. The large tumor size (>10 cm) in this case is associated with a relatively poorer prognosis, underscoring the importance of early detection. It often presents with nonspecific symptoms such as localized pain, swelling, and restricted shoulder movement, which can be easily mistaken for more common orthopedic conditions. Its atypical presentation can pose significant diagnostic challenges, often mimicking other soft tissue sarcomas. This case underscores the critical role of histopathology and immunohistochemistry in establishing an accurate diagnosis. A multidisciplinary approach, including complete surgical excision and adjuvant chemotherapy, is essential for effective management and improved prognosis.

Limitation:

A key limitation of this study is the absence of molecular confirmation due to unavailability of advanced diagnostic facilities in our institute. However, the diagnosis was consistent with WHO-based criteria for Ewing sarcoma family tumours, based on characteristic histopathological features, strong membranous CD99 expression, exclusion of other small round-cell tumours, and clinicoradiological correlation.

Conflict of Interest Statement

The authors declare no conflicts of interest.

Source of Funding

This study received no external funding

Statement of Ethics

Informed Consent was obtained from the patient for publication of this case report and accompanying images

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