Original Research Paper

Volume - 14 | Issue - 01 | January - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Radio-Diagnosis

EXTRAOSSEOUS EWING SARCOMA OF CHEST WALL – AN UNUSUAL PRESENTATION

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ABSTRACT Extraosseous Ewing's sarcoma (EES) is a malignant mesenchymal tumor much rarer than its intraosseous counterpart. Few case reports of EES have been published in the literature. Only one in five cases of Ewing's sarcoma occurs as Extraskeletal. The use of magnetic resonance imaging helped diagnose, plan procedural interventions, assess neoadjuvant chemotherapy effectiveness, and detect local recurrences and metastatic spread of the tumor. Immunohistochemical markers further differentiated the diagnoses. EES should be considered when evaluating soft tissue lumps of neoplastic characteristics, in children or adolescents. Early detection of EES is essential.

KEYWORDS : Ewing sarcoma, Extraskeletal, Soft tissue Tumor, Imaging, Case report

INTRODUCTION

Extraskeletal Ewing's sarcoma (EES) is an uncommon, rapidly growing, round-cell malignancy of uncharacterized mesenchymal cell origin. Tefft et al first described EES in 1969 to be histologically similar to primary Ewing's sarcoma of bone (1). EES is a rare soft tissue neoplasm, that can develop in the soft tissues at any location. They range from indolent to highly invasive and metastatic. ES and PNET are regarded as two extremes of a morphologic spectrum of the same tumor entity based on similar clinical, immunohistochemical, and cytogenetic profiles (1). Based on a previous report, the incidence of EES is 0.4 per million individuals, which is lower than that of ES of the bone by 10-fold. Although uncommon, the occurrence of EES seems to have a bimodal distribution, where there is a peak in the occurrence rate among children (<5 years) and adults (>35 years). Unlike Ewing sarcoma of the bone, no evidence supports a link between the tumor and race or biological sex. The origin of these tumors is unclear, but they appear to be derived from cells migrating from the neural tube, with different ectodermal or neuronal differentiation abilities (2).

Magnetic resonance (MR) and fluorodeoxyglucose positron emission tomography (FDG-PET) imaging are used for initial diagnosis and detection of metastasis, respectively (3). The management of EES includes surgery and chemotherapy in resectable tumors. Under unresectable conditions, radiotherapy is usually considered. According to the National Comprehensive Cancer Network (NCCN), the optimum management of EES remains not clearly defined, although some studies have highlighted an added value of surgery among EES cases compared to Ewing sarcoma of the bone in terms of better survival rates. (4)

Case Description:

- A 9 years old female child came with chief complaints of pain in right axillary region for 8 months, swelling in right axilla for 3 months, restriction of movements of right upper limb for 3 months. No history of fever, loss of weight or loss of appetite. No previous history of similar complaints. No family history of similar complaints in the past. No history of tuberculosis.
- Vitals: Pulse rate: 65/ minute, BP: 100/60 mmHg, SpO2: 98%@ room air.
- Routine blood investigations were within normal range. On examination A well-defined, smooth, fluctuant swelling of size ~ 7 x 4 cm is noted in the right axillary region.
- Proceeded with ultrasound and MRI.
- Later USG guided FNAC was done and patient was posted for excision biopsy.

USG FINDINGS:

- A heterogeneously hypoechoic well-defined lesion of size ~ 7.5 x 4.4 x 4.2 cm with densely packed internal echoes in the subcutaneous plane of the right axillary region. There are multiple cystic components and multiple thin septations within. This lesion is seen to abut the adjacent muscle posteriorly (Figure 1).

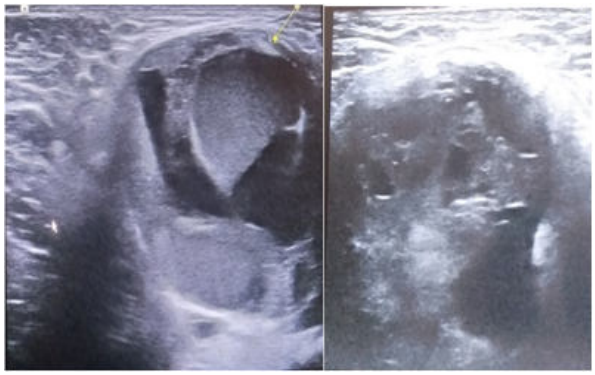

Figure 1: A heterogeneously hypoechoic well-defined lesion with densely packed internal echoes in subcutaneous plane of right axillary region.

Source: Pondicherry institute of medical sciences.

MRI Findings:

- Well-defined lobulated, T2/STIR hyperintense mass lesion of size ~ 8.5x4.1x4.4 cm with internal septations is noted in the right axilla extending to the right brachial plexus causing significant compression/ displacement of the neurovascular structures in the axilla (Figure 2).

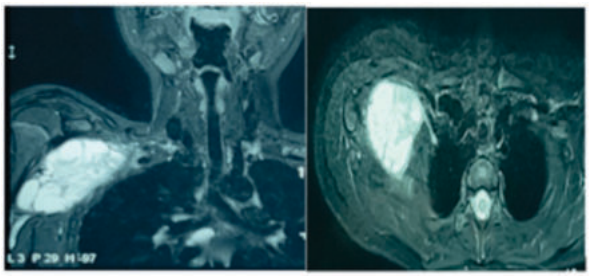

Figure 2: Well-defined lobulated, T2/STIR hyperintense mass lesion with internal septations is noted in the right axilla.

Source: Pondicherry institute of medical sciences.

Curvature of the spine is maintained, vertebral bodies, cervical intervertebral disc spaces, facet joints, ligamentum flavum, craniovertebral junction, and cord are normal.

Differential Diagnosis:

- Possibility of differentials of Lymphangioma / schwannoma with cystic degeneration was considered. Further, USG guided FNAC was advised and performed (Figure 3).

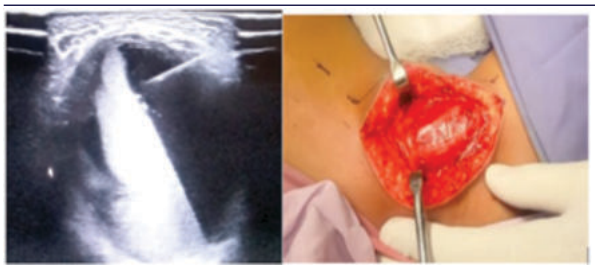

Figure 3: USG guided FNAC and excision biopsy performed.
Source: Pondicherry institute of medical sciences.

FNAC:

- FNAC showed clusters of Epithelial cells with some ductular pattern. Occasional spindles cells noted. Cell block showed hemorrhage and inflammatory cells. Further excision biopsy (Figure 3) was performed to relieve the mass effect of the lesion over the neurovascular bundle in right axilla. The excised lesion was sent for fluid cytology, histopathological and immuno histochemistry.

Biopsy And Fluid Cytology:
Fluid cytology:
fluid from the cystic lesion showed low cellularity which comprised of few clusters of small round cells – the possibility of a small round cell tumor was considered. **Biopsy** from the cyst wall and contents of the lesion showed fibrocollagenous tissue infiltrated by a malignant tumor arranged in sheets and nests. Increased mitosis with few atypical mitoses evident – features suggestive of malignant small round blue cell tumor. Subsequently, Immunohistochemistry was advised (Figure 4).

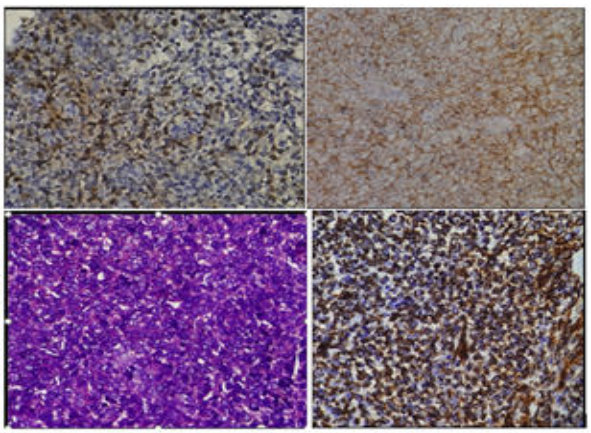

Figure 4:
Cyclin D positive (left upper corner), CD99 diffuse and membraneous positivity (right upper corner), Hematoxylin and eosin : small round blue cells (left lower corner), Vimentin positive (rigth lower corner).

Source: Pondicherry institute of medical sciences.

Final Diagnosis:
Final diagnosis of Ewing's sarcoma family of tumors – extraosseous Ewing's sarcoma was made. Patient is under chemotherapy.

DISCUSSION:
The Ewing sarcoma family of tumors (ESFT) is a collection of small, rounded tumor cells that have similar neural histological and genetic characteristics. It can affect different locations, without specific clinical signs, which delays the diagnosis (5).ESFT is categorized into four types based on the origin of the tumor: Ewing sarcoma of the bone, peripheral primitive neuroectodermal tumor (pPNET), Askin tumor, which originates from the chest wall, and, finally, the extraosseous or extraskeletal Ewing sarcoma (EES). EES, which occurs in around 20% of ES cases, typically originates from the soft tissues of the trunk and extremities(4). Extraskeletal tumors are more likely to arise from axial locations and less likely to arise from the pelvis(6)

In our case, Extraosseous Ewing sarcoma was noted on the right side of the chest wall. EES arises in the soft tissues of the trunk, paravertebral region, intercostal area, lower extremities, and pelvis

Around 75% patients present with rapidly growing painless mass, 30% patients exhibiting distant metastasis at the time of diagnosis. The tumor grows locally without any alarming inflammatory signs. This was the case with our patient, the tumor increased in size with time, without significant inflammatory signs.

With Doppler US, EES is usually a heterogeneous mass containing flow signals. Computed tomographic (CT) scan may show a low attenuation mass with heterogeneous enhancement with contrast administration. MRI findings include signal isointense to muscle on T1-W images and hyperintense on T2-W images with heterogeneous enhancement after gadolinium administration. Spontaneous tumor hemorrhage, adjacent bone destruction and regional metastatic adenopathy have been demonstrated in some cases using MRI or CT, but no evidence of tumoral calcification prior to treatment has been reported.(7).

The histology of ESFT can be described as “classical”, “atypical”, or “variant”(8).These tumors consist of primitive-appearing round cells with high nucleus-to-cytoplasm ratios. The immunohistochemical features of ES/PNET are positive for CD99 (a 32-kDa cell surface glycoprotein encoded by the MIC2 gene); however, expression of CD99 is by no means specific for ES/PNET among round-cell tumors. Although FLI-1 is a variable histochemical marker for ES/PNET.

The second approach is electron microscopic examination of tumor tissue. Electron microscopic features include a specific high nucleus-to-cytoplasm ratio and aggregated glycogen granules in the cytoplasm. The third approach are molecular genetic studies by polymerase chain reaction (RTPCR) or FISH to detect chromosomal translocation, such as t(11;22)(q24;q12) which is positive in 88–95% of ES/PNET cases.(1) Because EES represents a relatively rare clinical entity, there are limited data to guide optimal local therapy(9).

The golden standard of treatment is surgery. There is no potential for cure in sarcoma patients without margin-negative surgery. Chemotherapy is provided after surgery to improve overall survival rates and reduce the likelihood of tumor recurrence. A combination of several agents is used to obtain a higher response rate.

First-generation regimens consisted of the combination of vincristine, cyclophosphamide, actinomycin D and doxorubicin (VAcCD). Second-generation regimens incorporated ifosfamide and later etoposide with improved disease-free survival for patients with localized disease. Prognostic factors and optimal therapy are still unconfirmed.(10)

The current generation of clinical trials has attempted to improve survival by maximizing the chemotherapy dose per cycle, increasing the total number of cycles provided, or decreasing the interval between cycles ('dose-dense' therapies).(1)

CONCLUSION:
Extraskeletal Ewing sarcoma is a rare tumor mainly affecting young people(5). Radiological signs and histopathological analysis play major role in diagnosis and appropriate treatment plan for the disease entity. This entity must be kept in mind as differential diagnosis for identification and early treatment.

Teaching Points:
Extraosseous ewing sarcoma can occur in various forms and rare radiological presentations. Thus, radiological evaluation and clinicopathological correlation should include suspicion of this disease entity.

Careful interpretation and imaging characterization in such disease entity can help in diagnosis and prognosis of the patient.

Funding Information:
This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Ethical Consideration:
This article followed all ethical standards for carrying out research.

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