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Rare Case of Primary Extraskeletal Ewing Sarcoma on the Anterior Chest Wall Diagnosed by Fine Needle Aspiration Cytology

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Abstract: ***The rarity and deceptive presentation of extraskeletal Ewing sarcoma (EES) demand a sharper clinical eye and a multidisciplinary diagnostic approach. This case report details a rare incidence of subcutaneous EES in a 32-year-old male, highlighting how its subtle presentation initially as a firm, mobile chest wall swelling can obscure the underlying malignancy. What stands out is the role of fine needle aspiration cytology (FNAC), which, despite its limitations, proved effective in raising suspicion for a small round cell tumor. Histological evaluation and immunohistochemical markers such as CD99 and NKX2.2 solidified the diagnosis, aligning it with the broader Ewing/PNET spectrum. It is evident that modern diagnostics especially immunohistochemistry and molecular tools are redefining the diagnostic pathways for tumors like EES, which historically relied solely on histopathology. This case, although uncommon, illustrates a larger truth: early and minimally invasive diagnostic tools, when paired with aggressive treatment strategies, can meaningfully shift survival outcomes, even in aggressive tumors with initially grim prognoses. That said, the nuanced differential diagnosis against other round cell neoplasms reminds us that precision in pathology is not optional it's lifesaving. Taking this further, the discussion also underlines how FNAC, often underestimated, has quietly become a cornerstone in the early detection of rare but deadly malignancies like EES.***

Keywords: extraskeletal Ewing sarcoma, FNAC diagnosis, small round cell tumor, CD99 immunohistochemistry, aggressive soft tissue tumor

1. Introduction

In 1921, James Ewing first described what is now recognized as Ewing sarcoma. Initially classified as an undifferentiated type of bone sarcoma affecting children, Ewing sarcoma is now associated with Primitive Neuroectodermal Tumor (PNET). Thanks to recent advancements in cytological, histological, molecular, and genetic studies, the term *Ewing sarcoma/PNET* is now preferred to describe this group of tumors. (1)

Extraskeletal Ewing's sarcoma (ES) is a rare subtype within the broader family of Ewing's sarcoma tumors (ESFT). These tumors are characterized by small, round cells that exhibit varying levels of neuroectodermal differentiation. (2)

Ewing sarcoma accounts for 4 - 10% of all primary bone cancers, primarily affecting adolescents and young adults, with rare occurrences after the age of 30. The average age of onset in the head and neck region is 10.9 years. This cancer typically affects the white population and is more common in males, with a male - to - female ratio of 1.3 - 1.5: 1. Based on its anatomical location, Ewing sarcoma is classified into three types: (a) intraosseous (most common), (b) extraskeletal (less common), and (c) periosteal (rare). (3)

2. Case Report

A 32 year old man presented presented to the surgery OPD with complaints of swelling over right lower chest wall since 2 years. (Fig 1) Examination revealed a single. Tender, subcutaneous, mobile, firm swelling approximately measuring 6.5 x 5.2 cm.

Ultrasonography of local part of right chest revealed a well defined lobulated heteroechoic mass lesion measuring 5.2 x 6.5 x 2.0 cm in subcutaneous plane along left lateral chest wall protruding in between intercostal spaces. (fig 2)

FNAC was performed using aspiration, cellular smears were obtained show neoplastic cells arranged predominantly as scattered singly, loosely cohesive clusters and few tissue fragments. Individual cells are small round having round nuclei with mild irregular nuclear membrane, granular chromatin with indistinct nucleoli and scanty rim of cytoplasm. Few of the cells show moderate amount of cytoplasm with cytoplasmic vacuolations. At places nuclear overlapping, crowding and nuclear molding is seen. Many dispersed stripped nuclei are also seen. At places tumor cells are arranged in rosette like pattern. Background shows stromal fragments and RBCs.

Based on the above findings diagnosis of small round cell tumour was given with suggested differential of Ewings sarcoma.

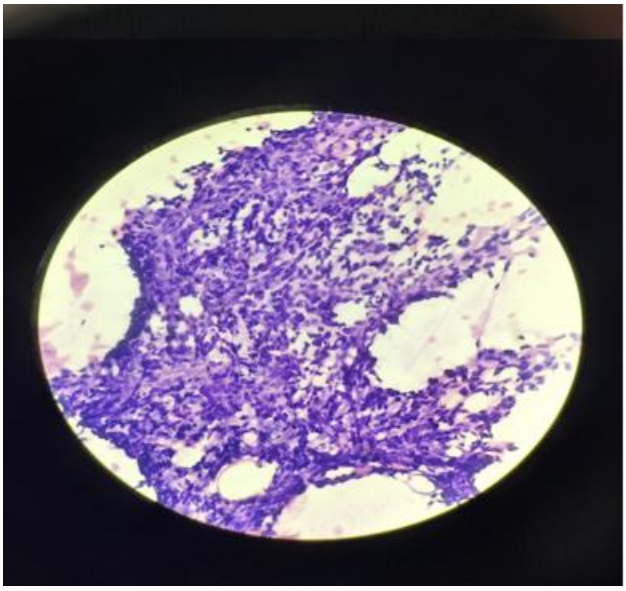
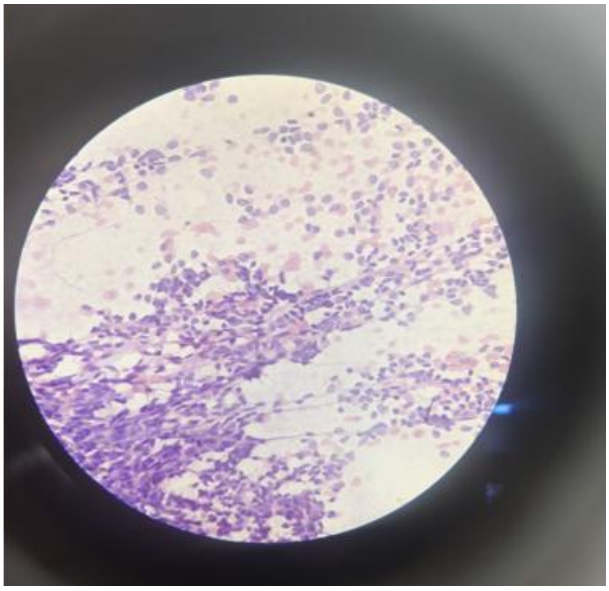

Figure 1 and 2: Tumor cells arranged in rosette like pattern (400X)

On histopathology microscopy revealed a tumor composed of sheets, nests of neoplastic cells having round morphology, uniform round nuclei with finely stippled chromatin. Rosette formation was also noted. differentiasl diagnosis of Ewing's sarcoma, Non hodgkin's lymphoma and other small round blue cell tumor was given. On immunohistochemistry the neoplastic cells expressed CD99 and NKX2.2 confirming the diagnosis of Ewing's sarcoma.

Pre - operative cytological diagnosis of Ewing sarcoma, including subcutaneous cases, is exceedingly rare, with only a limited number of instances reported based on fine needle aspiration (FNA) cytology findings.

3. Discussion

Ewing sarcoma serves as the prototype for small round blue cell tumors. It is now classified as part of the Ewing sarcoma/PNET (Primitive Neuroectodermal Tumor) family, reflecting its association with tumors that share similar histological and molecular features. (4)

Thanks to the advancements in immunohistochemistry (IHC), cytogenetics, and molecular genetic techniques, these tumors are now recognized as a spectrum known as the Ewing family of tumors. This spectrum includes Ewing's sarcoma of bone, extraskeletal Ewing sarcoma, peripheral primitive neuroectodermal tumors (PNET), and Askin tumors. The common cytogenetic abnormality observed in both Ewing's sarcoma and PNET is the translocation t (11; 22) (q24; q12), which plays a key role in the pathogenesis of these tumors. (5)

Extraskeletal Ewing sarcoma (EES) is a rare, fast - growing, and highly aggressive soft tissue tumor that can occur at any age, from infancy to old age. While it can develop in virtually any location, the most common sites are the paravertebral and intercostal regions, followed by the extremities. (4)

Ewing sarcoma (EES) typically originates in the soft tissues of the trunk. However, it can also affect other areas, including the larynx, nasal fossa, neck, lungs, retroperitoneum, perineum, and mediastinum. (6, 7)

The clinical manifestations of extraskeletal Ewing sarcoma (EES) are often nonspecific. Patients may present with a painless mass or swelling in any part of the body.

While histopathology remains the gold standard for diagnosing extraskeletal Ewing sarcoma (EES), aspiration cytology, followed by immunohistochemistry (IHC) or cell block analysis, has increasingly become a reliable diagnostic approach. EES is a highly malignant round cell tumor known for its aggressive clinical behavior. (7)

Because EES lacks distinct morphological features, its differential diagnosis includes other small round blue cell tumors, such as rhabdomyosarcoma (RMS), lymphoma, synovial sarcoma, and neuroblastoma. (7, 8, 9)

The EWS - FLI1 gene rearrangement is highly specific to Ewing sarcoma (ES) and peripheral primitive neuroectodermal tumors (PNET), occurring in over 90% of cases. (10)

These tumors exhibit strong CD99/MIC2 membrane positivity, which is useful in distinguishing them from neuroblastoma and other round cell tumors. (10)

The five - year survival rate for skeletal Ewing sarcoma is approximately 75%. In contrast, extraskeletal Ewing sarcoma (EES) generally has a poorer prognosis, with a five - year survival rate of around 38%. However, studies have shown that with aggressive treatment, including multi - agent chemotherapy and achieving a wide, tumor - free surgical resection margin, survival rates can improve significantly, reaching up to 61%. This highlights the importance of early detection and a comprehensive treatment approach to enhance outcomes in patients with EES. (7, 9) Conclusion:

Recent advancements in diagnostic techniques and treatment protocols, such as multidrug chemotherapy, have shown improved prognosis in cases of extraskeletal Ewing sarcoma

(EES). Fine needle aspiration cytology (FNAC) offers a less invasive alternative, providing a reliable diagnosis. As a result, FNAC has become an increasingly rapid, dependable, and patient - friendly diagnostic method.

References

[1] Ewing J. Diffuse endothelioma of bone. Proc N Y Pathol Soc.1921; 21: 17
[2] Bordia S, Meena S, Meena BK, Rajak V. Fine - needle aspiration cytology of Ewing's sarcoma of thoracic spine with extension into the intradural space Case Rep Oncol Med.2014; 2014: 351386
[3] Kollender Y, Shabat S, Nirkin A, Issakov J, Flusser G, Merimsky O, et al. Periosteal Ewing's sarcoma: Report of two new cases and review of the literature. Sarcoma 1999; 3: 85 - 8.
[4] Khatib WM, Jagtap SV, Kaleet P et. al. Primary extra skeletal Ewing sarcoma presenting as soft tissue mass diagnosed on Fine Needle Aspiration Cytology. Int J Health Sci Res.2015; 5 (10): 377 - 380.
[5] Malignant Soft tissue tumors of Uncertain type. Editors: John R Goldblum, Andrew L Folpe, Sharon W. Weise. In: Enzinger& Weiss Soft tissue tumors, 6ed. Philadelphia, Elsevier; 2014: p1028 - 1112.
[6] Ahmad R., Mayol B. R., Davis M. &Rougraff B. T. Extraskeletal Ewing's sarcoma. Cancer, 85 (3), 1999, 725 - 731.
[7] Buch A. C., Panicker N. K., Sarawagi S., Anwekar S., Kharat A. T. Fine needle aspiration cytology diagnosis of paravertebral extraosseous Ewing's sarcoma. Journal of cytology/ Indian Acedemy of Cytologists, 27 (4), 2010, 146.
[8] X Chun - Fang. M - Z Liu, and M Xi. "Extraskeletal Ewing's sarcoma: a report of 18 cases and literature review. " Chinese journal of cancer 29.4.2010, 420 - 424.
[9] Manduch M., Dexter D. F., Ellis P. M., Reid K., Isotalo P. A. Extraskeletal Ewing's sarcoma/primitive neuroectodermal tumor of the posterior mediastinum with t (11; 22) (q24; q12). Tumori, 94 (6).2008, 888.
[10] Dr. Bharathi M, Dr. NeelimaPrabha C, Dr. Vani D, Dr. Sharath Kumar H. KExtraosseous Ewing Sarcoma – Cytological Diagnosis of a Rare Entity IOSR Journal of Dental and Medical Sciences, 2015; 14 (2) 21–24.