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Abdominal Wall Extraskeletal Ewing Sarcoma - Case Report

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***Abstract**: Ewing sarcoma is most commonly occurring as bone tumor which has usually extended into the soft tissues at the time of diagnosis. Exceptionally, this tumor rarely can have an extra skeletal origin without relation to bone as a primary tumor. Clinical or imaging findings are non-specific and diagnosis is based on histology. We report a case of an extra skeletal Ewing sarcoma developed in the soft tissues of the abdominal wall in a 30-year-old woman who presented a painful abdominal wall tumefaction. Ultrasonography and computed tomography showed a large, well-defined soft tissue mass (? hematoma) developed in the anterior the abdominal wall. Surgical biopsy was performed and an extraskeletal Ewing sarcoma was identified histologically*

***Keywords:** Ewing's sarcoma, soft tissue tumor, extra skeletal, abdominal wall, rosettes.*

I. Introduction

Tumors morphologically indistinguishable from Ewing sarcoma of the skeletal system can present as soft tissue masses. In some cases, they simply represent soft tissue extensions of tumor originating in the underlying bone. In others, bone involvement is absent, and these are regarded as primary Ewing sarcomas of soft tissues [1,2,3]. Most of the patients are adolescents or young adults, and the usual sites of involvement are the deep soft tissues of the lower extremity and paravertebral region [5]. They have been described in many other sites, including digits and craniospinal vault[5,6]. Occasionally, the tumors are superficial, with primary involvement of subcutis or skin [7,8].

II. Case Report

A 30 year old female patient presented with swelling in anterior the abdominal wall. On ultrasonography and computed tomography showed a large, well-defined soft tissue mass developed in the anterior abdominal wall and they diagnosed as ? hematoma / ? cystic lesion. On FNAC (fine needle aspiration cytology) it is reported as non-specific inflammatory lesion because only inflammatory cell and hemorrhagic material is observed in smear prepared from aspirated material. In view of clinical suspension the patient is operated and the specimen is sent for histopathological examation.

On gross examination [Figure1] the specimen is globular in shape measuring 12×11×9cms with one side covered with skin flap measuring 9×5.5×5cms. External surface is dark-brown. On cut section cystic and solid grey-white areas present along with haemorrhagic and necrotic areas.

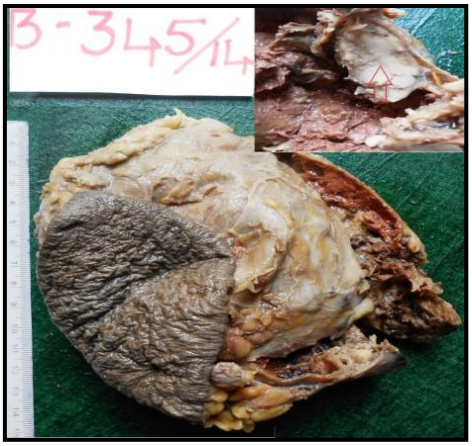

Figure1: Gross show globular skin covered soft tissue mass & cut section shows cystic and solid grey-white (arrow) areas (insert)

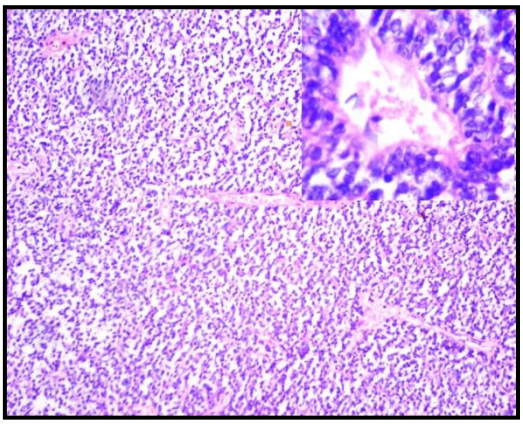

Figure2: Low power (100X) microscopic picture shows small blue round cells & most of the cells are arranged around blood vessel- perivascular(insert)

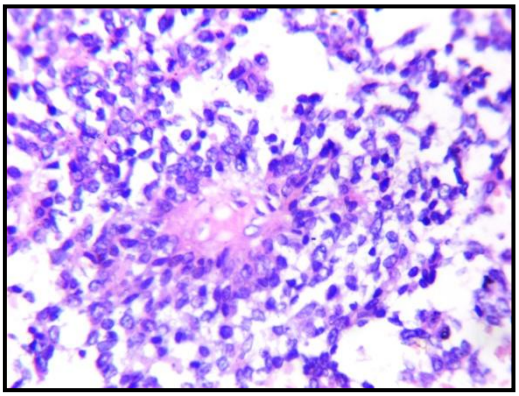

Figure3: High power (400X) microscoipc picture showing rosettes (central solid eosinophilic core surrounded by small blue cells like rosette)

On microscopic examination [Figure2, Figure3] shows small blue round cells with round nuclei containing fine chromatin, scanty clear or eosinophilic cytoplasm, and indistinct cytoplasmic membranes. Areas showing Homer–Wright rosettes (central solid eosinophilic core containing neurofibrillary material surrounded by small blue cell like rosette) and necrosis are present. Most of cells are perivascular in distribution.

III. Discussion

Ewing sarcoma commonly arises from bone but can rarely, Ewing sarcoma may have an extraskeletal soft tissue tumors of neuroectodermal origin. Soft tissue Ewing sarcoma is a rapidly growing, round-cell, malignant tumor which can reach 10 cm by the time of the diagnosis [5]. Young adolescents and adults between the ages of 10 and 30 years are predominantly affected with a slight predominance in males [4]. Commonly affected extraskeletal sites are the paravertebral spaces, lower extremities, head and neck, and pelvis [5,6]. Other rare reported locations of extraskeletal Ewing sarcoma are varies and include the retroperitoneum, orbit, skin, and chest wall, abdominal wall[7,8]. The imaging features of soft tissue Ewing sarcoma are non-specific [9]. Histological assessment shows that these tumors are composed of sheets of small round cells with hyperchromatic nuclei, and scanty cytoplasm.

Immunohistochemically, a number of markers have been used, of which MIC2 (CD99) is particularly useful in eliciting membranous positive staining of tumor cells [10] Cytogenetically, there is a chromosomal abnormality, with translocation of t (11:22) (q24:q12) being detected in 85% of cases using standard cytogenetic techniques, and in up to 95% of cases using reverse transcriptase and PCR[10]. There is no established predisposing, or genetic factors associated with PNET although there are anecdotal associations: in one case the sibling of a patient died of Ewings sarcoma of bone and in another the mother of the patient was treated for osteosarcoma of tibia and myxoid liposarcoma of retroperitoneum. PNET generally presents as a rapidly growing, deeply located mass that tends to metastasise early in the course of the disease. For this reason, although local control can be attempted with surgery and radiotherapy, PNET invariably requires systemic chemotherapy [11].

In conclusion, extraskeletal Ewing sarcoma arising from soft tissues is extremely rare. Although its radiological features are non-specific, Histopathological examination confirms the diagnosis.

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