Extraosseous Ewing's tumor of lateral abdominal wall

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Abstract

Extraosseous Ewings tumor (EES) is a rare entity. Few cases have been reported in literature. There are no specific guidelines for management of this disease. We are reporting a case of EES arising from left lateral abdominal wall. We did wide excision of tumor followed by chemoradiation. Patient is asymptomatic after 8 months of follow up.

Introduction

Extraosseous Ewings tumor (EES) has its origin from soft tissue and belongs to family of small round cell neoplasms /PNET'S.[1] About 82 cases of cutaneous or subcutaneous ES reported in the literature.[2] We are reporting this case because of the rarity of this tumor. Very few cases have been reported up till now; the treatment is still in its infancy.

Case Report

A 23-year female presented to surgical outpatients department with a painless lump in the left flank, which had been progressively increasing in size for seven months (Figure 1). There was no history of fever, cough, chest pain, burning micturition, hematuria, alteration of bowel habits, bleeding per rectum or malena. On examination, there was an approximatley 15×15 cm round variegated firm-hard intraparietal lump over the left lumbar and left iliac fossa with a 5×5 cm overlying non-healing ulcer. Clinically a diagnosis of soft tissue sarcoma was made. All routine blood investigations and chest X-ray were normal. Contrast enhanced CT scan of chest and abdomen revealed a 14×10×7 cm well defined lobulated, heterogenous, enhancing, hypodense lesion in the left lateral abdominal wall with ill-defined margins with underlying abdominal wall muscle with no evidence of intraperitoneal extension (Figures 2 and 3). There was no free fluid and no evidence of metastasis in chest or abdomen. Core biopsy suggested small round cell tumor. Wide excision of the tumor was planned under general anesthesia. Intraoperatively, the mass was found to be originating from the external oblique aponeurosis and involving few fibers of the internal oblique muscle; transversus abdominis muscle and tissues below were free. Reconstruction of the abdominal wall was performed using polypropylene mesh (15×15 cm) (Figure 4). The postoperative period was uneventful. Histopathology of the resected specimen showed small round cells with vesicular nuclei, small inconspicuous nulceoli with mild pleomorphism with pseudorossette formation (Figure 5). All the margins were free of tumor. Immunohistochemistry revealed CD99 and PAS positivity whereas it was negative for desmin, chromogranin, LCA and cytokeratin. Thus a diagnosis of EES was established. The patient received post-operative radiotherapy (Total 54Gy with 1.8Gy per fraction) and chemotherapy VAC with IE regime (vincristine, adriamycin, cyclophospamide, ifosfamide, etoposide). The patient is doing well after eight months of follow up.

Discussion

In 1921, James Ewing first described the tumor of bone, which is now known as Ewing's sarcoma.[3] Extraosseous Ewing's (EES) tumor is a rare entity; few cases have been reported in literature. These tumors occur most frequently during the second decade of life (64%).[2] There is no preferential site for these tumors; they have been reported on the trunk, limbs, face, and scalp and no gender or racial differences have been reported.[4] Confirmation of the diagnosis is made by histological examination and immunohisto-chemical examination; the diagnosis is often difficult.[5] The histological image of cutaneous or subcutaneous ES is a small blue round cell with a scanty cytoplasm, and it is often confused with other skin tumors.[6] The markers are of great importance for establishing the differential diagnosis with other entities.[7] Negative leukocyte common antigen (LCA), CD30, myosin, actin, myoglobulin, neu-

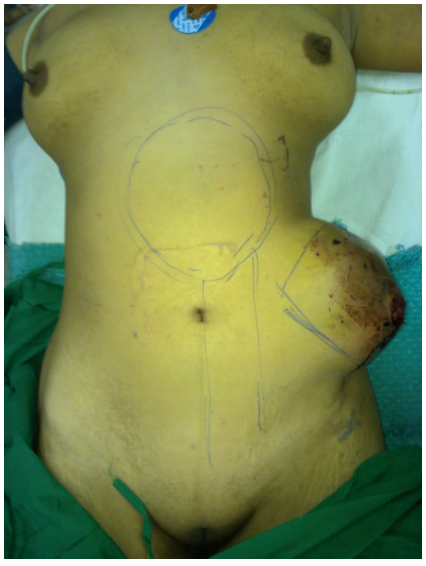

Figure 1. Lump in the left lumbar region.

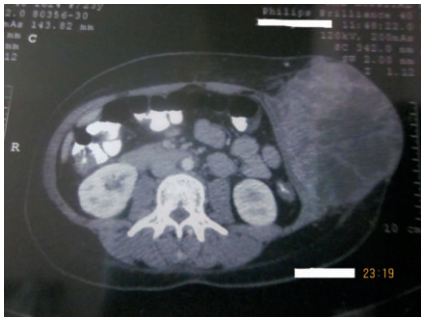

Figure 2. Contrast enhanced CT (transverse section) showing intraparietal mass in left lumbar region with no intraperitoneal extension.

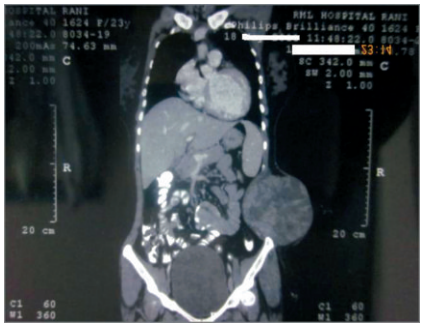

Figure 3. Contrast enhanced CT (longitudinal section) showing same mass with normal liver and lung fields.

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Key words: Ewings, extraosseous, chemoradiation.

Received for publication: 19 October 2011.
Revision received: 19 October 2011.
Accepted for publication: 2 November 2011.

Clinics and Practice 2011; 1:e135
doi:10.4081/cp.2011.e135

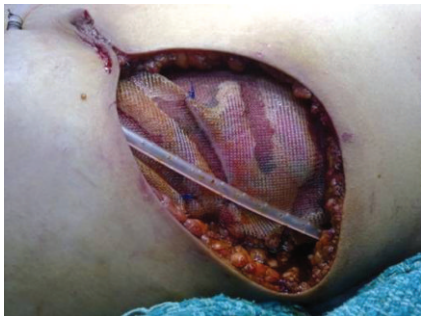

Figure 4. Reconstruction of abdominal wall using polypropylene mesh.

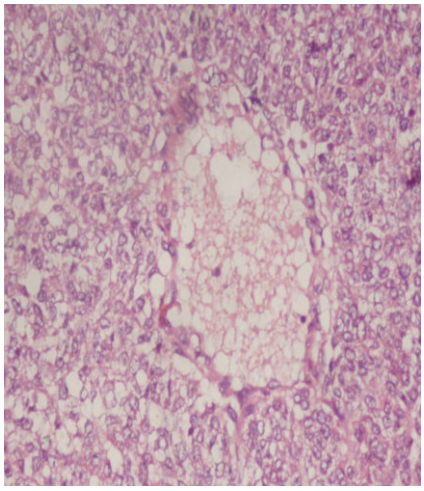

Figure 5. Histopathology showing small round cells with vesicular nuclei, small inconspicuous nucleoli. Cells showing mild pleomorphism with pseudorossette formation.

rofilament, neuron-specific enolase and S-100 protein can exclude lymphoblastic lymphoma, rhabdomyosarcoma and neuroblastoma.[8] Basaloid adnexal tumors of the skin are negative for CD99. Some pathologists regard ESS as a completely different disease entity from PNET. Although both ESS and PNET show expression of HBA-7 and the t11:22 translocation, only ESS contains PAS-stained glycogen in the cytoplasm.[9] Treatment for cutaneous Ewing's sarcoma, though not codified, consists of surgery associated with polychemotherapy and/or radiotherapy.[2,6] The current view is that surgical resection should be performed if the tumor can be completely resected. A local wide resection is the recommended operation, with the goal of a complete resection of tumor lesions and negative surgical margin, and the extent should include a 2-3 cm normal tissue surrounding the tumor if possible.[1,10] Adjuvant multidrug chemotherapy has a definitive survival advantage.[11,12] If the surgical margin is positive or insufficient, or the tumor location is at a special site or is bulky, RT may be practicable.[13]

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