Case Report

Extraskeletal Ewing sarcoma of the anterior abdominal wall

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ABSTRACT

Extraskeletal Ewing sarcoma (EES) is a relatively rare primary tumor of the soft tissues, it accounts for 20–30% of all reported cases of EES which is a neoplasm that usually occurs in children and young adults and presents as an undifferentiated primary bone tumor. Infrequently, this tumor can have an extraskeletal origin, known as EES. We report a case of Ewing's tumor involving the anterior abdominal wall without any associated skeletal location. The clinical examination suggested a benign neoplasm arising from the subcutaneous tissue. However, ultrasound, magnetic resonance imaging, and fine-needle aspiration that were done preoperatively suggested a malignant neoplasm. No evidence of metastasis was present. The diagnosis of EES was confirmed after surgical excision. Histopathological and immunohistochemical examinations were helpful in the confirmation of the diagnosis. The patient had an uneventful post-operative recovery. He was started on chemotherapy and is on regular follow-up.

Key words: Ewing sarcoma, Ewing sarcoma family tumor, Extraskeletal Ewing sarcoma, Sarcoma tumors

Extraskeletal Ewing sarcoma (EES) is a relatively rare primary tumor of the soft tissues, it accounts for 20–30% of all reported cases of Ewing sarcoma [1-3]. The incidence of EES is 0.4 per million which is 10 times less than Ewing sarcoma of the bone [4]. The prevalence EES follows a bimodal distribution, with peaks in those who are under 5 years and over 35 years [5]. EES develops rapidly into a painful mass within the soft tissues of any anatomic region, but the most common sites include the upper thigh, buttocks, upper arm, and shoulders [6]. Conversely, metastasis is commonly observed in the lungs, bones, and bone marrow [7]. In addition, EES does not show specific clinical signs causing a delay in the diagnosis [8]. Furthermore, the symptoms of EES depend on its primary site as well as the site of metastasis, which are found in 25% of all cases at presentation [6]. EES being rare, all members of the Ewing sarcoma family tumors are treated following the same general protocol of sarcoma tumors.

We present a similar patient who reported to our institute with a benign-looking swelling over the anterior abdominal wall, which was excised with a wide surgical margin, and confirmed on histopathological examination as EES. The diagnostic dilemma and the overall outcome are discussed in this case report. The rationale for reporting this case is the following: The rarity of the disease, diagnostic dilemma, surgical excision, and histopathology were needed for the confirmation of the diagnosis, and no standard protocol for treatment of these tumors is available due to the rarity.

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CASE REPORT

The patient was a middle-aged gentleman of average build and nutrition who came with complaints of a lump in the upper abdomen for 3 months which was gradually progressing in size.

His vitals at the time of reporting were pulse 86/min, blood pressure 130/72 mmHg, and the temperature was afebrile. There was no pallor or lymphadenopathy. On abdominal examination, there was a swelling of size 6 × 4 cm in the upper abdomen. It was firm to hard in consistency and was superficial to the muscles of the anterior abdominal wall (Fig. 1).

Ultrasonography of the abdomen revealed a heterogenous, predominantly hypoechoic lesion in the left periumbilical region in the subcutaneous plane. Magnetic resonance imaging (MRI) showed a mass lesion in the subcutaneous tissue plane of the left periumbilical region, having a lobulated outline measuring approximately 5.3 × 5.5 × 3.5 cm (TR × CC × AP), also showing evidence of T1/T2 hypo/isointensity changes. Loss of fat planes between the lesion and left-sided rectus abdominis muscle was noted (Fig. 2). A fine-needle aspiration cytology was performed and was suggestive of a small round blue cell tumor (Fig. 3).

Wide excision of the tumor was performed with a margin of 2 cm. The resulting defect was covered with a 15 × 15 cm

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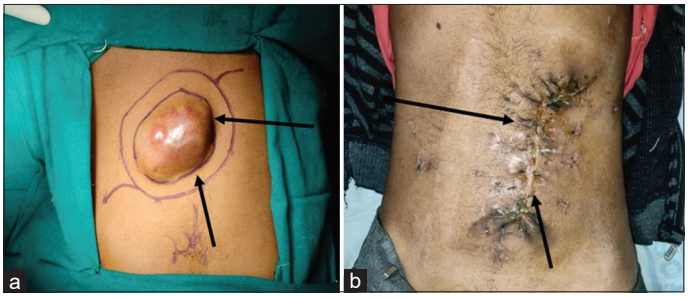

Figure 1: (a) Clinical photograph (pre-operative) showing a 6 × 5 cm oval lump in the anterior abdominal wall and (b) clinical photograph (3 weeks post-operative) showing healed surgical scar

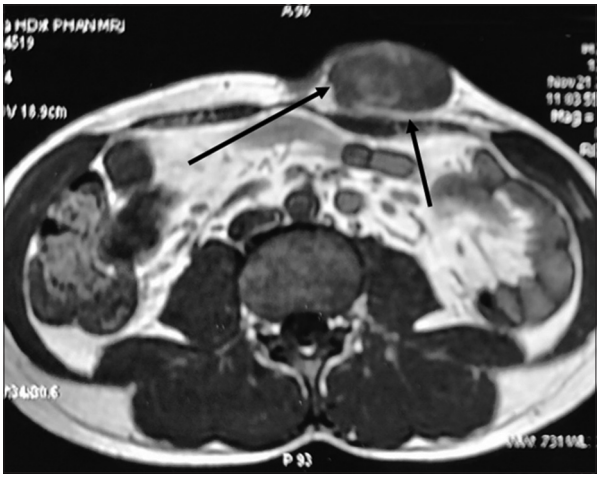

Figure 2: Magnetic resonance imaging (axial cut) showing a mass lesion in the subcutaneous plane of left paraumbilical region

polypropylene mesh placed anterior to the abdominal wall muscles. A 16F suction drain was placed over the mesh and the defect was closed primarily.

Histopathological examination showed a highly cellular tumor comprising sheets and nests of malignant small round cells (Fig. 4a). Pseudo rosette formation was also seen along with papilionid areas consisting of thick vascular channels with attached groups of tumor cells (Fig. 4b). Immunohistochemical examination showed diffuse CD99 positivity (Fig. 5) and Ki-67 being 35% in the tumor cells. The cells were negative for leukocyte common antigen, desmin, and PanCK, which excluded the diagnoses of lymphoma, rhabdomyosarcoma, and undifferentiated carcinoma, respectively. An impression of Ewing sarcoma was made on the basis of histopathology and immunohistochemistry.

The patient had an uneventful post-operative course and discharged in good general condition. He was advised chemotherapy with vincristine, adriamycin, and cyclophosphamide (VAC) alternating with ifosfamide and etoposide (IE). The patient is on regular follow-ups both in the surgical and oncology clinics.

DISCUSSION

Ewing sarcoma is the second most common primary sarcoma of the bone in children and adolescents after osteosarcoma [9]. EES is a rare entity that belongs to the Ewing sarcoma family of tumors,

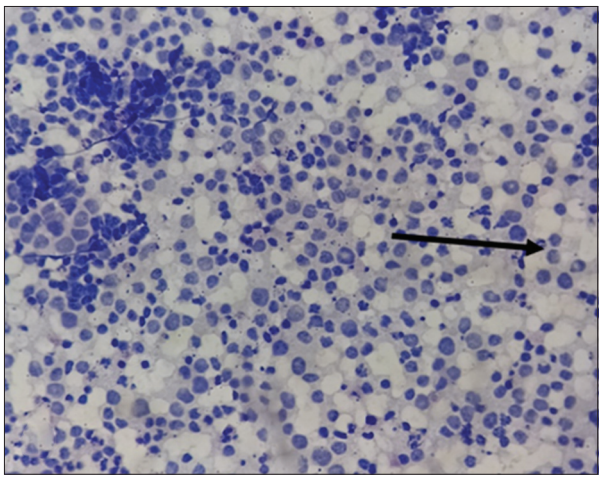

Figure 3: (a) Fine-needle aspiration cytology (×40): Pap smear shows predominantly singly scattered relatively monomorphic small round blue cells with focal resetting

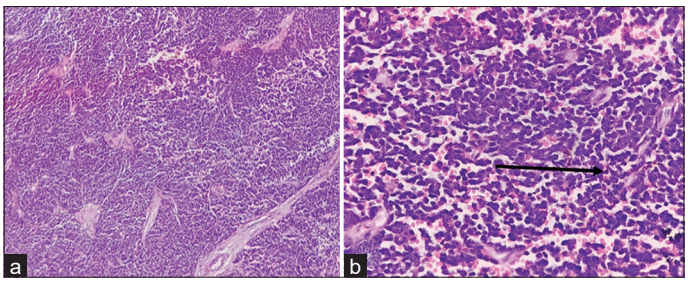

Figure 4: (a) H- and E-stained section shows solid sheets and nests of small blue round cells; (b) higher magnification (H and E) shows small round blue cells with minimal cytoplasm, medium size nuclei with coarse chromatin, and inconspicuous nucleoli. At places, the cells form rosette-like structures

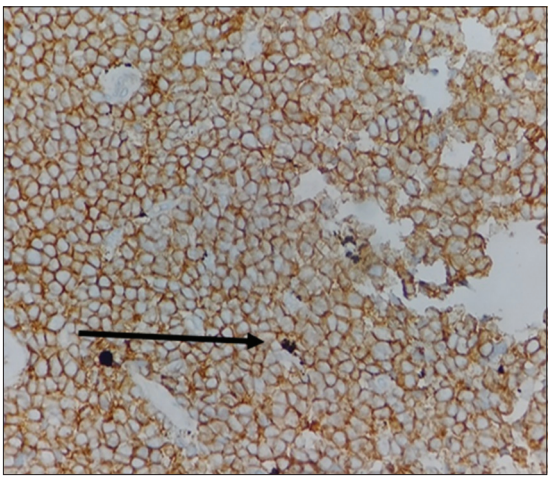

Figure 5: Immunohistochemical (×40) magnification shows diffuse strong and membranous positivity for CD99

which accounts for 20–30% of Ewing sarcoma. It is commoner in males with affection toward children, adolescents, and young adults [10]. Paravertebral spaces, pelvis, lower extremities, and head and neck are the common sites of its occurrence [11,12]. In our patient, EES was present in the anterior abdominal wall.

The overall incidence of EES is 0.4 per million [4]. Three cases of anterior abdominal wall EES were reported earlier, one

each by Aydinli *et al*. [13], Askri *et al*. [12], and Liu *et al*. [11]. In the current case, EES was diagnosed in a middle-aged male supporting the earlier reports on its age distribution [5].

The modality of choice for diagnostic imaging and local staging is MRI, on which this tumor is often of low to intermediate signal intensity on T1-weighted images, of high-signal intensity on T2-weighted images, and exhibits heterogeneous contrast enhancement. However, core-needle biopsy is required for a definitive diagnosis, which, irrespective of the primary site, comprises the characteristic small round blue cells [1,14]. Pathologically, it is a gray-yellow tumor with cystic, hemorrhagic, or necrotic areas, but calcifications are unlikely.

CD99 antigen is a highly sensitive marker for Ewing sarcoma, which was crucial in the diagnosis in the present case. Friend leukemia integration 1 was recently discovered as a DNA-binding transcription factor involved in translocation and has higher specificity than CD99. However, this marker could not be tested in our patient.

Differential diagnoses of Ewing sarcoma include primitive neuroectodermal tumor and other small, blue round cell tumors [14], but immunohistochemistry and histopathological examination is the cornerstone for diagnosis.

Ewing sarcoma is treated by surgery, radiation, and multidrug chemotherapy [9]. Single-modality local therapy alone may be sufficient for local disease control since the role of combined-modality local therapy in EES remains unclear [15]. Our patient underwent wide local tumor excision with primary closure of the defect followed by chemotherapy with VAC/IE regimen, as suggested in the treatment of non-retroperitoneal/visceral EES patients [16].

Common metastatic sites of Ewing sarcoma are lungs, bone, and bone marrow [7]. Therefore, a post-operative computed tomography of the thorax was done in our patient to look for metastasis and the imaging revealed no such finding. However, the patient is still at risk of development of metastasis; hence, he is on regular follow-up and metastatic workup including fluorodeoxyglucose-positron emission tomography scan.

CONCLUSION

The management of this EES is still in the evolving stage. Wide surgical excision, followed by chemotherapy using VAC/IE regime could be used as in the present case. Definitive radiotherapy is only indicated for inoperable lesions with a recommended dose of 54–55 Gy, depending on the site involved. The prognosis of EES is more favorable compared to the skeletal subtype. Risk factors associated with poor prognosis in EEs include older age, pelvic involvement, high white blood cells, elevated lactate dehydrogenase enzyme, and low hemoglobin at the time of diagnosis. Hence, patients without these risk factors, as in the present case report may have a better overall survival. However, it is necessary to advise frequent follow-up visits to monitor for the appearance of metastasis.

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