Primary Extra-Skeletal Ewing Sarcoma of the Inguinal Region a Rare Case Report and Review of the literature

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Abstract

Ewing sarcoma is most commonly a bone tumor which has usually extended into the soft tissues at the time of diagnosis. Exceptionally, this tumor can have an extra-skeletal origin. Clinical or imaging findings are nonspecific and diagnosis is based on histology. We report a case of an extra-skeletal Ewing sarcoma developed in the soft tissues of the inguinal region in a 28-year-old man who presented a painless right inguinal mass for 2 months. Ultrasonography showed a large, well-defined soft tissue mass developed in the right inguinal region with tender inguinal Lymph Node (LN) Surgical biopsy was performed and an extra skeletal Ewing sarcoma was identified histologically.

Keywords: Ewing sarcoma • Soft tissue neoplasms • Imaging • Ultrasonography

Introduction

Extra-skeletal Ewing sarcoma (EES) is a rare, round cell malignant neoplasm that is histologically indistinguishable from the more common osseous Ewing sarcoma [1, 2]. Both EES and Ewing sarcoma are derived from the same neuroectodermal cells that share the same cytogenetic marker, with translocation of chromosomes t(11;22) (q24;q12) [3]. The medical terminologies "Peripheral Primitive Neuroectodermal Tumor" (PNET) and "Askin tumour" (thoracopulmonary PNET) are no longer used. This is because, in terms of histological appearance and morphology, PNET and Askin Tumour are identical to EES. Consequently, for the sake of simplicity and clarity, the two terms mentioned above are avoided [4, 5].

Tefft et al first described this tumor in children with paravertebral soft tissue masses. Symptoms are non-specific and typically depend on the location of the tumor. EES are most commonly diagnosed via Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). The final diagnosis of EES requires a comprehensive analysis on the histopathology, immunoprofile and interphase Fluorescence In Situ Hybridization (FISH) results. In this study, we present a case of EES found in the right inguinal region which invade one of the superficial inguinal LN.

Case Report

A 28-year-old man presented with a 2- month

History of a painless swelling of the right inguinal region initially started as a small discrete painless lump, enlarged gradually and occupies the right inguinal region. He also noticed another hard painful lump developing below the right inguinal lump over 1 month prior. Physical examination, there is single lump, oval in shape, hard, 6 cmx7 cmx13 cm , in right inguinal region, attached to skin with ulcer on it not attached to underlying structure, well defined, not tender associated with tender superficial inguinal LN on right side (Figure 1).

Investigation

Ultrasonographic evaluation revealed a large hypoechoic and inhomoge neous well defined soft tissue mass, developed in the right side of inguinal region measuring about 13 cmx6 cm. The ultrasonography of the abdomino-pelvic cavity showed no anoma. MRI shows no abdominal or pelvic masses, no evidence of hepatic masses, ascites, or lymphadenopathy, no related soft tissue lesions noted at the pelvic cavity or sciatic notches, no evidence of bony lesions. The patient underwent an Excisional biopsy with 1cm margin and inguinal lymph node sampling (Figure 2a & 2b) and the wound closed in simple suture on edges and skin graft in center portion (Figure 3). titlIn outpatient follow up patient looks well, wound clean and healed in a possible good cosmetic way (Figure 4).

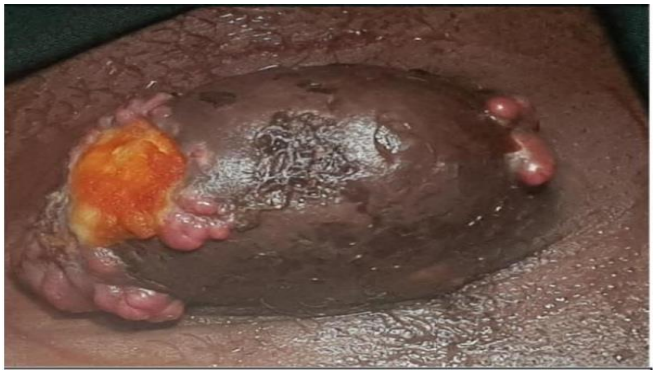

Figure 1. The right inguinal lump.

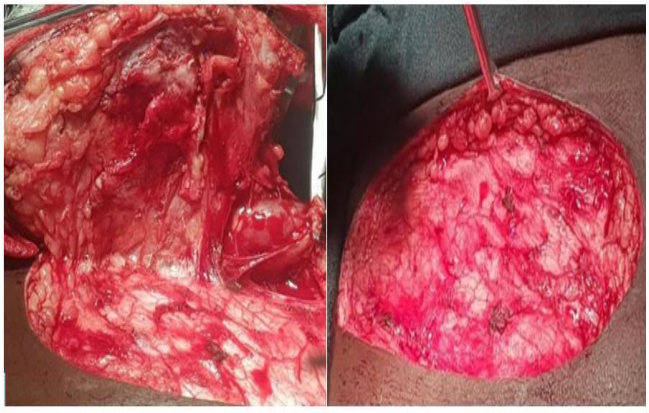

Figure 2.(a) Excisional biopsy (Intraofperative), **2(b)** Excisional biopsy (Intraoperative).

Figure 3. Skin Graft (60% taken).

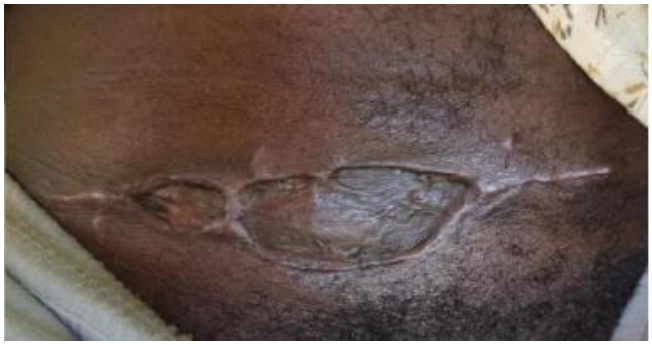

Figure 4. Outpatient follow up.

Pathological Finding

Dome shaped soft tissue covered by skin measuring 13 cmx6 cmx7 cm, measuring 6cm in greater diameter with skin ulcer, the tumor about 1.2.0.9 cm from all peripheral margins. A microscopy of groin right mass show deep fascia tumor consists of lobule of monotonous with abundant clear cytoplasm, having prominent nuclei with frequent mitosis. In a back ground of fibro-collengous stroma. The tumor extended from deep septal fascia up to dermis & with ulceration. The tumor invades one lymph node. The peripheral margins are free about 1 cm-0.9 cm away from tumor. The deep surgical margin is about 0.4 cm away from tumor. The feature is that morphological favor of extra-skeletal Ewing sarcoma. Microscopy of lymph node excision shows three lymph nodes free from neoplasm (0/3).

Discussion

Ewing sarcoma commonly arises from bone and it can involve soft tissues at the time of the diagnosis. Rarely, Ewing sarcoma may have an extra-skeletal origin. Soft tissue Ewing sarcoma is a rapidly growing, round-cell, malignant tumor which can reach 10 cm by the time of the diagnosis [6]. Young adolescents and adults between the ages of 10 and 30 years are predominantly affected with a slight predominance in males [7, 8]. However, some cases have been reported in patients between 14 months and 77 years of age [2]. Commonly affected extra-skeletal sites are the paravertebral spaces, lower extremities, head and neck, and pelvis [7-13]. Other rare reported locations of extraskeletal Ewing sarcoma are various and include the retro-peritoneum, orbit, skin, and chest wall [14-17]. To the best of our knowledge, only one previous paper reported a Ewing sarcoma involving the abdominal wall [18]. The most frequent presenting symptom is a rapidly growing mass with local pain.

The imaging features soft tissue Ewing sarcoma are non-specific [8, 12, 13, 15] It often presents as a well-limited mass which should not be confused with a benign lesion. Ultrasonography often shows a hypoechoic and heterogeneous mass. Hypodense foci are frequent in the large mass and are due to intratumor necrosis [8, 9, 13]. Spontaneous tumor haemorrhage and adjacent bone destruction have been reported in some cases [8]. The secondary osseous involvement is rare even when the mass is located near a bone segment. It can result in cortical erosion and/or a periosteal reaction [7]. Extra-skeletal Ewing sarcoma is confirmed by characteristic features on histologic analysis, histochemistry, immunohistochemistry and electron microscopy. Differential diagnoses include other Small Blue Round Cell Tumors (SBRCTs) and other members of the Ewing family of tumors such as the Primitive Neuroectodermal Tumor (PNET) [16].

The mainstay treatment should include multiagent chemotherapy and aggressive surgical treatment. Tumors that are not appropriate for surgical resection or have positive margins are treated with radiation [6, 9, 18] The prognosis for extra-skeletal Ewing sarcoma appears more favorable than that of Ewing sarcoma in bone [16]. Two previously published series devoted to the study of extra-skeletal Ewing sarcoma stated that it is a curable disease and has the best prognosis in young patients (age<16 years) treated with complete resection with wide surgical margins in conjunction with chemotherapy [7]. Ahmad et al. also demonstrated that tumor size did not have a significant effect on overall 5-year survival or disease-free survival [7]. There were no difference in overall diseased free survival between the patients and metastatic disease and those without diseases [7]. Lung, liver, brain and bone are the most common metastatic sites [8]. Imaging is useful for evaluation of the rate of resect ability and the tumor response to treatment [9]. Tumors that are not appropriate for surgical resection or have positive margins are treated with radiotherapy.

Conclusion

In summary, Ewing sarcoma arising from soft tissues is extremely rare. Although its radiological features are nonspecific, it should be kept in mind in the differential diagnosis of soft tissue tumors in young people.

Ethical statement

Written informed consent was provided by the patients to have their case details and images published.

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