



## Case report

## A large femoral primary cutaneous Ewing's sarcoma in a 35-year-old woman: Case report

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## ABSTRACT

**Introduction:** Extraskeletal Ewing's sarcoma is a rare malignant tumor of mesenchymal origin, which is histologically similar to primary osseous Ewing's sarcoma. It has been well described in deep soft tissues. However, location in cutaneous or subcutaneous tissue has rarely been reported. Being seen principally in children, it can be seen, rarely, in old men.

**Case presentation:** We present a case of large primary cutaneous Ewing sarcoma within the left thigh of a 35-year-old woman, without osseous involvement. Physical examination. Histologically, it was a small round cell tumor that marked strongly for CD99. The diagnosis of cutaneous Ewing sarcoma was performed.

**Discussion:** The things that distinguish our case are that it is the first case in Syria, in addition to the size of the large tumor with diameters of 15 × 20 cm, and it is in a 35-year-old woman.

**Conclusion:** Ewing sarcoma is a rare malignant small round cell tumor of the skin and subcutaneous tissue. It should be differentiated from other cutaneous neoplasms composed of small round cells.

## 1. Introduction

Ewing's sarcoma (ES) is a small round cell tumor, usually arising from flat bones and diaphyseal region of long bones. It is commonly found in the first two decades of life. It is curable when diagnosed in the localized stage and requires multimodality treatment. The extrasosseous Ewing sarcoma is a neuroectodermal tumor of clear cells and very rare, with few cases reported in the literature [1,2]. Primary cutaneous Ewing sarcoma is a very rare entity with less than 100 cases reported in the literature. The clinical picture is usually composed of a superficial tumoral mass of 2-3 cm, of soft consistency, mobile, sometimes painful. The tumor is located in the middle dermis, deep dermis or superficial subcutaneous tissue and may involve the dermal papilla, with pedunculated tumoral presentation. The average evolution time to complement the diagnosis is 5 months. The more frequently affected locations are the upper and lower extremities, trunk, head and neck or even multiple lesions [3,4]. The histology and molecular biology allowed us to perform the diagnosis of cutaneous Ewing's sarcoma. The treatment is surgical resection, being associated or not with chemotherapy and/or

radiotherapy, depending on the size and location of the tumor. Our patient came with a complaint of a large, painless tumor without any accompanying symptoms. The immunostaining of the specimens after surgical removal confirmed the presence of primary cutaneous Ewing sarcoma.

This case report has been reported in line with the SCARE criteria [5].

## 2. Case presentation

A 35-years-old woman visited the general surgery Department at Ibn Al Nafees Hospital with a complaint of a gradual mass growing two years ago, above the posterior aspect of the left thigh. She did not follow up on the condition of the mass during the two years at the surgeons due to the outbreak of the Covid\_19 epidemic. Clinical examination of the mass revealed that it is fixed on the skin and moving in depth without any other accompanying symptoms. BMI was 36 (sever obesity). In the patient's history, there were two Partus caesarius 14 years ago and allergic asthma treated with theophylline and salbutamol, and the patient was

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smoker with an average of 5 packs/year, this means a pack per day for 5 years. All tests of complete blood count, electrolytes, liver and kidney enzymes were within normal range. By examining the respiratory system, the chest x-ray was normal, but on chest auscultation bilateral wheezing were heard because the patient has asthma, so there was a high risk of performing general anesthesia. We took multiple core biopsies of the tumor, the longest one measuring 1.5 cm in order to send the core fragments for pathological autopsy, and the result was infiltration by tumor of predominant plasmacytic-like cells. Depending on above the primary diagnosis was extrasosseous plasmacytoma. Open surgery was the best option to remove the tumor, 5 general surgeons, a nurse and an anesthetist participated in the surgery. We performed lumbar anesthesia and the operation lasted an hour and a half in 45 degree position. Where a spindle incision was made around the mass, and the mass was dissected within the tissues around it with free safety edges (Fig. 1), and it was removed with a large size of 20 × 15 cm (Fig. 2), and a two-slit drainage tube was installed. Paracetamol and ceftaxime were administered intravenously after the operation.

Histological examination revealed a tan cut surface lobulated and contains brown hemorrhagic areas grossly and microscopic examination shows sheets and lobules of malignant cells with hemorrhage, necrosis, hemosiderin deposition and areas of fat necrosis (Fig. 3). The tumor showed only focal cytoplasmic globular periodic acid Schiff (PAS) and was immunohistochemically positive for CD99 in a characteristic membrane pattern (Fig. 4). The patient was discharged after 3 days. The drain tube was removed in the fifth day with a serous ooze of 500 ml in the previous days. A postoperative whole-body computed tomography (CT) scan revealed no metastases (Fig. 5). The patient did not receive any adjuvant chemotherapy or radiotherapy according to oncologist consultation and after year of follow-up period there was no evidence of recurrence but after one week delayed wound healing were suspected and a wound biopsy was taken to rule out malignancy (Fig. 6). Thus the delayed wound healing was attributed to patient sever obesity, as no signs of infection were observed also. The pathology report came out with no evidence of malignancy as normal supportive inflammation with granulation tissue was detected. After treatment she was followed every three months through clinical examination, while magnetic resonance imaging (MRI) was performed every 6 months during the first year. The time of following up was 2 years.

### 3. Discussion

Primary cutaneous Ewing sarcoma is a rare clinical presentation of

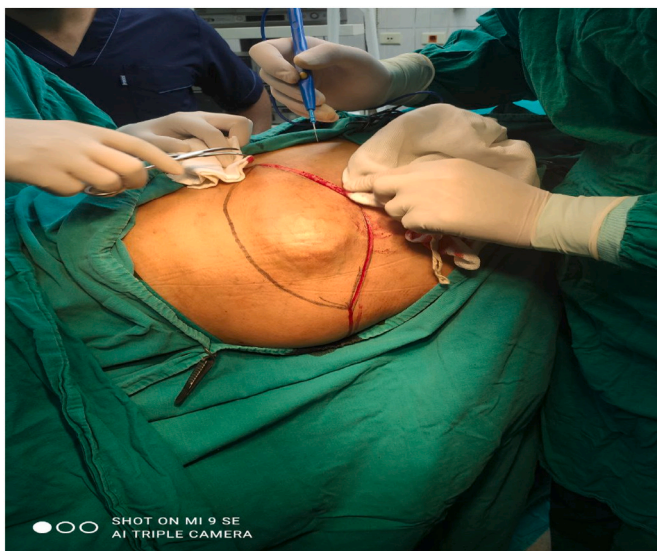


Fig. 1. The picture during operation that shows the large tumor.

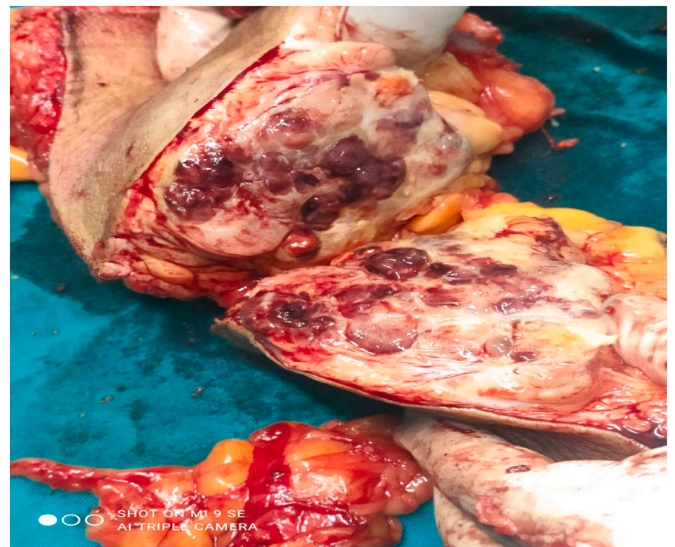


Fig. 2. The tumor after surgical removing.

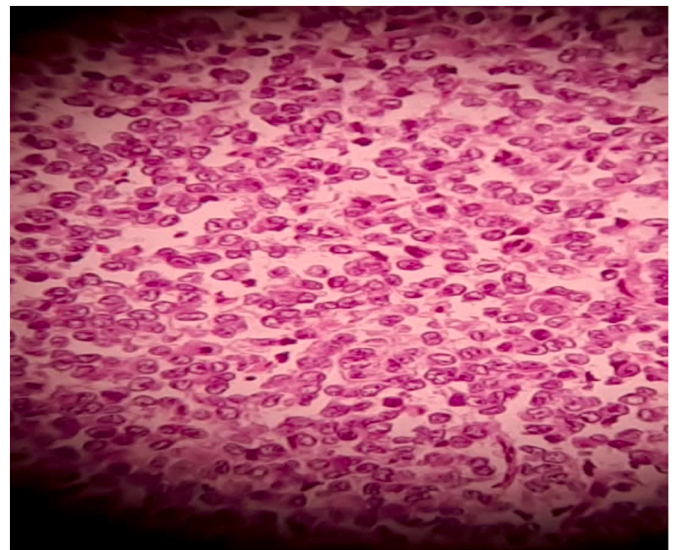


Fig. 3. Glass slide stained with HE, 400× magnification, showing diffuse proliferation of cells with indistinct cytoplasm and small basophilic nuclei.

Ewing sarcoma, usually occurring as a small, localized tumor on the extremities of young adults and associated with favorable prognosis [6,7]. In contrast to our case, which came in a very large size. Few case reports exist, with only 78 cases reported in the literature since it was first described in 1969 [8]. Patients were generally older (17 years old) and were female. Due to its rarity and morphological similarity to other cutaneous tumors, cutaneous ESs are subject to being clinically and pathologically subdiagnosed [9]. Clinically, lesions were described as slow growing, soft and hemorrhagic in appearance. Histologically, Ewing sarcoma is composed of small round cells, which express CD99. While sensitive, CD99 is not a specific marker and can be attributed to other disease processes that must be ruled out, such as Merkel cell carcinoma and lymphoma [10]. Depending on cases previously reported in the medical literature, the best treatment option is wide surgical excision with free edges. To our knowledge the present case is the largest primary cutaneous Ewing sarcoma of the thigh. Despite its large size, the patient did not report any metastases and these findings support the data already, in addition to being the first rare case diagnosed in Syria. We present a 35 year-old woman presented with a femoral lesion that had

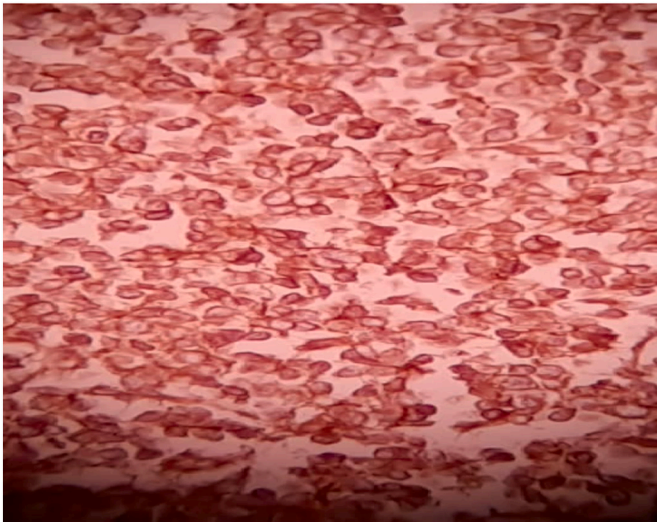


Fig. 4. Immunohistochemistry strongly positive for CD99.

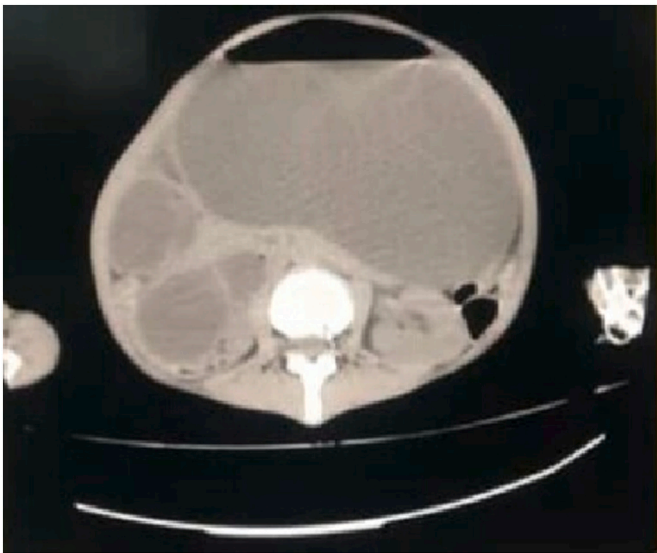


Fig. 5. computed tomography (CT) scan revealed no metastases.

been slowly developing for two year. The 20-cm lesion was removed and histological examination showed proliferation of small round cells in the dermis. Immunostaining revealed cytoplasmic membrane expression of CD99. A staging examination revealed no other abnormalities. It was decided to treat the lesion as for extraosseous Ewing's sarcoma with wide resection, accompanied by the follow-up of the patient's condition on the computed tomography and magnetic resonance imaging to investigate the relapse, fortunately we followed the patient for a whole year and we did not notice any relapse.

#### 4. Conclusion

Although many cases have been reported primary cutaneous Ewing's sarcoma in the medical literature but they remain very rare and our case is the first case in Syria, and what distinguishes it is the very large tumor size. A prompt clinical, histological, immunohistochemical and cytogenetic diagnosis and a proper treatment remain fundamental for their correct management.



Fig. 6. The delayed wound healing because of severe obesity.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

#### Consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Ethical approval

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#### CRediT authorship contribution statement

Weaam Ezzdean: contributed in study concept and design, data collection, and writing the paper.

Sarya Swed: contributed in writing the paper and data collection.

Mohammad Badr Almoshantaf: contributed in writing the paper.

Bisher Sawaf: contributed in reviewing the paper.

Talal Orabi: contributed in reviewing the paper.

#### Declaration of competing interest

All authors declare no conflict of interest.

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