Extraskeletal Ewing Sarcoma- A Case Report with Review of Literature

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(Ann Coll Med Mosul 2025; 47 (1):130-133).
Received: 7th Aug. 2024; Accepted: 10th Nov. 2024.

ABSTRACT

Extraskeletal Ewing sarcoma is a rare malignant soft tissue tumor. It belongs to a group of small round blue cell tumors known as the Ewing sarcoma family of tumors, with common histological and genetic features. Mainly encountered in adolescents and young adults, it commonly develops in the extremities (36%) of patients and, in others, arises in central locations (commonly paravertebral regions).

The Presnting case of a 13-year-old female presented to the private clinic after a history of trivial trauma to the back; her family noticed amass in the left paravertebral area, and the mass was painless. During the physical examination, the mass measured about 5*5 cm. It was not tender, hard inconsistency and fixed to the ribs, and the skin was not tethering; a radiological examination showed a mass of about 8*10*12 cm. involving three intercostal spaces and their ribs bulging into the thoracic cavity. A tru cut biopsy and histopathological examination revealed round cell tumors (Extraskeletal Ewing sarcoma) confirmed by immunohistochemical marker CD99 strongly positive. The patient was treated by chemotherapy and surgery, and she responded well to treatment. After eight cycles of chemotherapy, the patient was referred to the surgeon for resection of the residual mass; a left posterolateral thoracotomy was done, and the remnant of the mass was mainly fibrous tissue removed with 10 cm. of the posterior part of the 8th rib removed and sent again for histopathological examination, then the patient sent to an oncologist for prophylactic chemotherapy the patient followed for 5 months with no evidence of recurrence

keywords: Ewing sarcoma, extraskeletal Ewing sarcoma, round blue cell tumor.

ساركوما إيوينغ خارج الهيكل العظمي ـ تقرير حالة مع مراجعة الأدبيات

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الخلاصة

ساركوما إيوينغ خارج الهيكل العظمي هي ورم خبيث نادر في الأنسجة الرخوة ، وهي تنتمي إلى مجموعة من أورام الخلايا الزرقاء الصغيرة المستديرة المعروفة باسم عائلة أورام ساركوما إيوينغ ، ولها سمات نسيجية ووراثية مشتركة. تصادف بشكل رئيسي في المراهقين والشباب ، وعادة تصيب الأطراف بنسبة (٣٦ ٪) من المرضى وفي الآخرين تنشأ في المواقع المركزية (عادة المناطق شبه الفقرية) .

الحالة المقدمة لأنثى تبلغ من العمر ١٣ عاما قدمت إلى العيادة الخاصة بعد فترة من تعرضها لضربة بسيطة في الظهر ، لاحظت عائلتها أن الكتلة كانت غير مؤلمة في المنطقة الفقرية اليسرى. أثناء الفحص ، كانت الكتلة تقيس حوالي ٥ * ٥ سم ، ولم تكن طرية ،مثبتة على الأضلاع ولم يكن الجلد مرتبطا بها، وأظهر الفحص الإشعاعي كتلة حوالي ٨ * ١٠ * ١٢ سم تتضمن ثلاث مساحات وأضلاعهم منتفخة في التجويف الصدري. تم أخذ خزعة مقطوعة وكشف الفحص النسيجي المرضي عن أورام الخلايا المستديرة (ساركوما إيوينغ خارج الهيكل العظمي) والتي أكدتها العلامة المناعية الكيميائية CD99 التي كانت إيجابية بقوة. تم علاج المريضة بالعلاج الكيميائي والجراحة واستجابت بشكل جيد للعلاج. بعد ثماني جلسات من العلاج الكيميائي ، أحيلت المريضة إلى الجراح لاستئصال الكتلة المتبقية ، تم اجراء بضع الصدر الخلفي الجانبي الأيسر ، وكانت بقايا الكتلة عبارة عن نسيج ليفي تمت إزالته بشكل أساسي مع إزالة ١٠ سم من الجزء الخلفي من الضلع الثامن وإرساله مرة أخرى للفحص النسيجي المرضي ، ثم أرسلت المريضة إلى طبيب الأورام للعلاج الكيميائي الوقائي الذي تابعه المريض لمدة ٥ أشهر دون أي دليل على تكراره.

الكلمات المفتاحية : ساركوما إيوينغ، ساركوما إيوينغ خارج الهيكل العظمي، ورم الخلايا الزرقاء المستدير.

INTRODUCTION

Extraskeletal Ewing sarcoma (ESES) was first discovered in 1969, and it is a rare tumor forming about 20-30% of all reported cases of Ewing sarcoma; it belongs to Ewing sarcoma (ES) family of tumors [1], which is a group of small round blue cell tumors that have a common neural histological picture and genetic mechanism [2].

The Ewing Sarcoma Family of Tumor includes the classical bone Ewing sarcoma, Extraskeletal Ewing sarcoma, peripheral primitive neuroectodermal tumor, and Askin tumor of the chest wall [3]. Ewing sarcoma forms 3% of all pediatric malignancies, and so it is considered the second most common bone cancer [3]; by development in the Ewing sarcoma treatment with both local surgery and adjuvant chemotherapy, the 5-year survival rate raised from less than 20% to above 70% [4].

Case Presentation

A 13-year-old female presented to the private clinic after a history of trivial trauma to the back. Her family noticed a mass in the left paravertebral area, and the mass was painless. During the physical examination, the mass measured about 5×5 cm. It was not tender, hard in consistency, and fixed to the ribs, and there was no skin tethering; the radiological examination done, including a CT scan (figure 1), showed a mass of about 8×10×12 cm involving three intercostal spaces and their ribs bulging into the thoracic cavity.

A true-cut biopsy was taken, and a histopathological examination was done for multiple small pieces of soft tissues measuring 1.5 cm. All microscopically revealed fibrofatty tissue infiltrated by a nest of small, round blue cells separated by septa. This picture suggests extraskeletal ewing sarcoma (figure 2), and the diagnosis is confirmed by the immunohistochemical marker CD99, which was strongly positive with diffuse membranous expression (figure 3). At the same time, CD45, myogenin, and synaptophysin were negative.

The patient was treated with chemotherapy, and surgery was done, and she responded well to treatment. After eight cycles of chemotherapy, the patient was referred to the thoracic surgeon for resection of the residual mass; a left posterolateral thoracotomy was done, and the remnant of the mass was mainly fibrous tissue removed with 10 cm. of the posterior part of the 8th rib removed and sent again for histopathological examination where multiple fragments of soft and bony tissues examine. All revealed fragments of skeletal muscle fibers, fibrous tissue, and mature fat cells with extensive hyaline degenerative changes, infiltrated by only a few scattered small nests of small round blue cells that represent few remnants of Ewing sarcoma with marked post-chemotherapy effects (figure 4) then the patient sent to an oncologist for prophylactic chemotherapy the patient followed for 5 months with no evidence of recurrence.

DISCUSSION

Ewing sarcoma, in general, is a rare tumor and extraskeletal Ewing sarcoma is a very rare cancer; it has an incidence of 0.4 per million (ten times less common than osseous Ewing sarcoma) [5]. It mainly affects young individuals (10 - 30 years) without gender predominance [6,7]. Extraskeletal Ewing sarcoma can arise from different locations, including the lower limbs, pelvis, paravertebral region, and chest wall [1]. This case is EES in the paravertebral area of a 13-year-old female. Extraskeletal Ewing sarcoma clinical symptoms depend on its site; usually, it causes local pain caused by rapid expansion as well as to the site of metastases, which are found in 25% and mainly affect the lung [8]. For the case being discussed, the patient was asymptomatic and discovered incidentally after a history of minor trauma without evidence of metastasis. The imaging characteristics of EES are non-specific, and the imaging test of choice for assessing the tumor's size, site, and extension of the mass and assessing if the tumor has metastasis to the lungs is a CT scan or not [1]. In this case, a CT scan showed a mass of about 8×10×12 cm^3 involving three intercostal spaces and their ribs bulging into the thoracic cavity. The diagnosis is confirmed by histopathological and immunohistochemical analysis, which confirms the diagnosis of ESES; the presence of the t(11;22) chromosomal translocation also helps diagnose. However, in the presenting case, it was not performed for economic cause.

The Treatment for EES includes chemotherapy and surgical resection or, less commonly, adding radiotherapy, also helps in confirming the diagnosis by sending biopsy to the histopathologist and provides a long-term cure rate with decreased incidence of recurrence [1]. lateral thoracotomy is the approach of choice when the lesion affects the hemithorax [9]. In this case, complete surgical excision was done through left posterolateral thoracotomy; the remnant of the mass was mainly fibrous tissue removed with 10 cm. of the posterior part of the 8th rib.

Imaging tests for detecting metastatic tumors include CT scans of the chest, which are more favorable than X-rays in detecting lung metastasis; metastatic lumps are usually well-circumscribed, oval, or round in shape and located peripherally in the lung [10]. CT scan of the chest for the presenting case revealed no lung metastasis.

The prognosis of EES is better than that of the skeletal one [7,11]. The 5-year survival rate for focal EES is superior to that of the focal skeletal subtype [8]; the worse prognosis in EES is related to tumor size, which is considered a strong prognostic marker in focal cancer, older age, and pelvic involvement [1]. However, histological response for patients treated with neoadjuvant chemotherapy is considered the strongest prognostic factor [1]. Metastatic disease is a bad prognostic factor, with a 5-year survival rate of less than 30% [12]. Recurrence of the malignancy is almost always fatal [1]. This case histologically responded to chemotherapy as it showed fibrosis, and the tumor had no feature of metastasis radiologically with no history of metastasis or recurrence at follow-up.

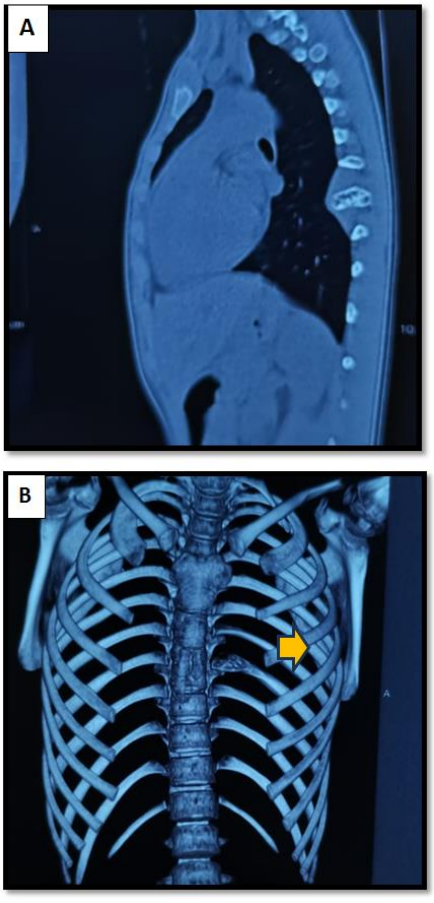

Figure 1: CT scan showing a mass of about 8×10×12 cm involving three intercostal spaces and their ribs bulging into the thoracic cavity (A: lateral view, B: anterior view)

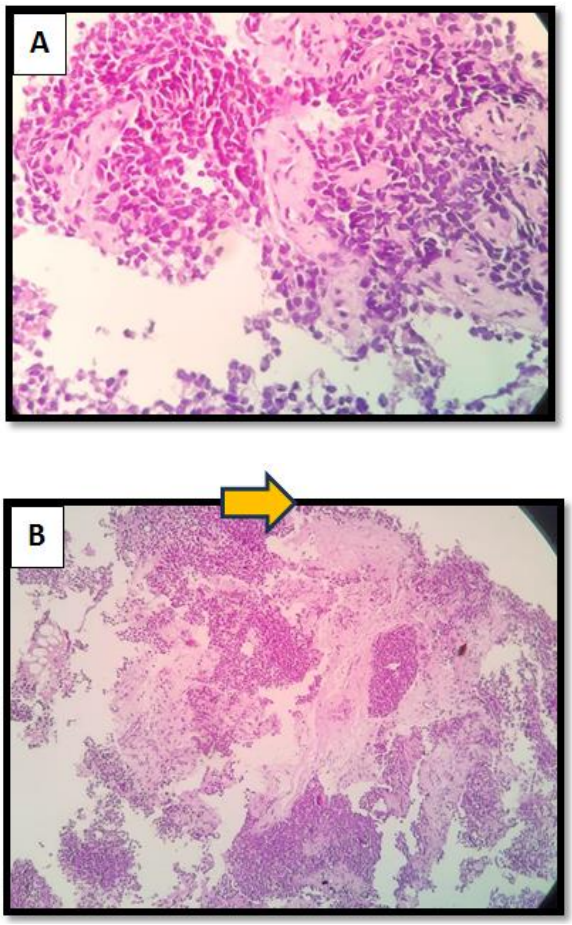

Figure 2: fibrofatty tissue infiltrated by a nest of small, round blue cells separated by septa (A: X10, B: X40).

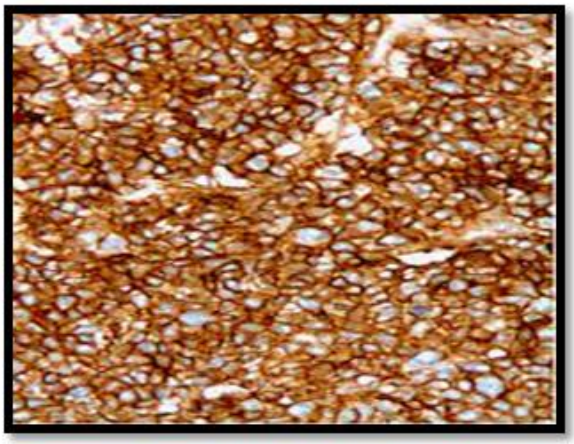

Figure (3): immunohistochemical marker CD99 strongly positive with diffuse membranous expression.

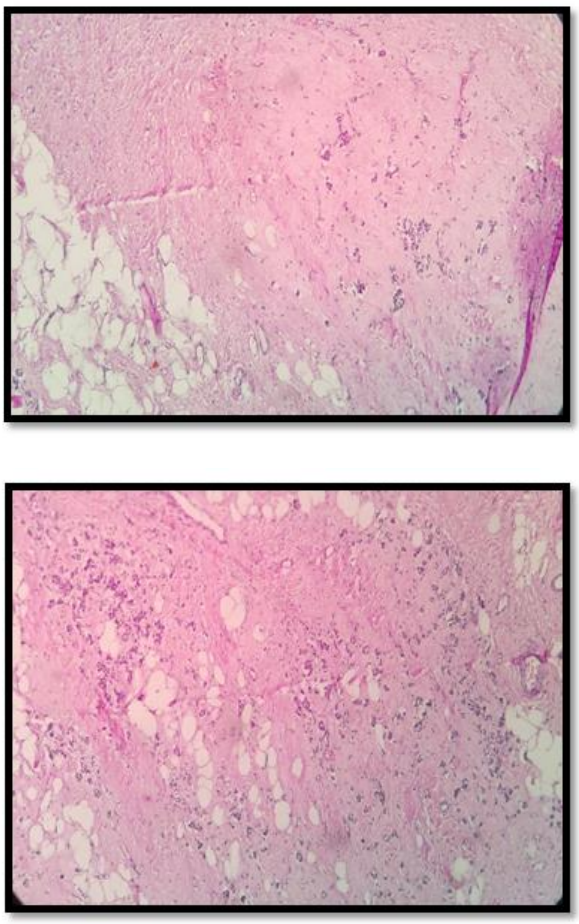

Figure 4: fragments of skeletal muscle fibers, fibrous tissue, and mature fat cells with extensive hyaline degenerative changes, infiltrated by only a few scattered small nests of small round blue round cells representing few remnants of Ewing sarcoma due to chemotherapy effects.

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