

Primary Cutaneous Ewing Sarcoma: Case Report and Review of Literature

Jlalia Z^{1*}, Sboui I², Bouhajja L², Abid L², Smida M², Klibi F² and Daghfous MS²

¹Department of Anatomopathology, Kassab institute of orthopedic surgery, Ksar said, Tunisia

²Department of Kassab institute, University of Tunis, El Manar, Ksar said, Tunisia

*Corresponding authors: Jlalia Z, Department of Orthopedic and traumatology, Kassab institute of orthopedic surgery, Ksar said 2010 Tunis, Tunisia, Tel: +00 216 95562337; E-mail: sboui_imedk@yahoo.fr

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Abstract

Ewing sarcoma (ES) is a primitive neuroectodermal tumor. It's usually a primary bone tumor but rarely occurs in the skin and subcutaneous tissues. Current literature reports only a few isolated cases or small series. To date, less than 100 cases have been reported. The diagnosis is made by aspiration cytology, histochemical stains, immunohistochemistry, electron microscopy, cytogenetics and molecular genetics of translocations. Cutaneous ES has better prognosis than primary bone or soft tissue ES with a survival rate of 91% in 10 years. The presence of metastasis is really rare. Currently, no specific recommendations to primitively cutaneous Ewing tumors, these latter are treated as bone Ewing's sarcomas: neoadjuvant chemotherapy, surgery, adjuvant chemotherapy (+/- radiotherapy), and autologous bone marrow transplantation in high risk patients. We report a new case in a 20-year-old female with a lesion in the left elbow.

Keywords: Ewing Sarcoma; Skin; Chemotherapy; Radiotherapy

Introduction

Ewing sarcoma (ES) is a primitive neuroectodermal tumor. It's usually a primary bone tumor but rarely occurs in the skin and subcutaneous tissues [1], and generally appears as a single small lesion, circumscribed mid to deep dermis or involving subcutis. Due to their rarity and morphological similarity to other cutaneous tumors, cutaneous Ewing sarcoma (CES) are subject to being clinically and pathologically sub-diagnosed [1]. We report a new case of a 20-year-old female with a lesion in the left elbow.

Case report

A 20-year-old female, with no past medical history, presented with a progressively increasing lump, of the external face of the left elbow for 7 months. He was asymptomatic with no fever or weight loss. On physical examination, the mass was superficial, firm, oval shaped, mobile, painless, and measuring 2 cm in greatest diameter. There was no lymphadenopathy detected. The standard radiographs of the elbow was normal (Figure 1). The magnetic Resonance Imaging (MRI) revealed a susaponevrotic lesion, invading the subcutaneous tissue of the external face of the left elbow (Figure 2). A surgical biopsy was done. Histological examination showed a subcutaneous (hypodermic and dermal) sarcomatous nodular proliferation (Figure 3) It is composed of monomorphic small round blue cells, with scanty cytoplasm, and round or ovoid hyperchromatic nuclei, mitotic Figures were rare (Figure 4). The epidermis was normal. The immunohistochemical study revealed a strong and diffuse membranous immunostaining for CD 99, and lack of stain for lymphoid markers (CD3, CD 20) and muscular markers (desmin and myogenin) (Figure 5). The molecular study showed a chromosomal translocation t (11; 22). These features confirmed the diagnosis of extraskeletal Ewing's sarcoma. Thoracic CT was normal. The patient received 6 courses of chemotherapy (Ewing 99 protocol). After

chemotherapy, MRI was done and revealed a stable lesion. She underwent a surgical excision with negative margins. The skin fragment measured 6.5 × 4 × 1.5 cm. On the cut, the tumor was white-grayish and measured 2 × 2 × 0.8 cm of greatest diameter. Histologically, tumour remnants corresponded to subcutaneous Ewing's sarcoma. Afterwards, she received an adjuvant chemotherapy. At 2 years of follow-up, there was no recurrence and no metastasis.

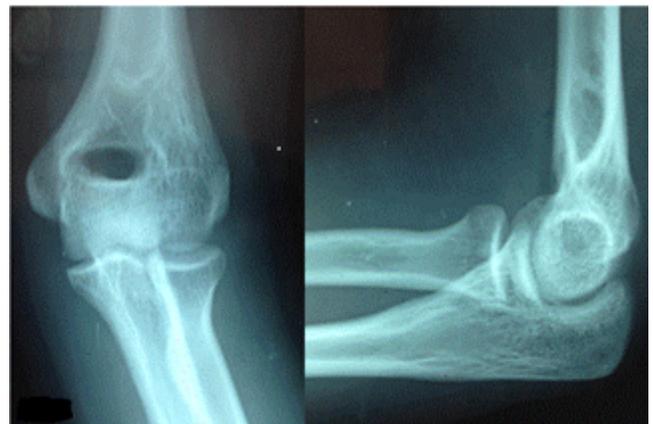


Figure 1: The standard radiographs of the elbow. No abnormalities.

Discussion

Current literature reports only a few isolated cases or small series. To date, less than 100 cases have been reported [2]. Extraskeletal Ewing's sarcoma family of tumors (EESFT) most frequently occur in the deep soft tissues of children and young adults, such as paraspinal muscles, chest wall, and the lower extremities [2]. Nevertheless, it occasionally presents in a superficial location either as a primary

tumor or a metastasis from osseous or deep seated EESFT. Those superficially located lesions, the so called Primary cutaneous EESFT are exceedingly rare and they are limited to the skin and generally present as a single small lesion, circumscribed mid to deep dermis or involving superficial subcutis They were first described by Angerwall and Enzinger in 1975.

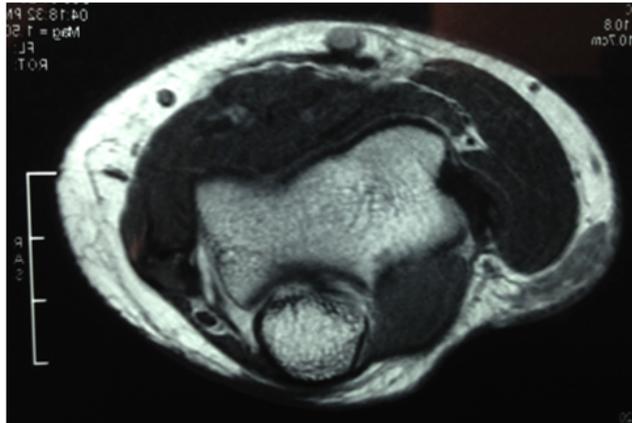


Figure 2: MRI. Sus-aponevrotic lesion, invading the subcutaneous tissue of the external face of the left elbow.

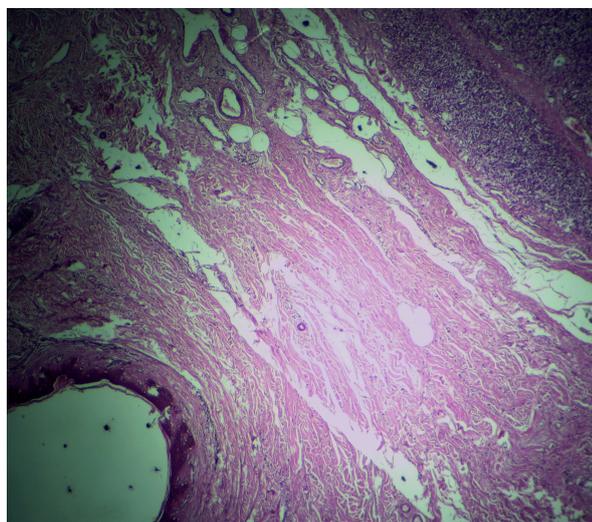


Figure 3: Hypodermal monomorphous round blue cell proliferation. (HEX100).

The diagnosis is made by aspiration cytology, histochemical stains, immunohistochemistry, electron microscopy, cytogenetics and molecular genetics of translocations [1,3,4]. The differential diagnosis of ES include primary cutaneous small round cell tumors like Merkel cell carcinoma, eccrine spiradenoma, lymphoma, melanoma, clear cell sarcoma, rhabdomyosarcoma, malignant rhabdoïd tumor, malignant primitive neuroectodermal tumor and poorly differentiated adnexal tumors [5,6]. It includes also cutaneous metastases from osseous ES, large cell neuroendocrine carcinoma, small cell lung carcinoma and neuroblastoma [1,7]. The ES is composed of small round cells which

express the CD99 and molecular study shows a specific chromosomal translocation t(11;22) involving gene EWSR1 in chromosome 22q12 or a fusion or combination between EWSR1 gene and gene of ETS family [1,3,8].

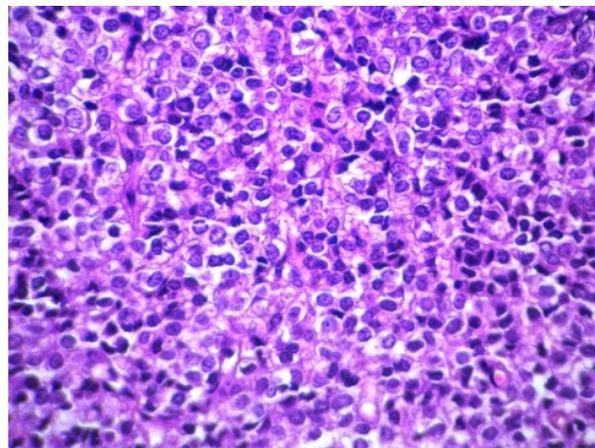


Figure 4: Small uniform cells with scanty cytoplasm and round hyperchromatic nuclei. (HE x 400).

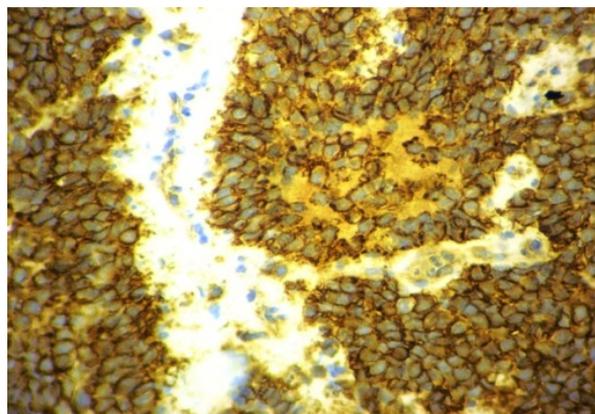


Figure 5: Immunohistochemical expression of CD99 showing characteristic reactivity of the cell membranes.

This study is made by FISH or real time polymerase chain reaction (RT-PCR) [8]. Cutaneous ES has better prognosis than primary bone or soft tissue ES with a survival rate of 91% in 10 years [1].

The less aggressive behavior is due to superficial location, small size and easy access [1,8]. One case of metastasis and one with local node involvement have been reported [9]. The presence of metastasis is really rare [4,10]. Currently, no specific recommendations to primitively cutaneous Ewing tumors, these latter are treated as bone Ewing's sarcomas: neoadjuvant chemotherapy, surgery, adjuvant chemotherapy (+/- radiotherapy), and autologous bone marrow transplantation in high risk patients [11]. This approach has been used successfully in bone tumors and deep tissue tumors, but there are data that supports the theory in the cutaneous and subcutaneous Ewing's sarcoma, the intensity should be lowered [12].

Conclusion

Cutaneous Ewing sarcoma is a very rare entity, confined to the epidermis, dermis and subcutaneous tissue. It's clinically and pathologically sub-diagnosed because of its rarity and morphological similarity to other cutaneous tumors. The molecular biology can lead us to the correct diagnosis. It seems to have better prognosis than osseous ES. The treatment consists on local surgery and chemotherapy and/or radiation.

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