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Extraskeletal Ewing sarcoma: report of a rare and exuberant case simulating lipoma

Sarcoma de Ewing extraesquelético: relato de um caso raro e exuberante simulando lipoma

Case report

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ABSTRACT
Extraskeletal Ewing sarcoma (EES) is a rare tumor that affects men between 10-20 years old and represents 25% of Ewing sarcomas. We report a case due to the previous diagnosis of a fast-growing lipoma to highlight the importance of histopathological diagnosis. A 14-year-old boy presented 10x10 cm tumor in the right shoulder for a year with ultrasonography suggesting lipoma. The lesion was excised and EES was confirmed by immunohistochemical analysis. EES diagnosis in young people is challenging compared to subcutaneous tumors, and histopathology is essential. The rapid progression of the tumor and high metastatic rates highlight the significance of early treatment.
Keywords: Sarcoma, Ewing; Neuroectodermal tumors; Sarcoma

RESUMO
Sarcoma de Ewing extraesquelético (EES) é tumor raro e agressivo, acomete mais homens entre 10-20 anos e representa 25% dos sarcomas de Ewing. Caso relatado pelo diagnóstico prévio de lipoma de crescimento acelerado, destacando a importância do diagnóstico histopatológico. Masculino, 14 anos, apresentando tumoração de 10x10cm no ombro direito há um ano, com ultrassonografia sugestiva de lipoma. Realizada exérese da lesão, sendo confirmado EES pela imuno-histoquímica. O diagnóstico de EES em jovens é desafiador frente a outras tumorações subcutâneas, sendo a histopatologia imprescindível. A rápida progressão do tumor e os elevados índices metastáticos evidenciam a importância da terapêutica precoce.
Palavras-chave: *Sarcoma de Ewing; Tumores neuroectodérmicos primitivos; Sarcoma*

INTRODUCTION

Ewing sarcoma is the second most common malignant bone neoplasm in the pediatric population after osteosarcoma. It occurs mainly in adolescents in the second decade of life and has high metastasis rates.[1] James Ewing described it in 1921, and it constitutes 10-15% of all bone sarcomas, including classic Ewing sarcoma, extraskeletal Ewing sarcoma (EES), malignant small cell tumor of the chest wall, and primitive neuroectodermal tumors of soft tissues (PNET), all with differentiation from mesenchymal stem cells.2 Its histological origin is not well elucidated, and, to date, there are no well-established associations with family history, environmental exposure, or radiation history.[1,2] Genetically, they are characterized by spontaneous chromosomal translocations, and the most specific for EES are t(11;22)(q24;q12) and t(21;22)(q24;q12).[1,2,3]

EES presents as a rare, primary soft tissue tumor, accounting for approximately 25% of the reported cases of Ewing Sarcoma.[3] It has a poor prognosis depending on the stage, and it is prevalent in men in the second and third decades of life.[4] We report the case of EES in a 14-year-old patient with a previous diagnosis of rapidly evolving lipoma to highlight the importance of early investigation, differential diagnoses, and histopathological study to achieve curative therapeutic possibilities and a better prognosis.

CASE REPORT

A 14-year-old man, eutrophic and without comorbidities, was referred to the tertiary service of Dermatology due to the appearance of a tumor in the right shoulder one year ago, with progressive growth, presenting pain, and limitation of movement. Previous ultrasonography showed a hypoechoic nodular formation measuring 4.5 x 1.4 cm, intermingled with fine fibrous strands, without significant vascular flow, located in the subcutaneous region of the right shoulder, with characteristics suggestive of lipoma. Due to the Covid-19 pandemic, the patient was seen nine months after the imaging exam, showing exuberant enlargement of the tumor, measuring 10 x 10 cm in the posterior region of the right shoulder (Figure 1). Immediate exeresis of the lesion was performed, which, during the intraoperative period, appeared as a violaceous tumor, somewhat friable and adhered to deep planes (Figure 2).

Histopathological analysis revealed a multinodular growth pattern composed of small, monotonous, round to ovoid cells, containing scarce cytoplasm and hyperchromatic nuclei organized in nests delimited by thin fibrous septa. We observed a markedly hypercellular tumor with an exuberant pseudoangiomatous pattern. Abundant mitotic figures were identified, in addition to multiple foci of necrosis (Figure 3). The immunohistochemical study showed immunoexpression for CD99 and FLI1, focal expression of cytokeratin (AE1/AE3) and S100, in addition to negativity for CD34, smooth muscle actin, desmin, myophenin, synaptophysin, EMA, and TLE1 (Figure 4). The set of findings confirmed the diagnosis of EES.

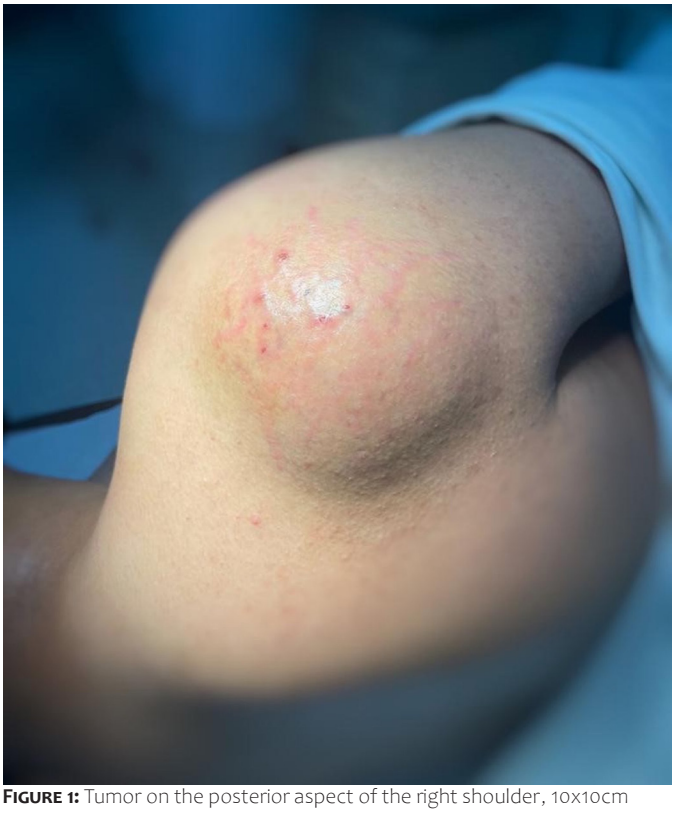

Figure 1: Tumor on the posterior aspect of the right shoulder, 10x10cm

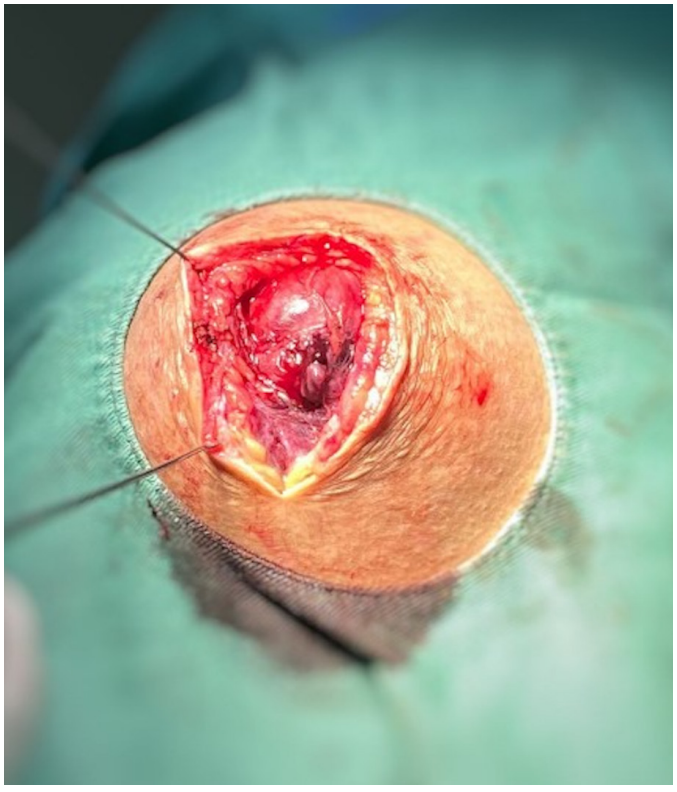

Figure 2: Intraoperative: friable lesion with violaceous coloration

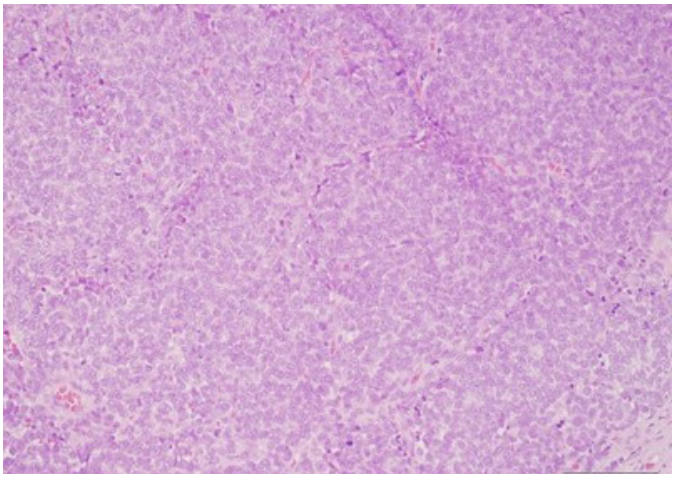

Figure 3: High-grade sarcoma: histopathological examination revealing a tumor composed of small, monotonous round cells. Hematoxylin & eosin, 200x

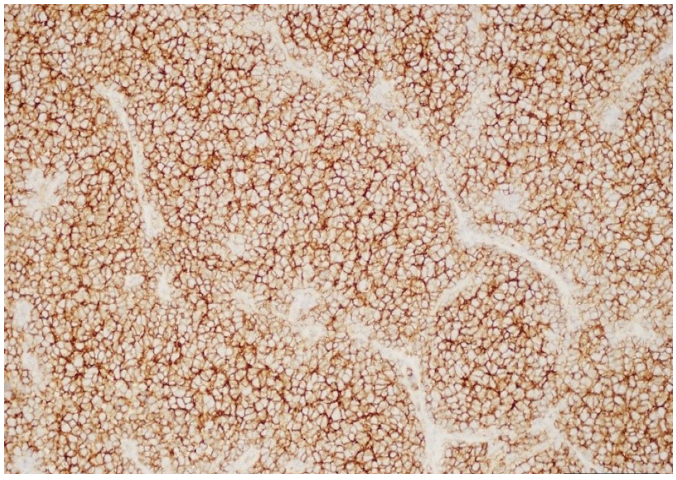

Figure 4: Extraskeletal Ewing sarcoma. Immunohistochemistry for CD99, 200x

Oncological staging revealed bone marrow metastasis. We conducted extensive surgical expansion and chemotherapy with the stability of the condition. The patient continues with 12 months of follow-up in the 13th cycle of chemotherapy and the absence of other neoplastic foci/metastases.

DISCUSSION

EES is a rare malignant neoplasm that presents as a painful, fast-growing nodule, frequently located in the paravertebral regions and lower limbs.[5] Histologically, it presents compact, homogeneous, small, round to oval cells arranged in sheets, showing in immunohistochemistry marked expression of CD99 on its surface.[2,5] It tends to local dissemination and high rates of metastasis, with prognosis dependent on tumor extension and the presence of metastases at diagnosis.[6] Thus, early diagnosis through histopathology becomes essential for the aggressive approach to the tumor and improvement of survival.[6]

Appropriate treatment involves surgery and/or local radiotherapy associated with chemotherapy.[6,7] Although EES is considered radiosensitive currently, wide surgical excision is the local therapeutic method of choice due to the adverse events associated with radiotherapy.[6,7] The complete resection of the tumor associated with early age at diagnosis is considered a predictor of better prognosis and longer survival.[6,7] On the other hand, age greater than 14 years at diagnosis, primary tumor volume greater than 200 mL, and metastases (mainly in the bone marrow and lung) are the main factors associated with a worse prognosis.[6,7] Chemotherapy with anthracyclines can be used as adjuvant therapy, and the role of local adjuvant radiotherapy after complete resection is still inconclusive, although it has been shown to improve survival. Neoadjuvant chemotherapy seems to achieve promising results, but it depends on prospective clinical trials.[8]

The diagnosis of EES in a young patient is challenging, both because of its rarity and the plethora of diseases that can manifest as subcutaneous nodules. Differential diagnoses in adolescence make the histopathological study essential, and the rapid progression of the tumor associated with high metastatic rates highlights the importance of early multimodal therapy. Despite marked improvements in survival, a better understanding of the complex biology of extraskeletal Ewing sarcoma may provide a roadmap for the successful development of biologically targeted therapies. ●

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