

# Extraskeletal Osteosarcoma. Case Report

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## ABSTRACT

Extraskeletal osteosarcoma (EOS) is a high-grade malignant tumor that accounts for less than 2% of soft tissue sarcomas and mainly engages people between the fifth and sixth decade of life. It is typically of late diagnosis, with an extensive symptom-diagnosis period greater than 6 months compared to conventional osteosarcoma. We report the case of a 43-year-old patient who presented in our service with a 1-year-old tumor localized in the gluteal and left thigh region. After inconclusive paraclinical studies and biopsy, the tumor was surgically resected with wide margins and diagnosed as EOS after the histological analysis of the piece. The patient showed a very good clinical and paraclinical progression without signs of local or distant recurrence after 20 months of follow-up.

**Keywords:** Extraskeletal osteosarcoma; soft-tissue sarcoma.

**Level of Evidence:** IV

## Osteosarcoma extraesquelético. Presentación de un caso

## RESUMEN

El osteosarcoma extraesquelético es un tumor maligno de alto grado que representa menos del 2% de los sarcomas de partes blandas y que afecta principalmente a personas entre la quinta y sexta década de la vida. Típicamente el diagnóstico es tardío, con un período síntomas-diagnóstico extenso, >6 meses en comparación con el del osteosarcoma convencional. Comunicamos el caso de un hombre de 43 años que acudió con una tumoración en la región glútea y el muslo izquierdo de un año de evolución. Luego de estudios complementarios y una biopsia no concluyentes, se procedió a la resección quirúrgica del tumor con márgenes amplios. El estudio histológico de la pieza indicó osteosarcoma extraesquelético. El paciente tuvo una evolución muy buena, sin elementos de recidiva local ni compromiso a distancia luego de 20 meses de seguimiento.

**Palabras clave:** Osteosarcoma extraesquelético; sarcoma de partes blandas.

**Nivel de Evidencia:** IV

## INTRODUCTION

Soft tissue osteosarcoma or extraskeletal osteosarcoma (ESO) is a rare, high-grade malignant tumor composed of neoplastic cells that secrete organic bone matrix that can mineralize.<sup>1</sup> It represents less than 2% of soft tissue sarcomas.<sup>2-5</sup> The objective of this article is to report the case of a 43-year-old patient with soft tissue osteosarcoma of the thigh and to analyze its clinical-pathological aspects, differential diagnoses, and treatment.

## CLINICAL CASE

The patient was a 43-year-old man with high blood pressure who had undergone a splenectomy 12 years before due to abdominal trauma. Nine months before, he had suffered trauma to the posterolateral aspect of the left thigh with a significant hematoma. Once the acute period of pain had resolved, he consulted at our Center for a tumor on the left thigh that had been growing for a year, progressive and painful. The physical examination revealed a tumor located on the posterolateral aspect of the proximal third of the left thigh, with undefined limits, approximately 10

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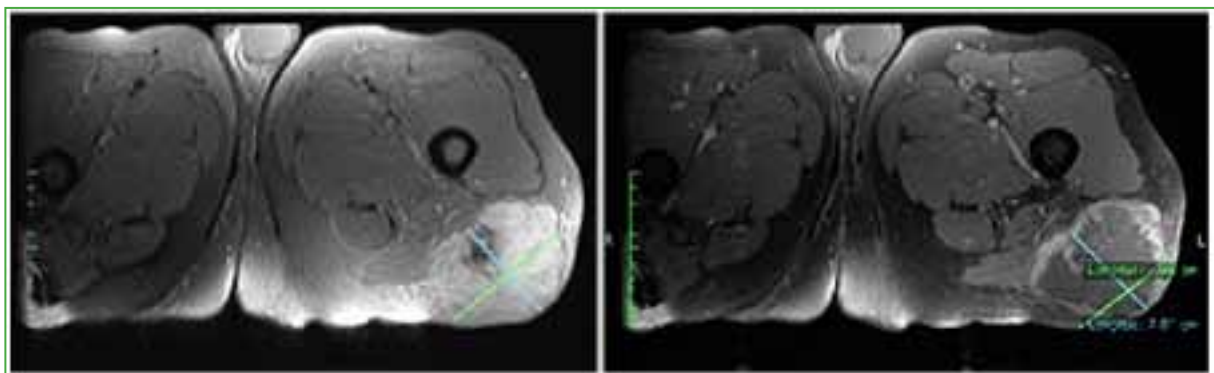
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x 15 cm in diameter, rounded, hard, attached to superficial planes and deforming said region. Erythema of the most compromised skin sector was observed. No alterations were detected in the neurovascular examination. The rest of the physical examination showed no particularities and no elements of general repercussion (Figure 1).



**Figure 1.** Patient images. On the posterolateral side of the left thigh, the tumor stands out, with little skin erythema, deforming said region.

In the MRI of the left thigh, a mass was visualized on the posterolateral side, with a gadolinium-enhancing cystic image in its center (Figure 2). The abdominopelvic MRI and body computed tomography did not show any particularities.

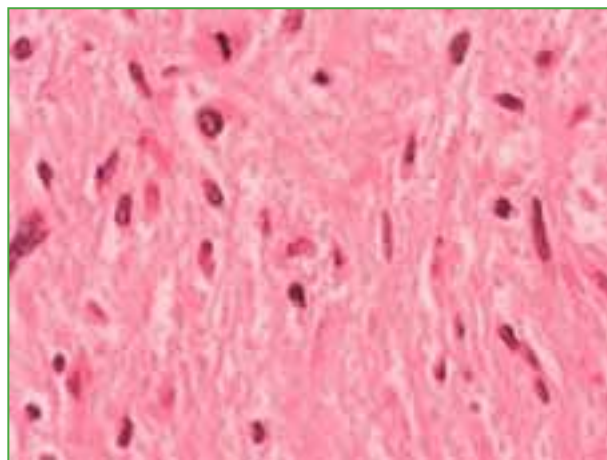


**Figure 2.** MRI of the thigh before treatment. In the axial planes in T1 and T2 sequences, a mass is visualized in the posterolateral aspect of the left thigh with a cystic image inside.

A percutaneous Tru-Cut biopsy was performed longitudinally, following the plan for definitive treatment, and eight cylinders of tissue were obtained that measured between 4 and 20 mm in length (Figure 3). The result of the histological study revealed proliferation of mesenchymal spindle cells, with moderate pleomorphism, and a presumptive diagnosis of intermediate-grade spindle cell sarcoma (Figure 4).



**Figure 3.** Tru-Cut needle biopsy. It is made longitudinal to the future incision for surgical resection, marked with a dermographic pencil.



**Figure 4.** Microscopy. The proliferation of mesenchymal spindle cells with moderate pleomorphism is observed.

Immunohistochemical staining techniques in the resected piece confirmed the diagnosis. The surgical resection of the tumor was carried out in the presence of the Plastic Surgery team, and the biopsy region was resected, with wide margins (Figure 5).

An adjuvant chemotherapy schedule with adriamycin 50 mg, days 1-3, and ifosfamide, days 1-3, was administered. The patient then received a 66 Gy radiotherapy plan.

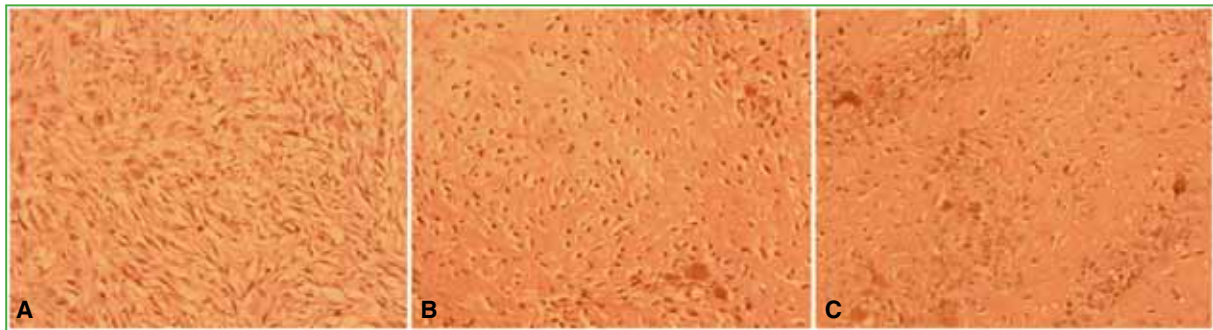
The resection piece weighed 1140 g and measured 182 x 115 x 107 mm. A radiograph showed some central calcifications (Figure 5).



**Figure 5.** A. Surgical resection with wide margins, along with the path of the sample collection sector. B. Resection piece and corresponding radiograph showing intrasubstance calcifications.

The microscopy analysis revealed diverse histological patterns, spindle cell areas with myxoid areas, chondroid differentiation, and the presence of an osteoid substance (Figure 6). Extensive areas of necrosis and hemorrhage with elevated cellularity were observed. The tumor was in contact with the dermis and deeply infiltrated the muscle bundles. The deep edge of the resected piece was 5 mm away from the tumor. ESO with areas of chondroblastic differentiation was diagnosed. According to the functional scale of the *Musculoskeletal Tumour Society*, the patient had a good evolution, without pain, with good function for activities of daily living.

At the 20-month follow-up visit, there was no evidence of local recurrence or metastatic disease.



**Figure 6.** Microscopy of the resected piece. **A.** Spindle cell pattern. **B.** Chondroid differentiation. **C.** Osteoid substance and osteoclasts. The diagnosis of extraskelatal osteosarcoma is considered.

## DISCUSSION

ESO was first described in 1941 by Wilson et al.<sup>6</sup> It is a rare mesenchymal tumor that develops from soft tissue, without continuity to the bone or periosteum, and produces osteoid material.<sup>7</sup> It represents about 1% of soft tissue sarcomas, and 4% of all osteosarcomas.<sup>8,9</sup> It presents, on average, between the fifth and sixth decades of life. Its etiology is unknown. In a retrospective series of 88 cases analyzed, only 12.5% had suffered a previous trauma, and 5.7% had undergone radiotherapy, on average, 15 years before the appearance of the tumor.<sup>9</sup> The development of OSE from myositis ossificans has also been reported.<sup>10</sup> Unlike osteosarcoma, the period between the onset of symptoms and the diagnosis of OSE is approximately 6 months, or three times longer.<sup>11</sup> Longhi et al. published a retrospective series of 266 cases of OSE with an average size of 10 cm, in which men predominated (1.5:1).<sup>12</sup> 18% had distant metastases when diagnosed. It predominates in the lower limb, especially in the thigh, and in most cases, it develops in the deep planes.<sup>3,13</sup> It also affects the retroperitoneum, the thoracic wall and, frequently, the breasts.<sup>10</sup> In radiographic images, a soft tissue mass with variable degrees of mineralization can be observed, and calcification in only 50% of cases.<sup>5,14</sup> Magnetic resonance imaging shows an intermediate to hypointense signal on T1 sequences and hyperintense on T2 sequences in unmineralized areas. Necrosis and hemorrhage foci are also frequent, and they were present in the cystic image described in our case.<sup>15,16</sup>

In the macroscopy study, a mass of 8-10 cm in diameter, grayish-white and with characteristic central calcifications is usually observed.<sup>8</sup> As for the microscopy study, all the subtypes of conventional osteosarcoma can be visualized. The osteoblastic type is the most frequent, followed by fibroblastic, chondroblastic, telangiectatic, small cell, and other differentiated subtypes, each with characteristic structures.<sup>8</sup>

Differential diagnoses are divided into benign lesions, such as myositis ossificans, which presents peripheral calcifications, and malignant lesions, such as synovial sarcoma and epithelioid sarcoma.<sup>17</sup> Undifferentiated pleomorphic sarcoma is very difficult to differentiate from ESO. Parosteal osteosarcoma can also manifest as a soft tissue mass with calcification of the tumor matrix, but it is adhered to the bone by a broad base and with cortical erosion, the same occurs with periosteal osteosarcoma.<sup>8</sup>

Treatment consists of surgical resection and subsequent adjuvant treatment, which represents the most controversial topic in the treatment of this tumor.<sup>3-5,14</sup> Numerous combinations of cancer treatment have been described, such as neoadjuvant or adjuvant chemotherapy and associated or isolated radiotherapy. In the large retrospective series by Longhi et al, there was a better response to chemotherapy for osteosarcoma. This includes doxorubicin, cisplatin, and isophosphamide, even if it is a soft tissue tumor.<sup>12</sup> Other studies obtained similar results, with a better therapeutic response for conventional osteosarcoma.<sup>17</sup> Adjuvant radiotherapy could have a role in an ESO >5 cm and with R0 margins.<sup>12</sup>

The recurrence rate of ESO ranges between 47% and 77% according to different authors, and metastases occur mainly at the lung, lymphatic, and bone levels.<sup>7,12,13</sup> A poor survival rate at 5 years has been described (25-76%).<sup>5,7</sup> Some factors for a better prognosis are: age <40 years, size <5 cm, chondroblastic or fibroblastic subtype, a resection with wide margins and also chemotherapy with multiple drugs.<sup>3,9</sup>

One of the important limitations in our case is not having a preoperative radiograph. We know that it is a fundamental study, since, as observed in the radiograph of the piece, the presence of intrasubstance calcifications can contribute to the diagnosis. It should be noted that ESO represents less than 2% of soft tissue tumors, is difficult to diagnose, as in our case, and has a long period from the onset of symptoms to diagnosis, as reported in the literature.

What is particular about this case is the diagnostic problem that it posed, since the biopsy was not conclusive for the definitive diagnosis and, instead, the study only reported a high-grade spindle cell sarcoma. For this reason, the study and radiograph of the piece were essential to arrive at the diagnosis of soft tissue osteosarcoma. Regarding the therapeutic problem that this case proposed, the treatment of soft tissue osteosarcoma is similar to that of bone osteosarcoma. Unlike other histologies, it is sensitive to chemotherapy as a rule; therefore, if the patient tolerates it, adjuvant chemotherapy is performed, as in this case.

## CONCLUSION

When faced with a tumor mass, it is essential to carry out a multidisciplinary assessment and promptly indicate the appropriate complementary studies, among which the importance of radiography and study of the piece stands out in order to reach a diagnosis as early as possible and improve the patient's prognosis.

Conflict of interest: The authors declare no conflicts of interest.

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