A Rare Case of Primary Extraosseous Osteosarcoma (EOO) of The Thigh: A Case Report

Chiheb Louizi[1], Ali Seif Amir Hosseini[1], Günther Engel[1], Omar Al-Bourini[1]*

Department of Clinical and Interventional Radiology, University Medical Center Göttingen, Germany

***Correspondence:** Omar Al-Bourini, Department of Clinical and Interventional Radiology, University Medical Center Göttingen, Robert-Koch-Str. 40, 37075 Göttingen, Germany

omar.al-bourini1@med.uni-goettingen.de

Radiology Case. 2024 December; 18(12):34-39 :: DOI: 10.3941/jrcr.5528

ABSTRACT

Background: Extraosseous osteosarcoma (EOO) is a rare mesenchymal malignancy, which produces osteoid, bone, or chondroid material and is located in the soft tissue without attachment to skeletal bones.

Case presentation: A 57-year-old male patient presented with extraosseous osteosarcoma located in the left rectus femoris muscle. The external magnetic resonance imaging revealed a large, irregular non-homogeneous contrast enhanced mass (largest diameter 9.5 cm). The final pathological diagnosis yielded extraosseous osteosarcoma. After interdisciplinary tumor board discussion, the following procedure was recommended: neoadjuvant systemic therapy with subsequent resection of the tumor and postoperative continuation of systemic therapy as well as discussion of adjuvant radiotherapy.

Conclusion: EOO should be treated as a soft tissue sarcoma with aggressive behavior and multimodality treatment should be actively sought to improve treatment outcome.

CASE REPORT

CASE REPORT

A 57-year-old male presented to our clinics sarcoma consultation in November 2023 for the first time with a swelling in the left thigh that has been increasing in size since the beginning of 2023.

The patient did not report any pain and the swelling did not affect his daily activities. His vegetative history is unremarkable apart from a “smoker’s cough” (40 py). The patient drinks alcohol occasionally and has no known allergies.

There was no history of weight loss, B symptoms or neurological failures in the months prior. The family history reveals that a sister suffers from a rheumatological disease and that the father had prostate cancer. The patient works as a plumber and had contact with composite materials containing asbestos. There was no previous history of trauma or radiation to that part of the thigh.

On examination, a 10 cm swelling presented on the anterior aspect of the left thigh at the level of the inguinal ligament. There was no palpable inguinal lymphadenopathy.

Imaging findings

The external MRI examination showed a 9.5 x 6.2 x 8.0 cm irregular tumor located intramuscularly in the left thigh with marginal enhancement, a partially cystic and an irregularly solid internal structure (Figure 1). The tumor was encapsulated and showed inhomogeneous contrast enhancement with peripheral mottled calcification on computer tomography (CT) images (Figure 2). There was no attachment to the bone and no neurovascular involvement.

Management

On the same day as the MRI examination a sonography-guided biopsy of the tumor was performed. Histology results were consistent with the diagnosis of extraosseous osteosarcoma.

The staging examination showed no evidence of distant metastases. After interdisciplinary tumor board discussion in early December, the following procedure was recommended: neoadjuvant systemic therapy with subsequent resection of the tumor and postoperative continuation of systemic therapy as well as discussion of adjuvant radiotherapy.

DISCUSSION

Etiology & demographics

Extraosseous osteosarcoma (EOO) is a rare mesenchymal neoplasm characterised by production of osteoid or bone but has no connection with the skeletal bones and periosteum. EOO was first described in 1941 by Wilson [1]. The incidence of extraosseous osteosarcoma is low, in one center accounting for only 4% of osteosarcomas and approximately 1% of soft tissue sarcomas [4]. This tumor usually occurs in the sixth or seventh decade of life and most frequently in the deep soft tissues of lower extremities, as well as in the upper extremities and retroperitoneum [3]. The pathogenesis of EOO remains unclear; however, long-term adverse factors, such as trauma, myositis ossificans, chronic bursitis and local radiotherapy, can be considered risk factors [4]. Here, we report a case of extraosseous osteosarcoma of the left thigh in a 57-year-old male.

Osteosarcoma in soft tissues is an uncommon type of soft tissue sarcoma. Extraosseous osteosarcoma is more frequently found in older patients than the skeletal form. Regarding aetiology many factors have been implicated. Such as radiation [4], articular implantation [10], previously benign lesions [11] and trauma [4].

Differential Diagnoses

Differential diagnosis such as myositis ossificans in its pseudomalignant form in which the microscopic findings may be mistaken for those of extraosseous osteosarcoma need to be ruled out [13]. In EOO, the active tumor cells forming osteoid, are in the periphery, while mature osteoid is present in the center. This is in contrast to the appearance of myositis ossificans [5].

Treatment & prognosis

The prognosis in extraosseous osteosarcoma is similar to that of skeletal osteosarcoma. Significant factors are patient age, tumor localization, histologic subtype and duration of symptoms before onset of treatment [6]. Tumour size has been found to be the only predictor of final outcome [3].

Radical surgery is advised since radiotherapy and/or chemotherapy are not always effective [2, 5]. EOO exhibit relative chemoresistance to doxorubicin-based chemotherapy [7]. The largest retrospective analysis on 60 EOO patients yielded a response rate to doxorubicin-based chemotherapy of only 25%, while doxorubicin combined with ifosfamide seemed to confer significant survival benefits [7]. The use of neoadjuvant chemotherapy and improvement in surgical technology have increased the survival rate to 65–75% [12].

The role of postoperative radiotherapy or chemotherapy in curatively resected soft tissue sarcomas remains controversial. The largest meta-analysis regarding adjuvant chemotherapy for soft tissue sarcomas was published in 1997 [8] and was updated in 2008 [9]. Adjuvant chemotherapy demonstrated a small but definite risk reduction in local recurrence, distant recurrence and overall recurrence.

TEACHING POINT

Extraosseous osteosarcoma should be treated as an aggressive soft tissue sarcoma and multimodal treatment should be actively sought to improve treatment outcome. The impact of adjuvant chemotherapy on survival of EOO should be further investigated.

QUESTIONS

1. Which of the following statements about extraosseous osteosarcoma (EOO) is false?

a. EOO is a rare mesenchymal malignancy that produces osteoid, bone-like, or chondroid material.

b. EOO is located in soft tissue without attachment to skeletal bone.

c. EOO is a benign tumor.

d. EOO is more common in older patients than the skeletal form.

e. EOO has no attachment to skeletal bone and periosteum.

Correct answer: c

Explanation: *Wilson H. Extraskeletal ossifying tumors. Ann Surg. 1941;113:95–112. doi: 10.1097/00000658-194101000-00013.*

2. EOO usually occurs in

a. Second decade of life

b. Third decade of life

c. Fourth decade of life

d. Sixth decade of life

e. Seventh decade of life

Correct answer: d and e

Explanation: *Bane BL, Evans HL, Ro JY, et al. Extraskeletal osteosarcoma. A clinicopathologic study of 26 cases. Cancer. 1990;65:2762–70.*

3. What risk factors can be considered for EOO?

a. Myositis ossificans

b. Chronic bursitis

c. Local radiotherapy

d. Chemotherapy

e. Chronic bursitis

Correct answer: a, b, c, and e

Explanation: *P.P. Sordillo, S.I. Hajdu, G.B. Magill, R.B. Golbey, Extraosseous osteogenic sarcoma. A study of 48 patients.*

4. Which of the following statements about the etiology of extraosseous osteosarcoma (EOO) is false?

a. Radiation

b. Joint implantation

c. Benign lesions

d. Trauma
e. Positive family history

Correct answer: a, b, c and d

Explanation: *P.P. Sordillo, S.I. Hajdu, G.B. Magill, R.B. Golbey, Extraosseous osteogenic sarcoma. A study of 48 patients.*

Penman HG, Ring PA (1984) Osteosarcoma associated with total hip replacement. J Bone Joint Surg [Br] 66: 632-634.

Kauffman SL, Stout AP (1963) Extraskeletal osteogenic sarcoma and chondrosarcomas in children. Cancer 16: 432-439.

5. What are the prognostic factors in EOO?
a. Patient age
b. Tumor location
c. Histologic subtype
d. Duration of symptoms before starting treatment
e. Gender

Correct answer: a, b, c and d

Explanation: *Bentzen SM, Poulsen HS, Kaae S, Jensen M, Johansen H, Mouridsen HT, Daugaard S, Arnold C (1988) Prognostic factors in Osteosarcomas. A regression analysis. Cancer 62:194-202REFERENCES*

AUTHORS' CONTRIBUTIONS

All authors contributed to this case report.

ACKNOWLEDGEMENTS

We would like to thank all of the Department of Radiology at University Medical Center Goettingen for their support in the treatment of patients.

DISCLOSURES

All the authors have indicated they have no financial relationships relevant to this article to disclose.

CONSENT

Yes

REFERENCES

1. 1. Wilson H. Extraskeletal ossifying tumors. *Ann Surg.* 1941; 113(1): 95–112. PMID: 17857721.
2. 2. Allan CJ, Soule EH. Osteogenic sarcoma of the somatic tissues: clinicopathologic study of 26 cases and review of literature. *Cancer.* 1971; 27(5): 1121-1133. PMID: 5281245.
3. 3. Bane BL, Evans HL, Ro JY, et al. Extraskeletal osteosarcoma. A clinicopathologic review of 26 cases. *Cancer.* 1990; 65(12): 2762–2770. PMID: 2160317.
4. 4. Sordillo PP, Hajdu SI, Magill GB, Golbey RB. Extraosseous osteogenic sarcoma. A review of 48 patients. *Cancer.* 1983; 51(4): 727-734. PMID: 6571801.
5. 5. Rao U, Cheng A, Didolkar MS. Extra-osseous osteogenic sarcoma: Clinico-pathological study of eight cases and review of literature. *Cancer.* 1978; 41(4): 1488-1496. PMID: 346199.
6. 6. Bentzen SM, Poulsen HS, Kaae S, et al. Prognostic factors in Osteosarcomas. A regression analysis. *Cancer.* 1988; 62(1): 194-202. PMID: 3164231.
7. 7. Ahmad SA, Patel SR, Ballo MT, et al. Extraosseous osteosarcoma: response to treatment and long-term outcome. *J Clin Oncol.* 2002; 20(2): 521-527. PMID: 11786582.
8. 8. Adjuvant chemotherapy for localised resectable soft-tissue sarcoma of adults: meta-analysis of individual data. Sarcoma Meta-analysis Collaboration. *Lancet.* 1997; 350(9092): 1647-1654. PMID: 9400508.
9. 9. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. *Cancer.* 2008; 113(3): 573–581. PMID: 18521899.
10. 10. Penman HG, Ring PA. Osteosarcoma in association with total hip replacement. *J Bone Joint Surg Br.* 1984; 66(5): 632-634. PMID: 6594340.
11. 11. KAUFFMAN SL, STOUT AP. Extraskeletal osteogenic sarcoma and chondrosarcomas in children. *Cancer.* 1963; 16: 432-439. PMID: 14031358.
12. 12. Mankin HJ, Hornicek FJ, Rosenberg AE, Harmon DC, Gebhardt MC. Survival Data for 648 Patients with Osteosarcoma Treated at One Institution. *Clin Orthop Relat Res.* 2004; (429): 286-291. PMID: 15577500.
13. 13. Lattes R (1981) Tumors of the soft tissues. Armed-Forces Institute of Pathology, Washington.

FIGURES

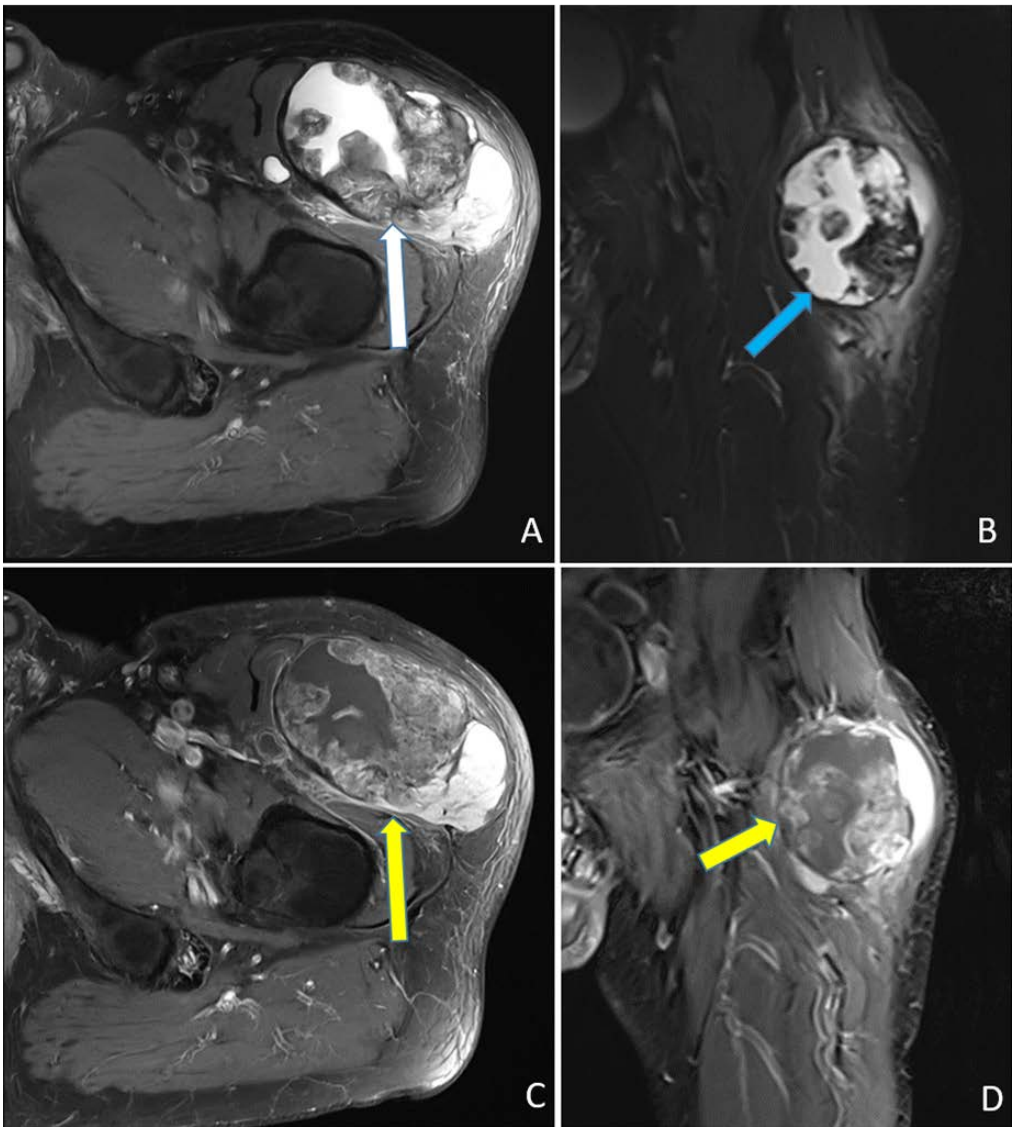

Figure 1: EOO in the left thigh. A (axial), B (coronal): Proton density sequence images without contrast demonstrate a 9.5 x 6.2 x 8.0 cm mass with mixed solid (yellow arrow) and liquid (blue arrow) components. C (axial), D (coronal): Contrast enhanced T1-weighted MRI images show inhomogeneous contrast uptake (yellow arrows).

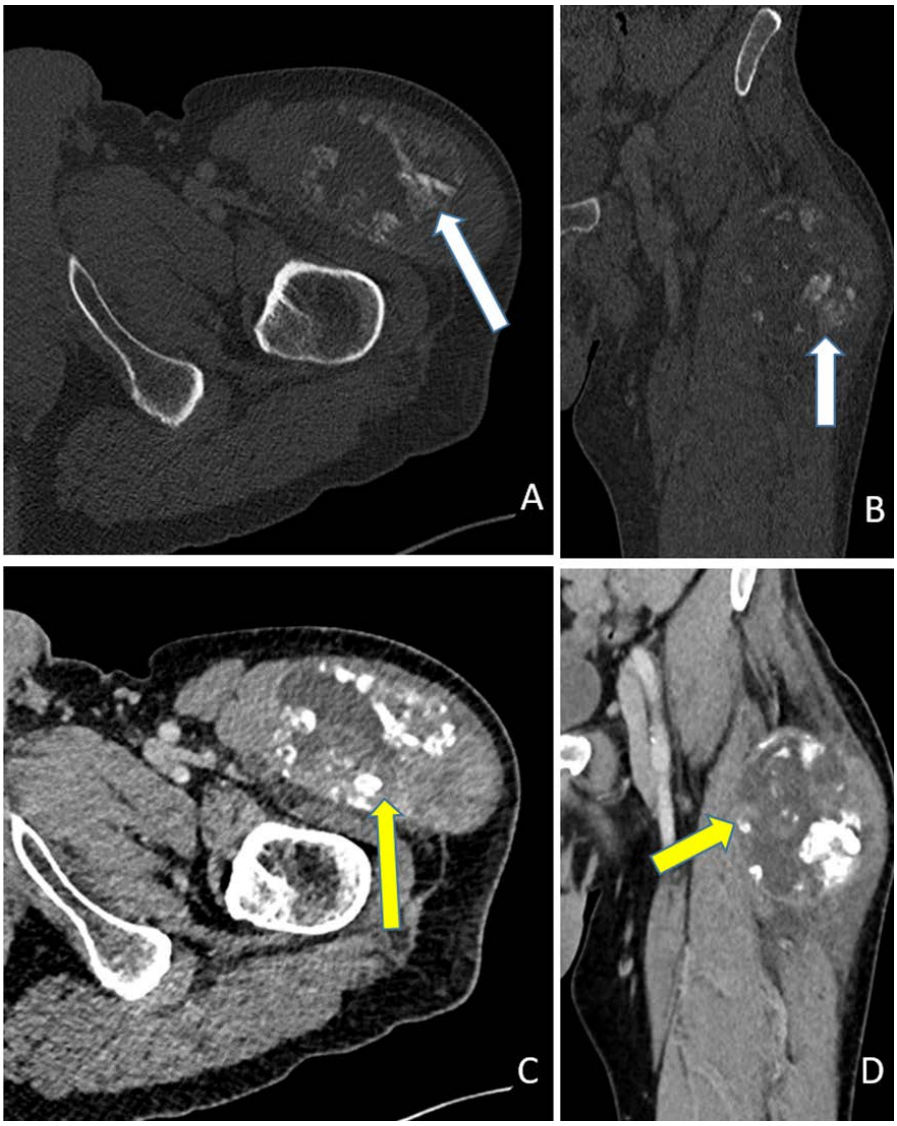

Figure 2: EOO in the left thigh (9.5 x 6.2 x 8.0 cm). A (axial), B (coronal): a large mass with peripheral mottled calcification (white arrows) could be seen on computer tomography (CT) images. C (axial), D (coronal): heterogeneous contrast enhancement (yellow arrows) can be observed on post-contrast CT images.

KEYWORDS

Extraosseous, Osteosarcoma, Cancer, Soft tissue osteosarcoma, EOO.

ABBREVIATIONS

EOO = EXTRAOSSEOUS OSTEOSARCOMA
MRI = MAGNETIC RESONANCE IMAGING
CT = COMPUTED TOMOGRAPHY

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/5528

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad

www.EduRad.org