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CLINICAL CASE

Dedifferentiated Parosteal Osteosarcoma of the Ulnar Diaphysis

Miguel Ángel Clara-Altamirano^{1*}, Dorian Yarih García-Ortega², Alejandro Maciel-Miranda³, Héctor Martínez-Said², Jorge Luis Martínez-Tlahuel⁴, Claudia Haydee Sarai Caro Sánchez⁵ and Mario Cuéllar-Hubbe⁵

¹Orthopedic Oncologist; ²Surgical Oncologist; ³Plastic Surgery Oncologist; ⁴Medical Oncologist; ⁵Pathology Oncologist; ⁶Surgical Oncologist, Head of Department, Department of Skin and Soft Tissues, Instituto Nacional de Cancerología, Ciudad de México, Mexico

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KEYWORDS

Dedifferentiated parosteal osteosarcoma; Intercalary resection; Microvascularized autologous graft

Abstract Introduction: Dedifferentiated parosteal osteosarcoma is a variant where a high-grade osteosarcoma coexists with a parosteal osteosarcoma. **Clinical Case:** A 20-year-old female patient presented with a 6-month history of right forearm pain and functional limitation, with no apparent cause. X-rays revealed a right ulnar diaphysis tumor lesion. On physical examination, there was pain on palpation at right ulna diaphysis and pronosupination limitation. Chest computed axial tomography showed metastatic disease to the left lung upper lobe. An incisional biopsy of the right ulna tumor reported a dedifferentiated parosteal osteosarcoma. Neoadjuvant chemotherapy with cisplatin and doxorubicin management was therefore started until 3 cycles were completed. Surgical treatment involved right ulna diaphysis intercalary resection plus microvascularized autologous right fibula diaphysis graft reconstruction and graft stabilization with a 3.5-mm DCP plate and a one-third tubular plate. In the same procedure, pulmonary metastasectomy was carried out by thoracoscopy. Postsurgical histopathological examination reported 100% necrosis. Currently, the patient is asymptomatic and with no evidence of tumor activity. **Conclusion:** Dedifferentiated parosteal osteosarcoma is a rare condition, but it should be suspected as differential diagnosis when a parosteal osteosarcoma is found. The fact that this condition can generate metastasis owing to its dedifferentiated pattern should be taken into account. It is important to plan a surgical treatment that enables appropriate functional reconstruction, always taking into account the oncologic principle. (creativecommons.org/licenses/by-nc-nd/4.0/).

*E-mail for correspondence: drmiguelclara@gmail.com (M.Á. Clara-Altamirano)

INTRODUCTION

Surface osteosarcomas comprise a varied group of malignant bone tumors with different degrees of malignancy^{1,2}. Within this group, dedifferentiated parosteal osteosarcoma is a variant where a high-grade osteosarcoma coexists with a parosteal or low-grade osteosarcoma either at the same time (synchronously) or as a recurrence (metachronally)^{2,3,4}.

Typical parosteal osteosarcoma is a superficial lesion formed by low-grade fibroblasts that produce laminar bone⁵. Usually, it occurs at between 20 and 40 years of age, with most common location being the distal femur posterior region^{1,6}.

There are only a few cases of dedifferentiated parosteal osteosarcoma published in the literature. Clinicopathological findings, diagnosis, treatment and evolution of patients with this rare osteosarcoma have not been clearly defined^{4,5,6}.

CLINICAL CASE

A 20-year-old female patient presented with a 6-month history of right forearm pain that limited its mobility, with no apparent cause. On physical examination she had tenderness at the right ulna diaphysis and pronosupination limitation owing to pain. Forearm radiographic study showed



Figure 1. Right forearm anteroposterior radiograph, where a radio-opaque image is observed at ulnar diaphysis middle third, with extension to soft tissues, consistent with parosteal osteosarcoma by imaging.

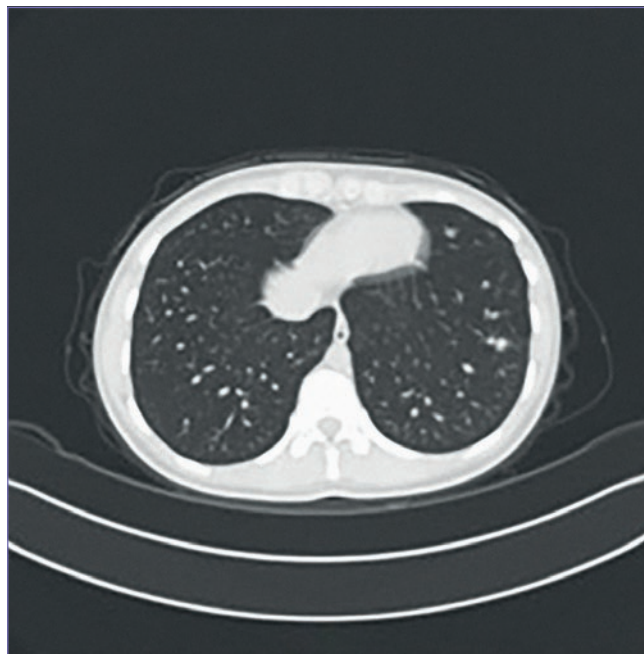


Figure 2. Chest CAT scan, with metastatic lesion present at left lung upper lobe.

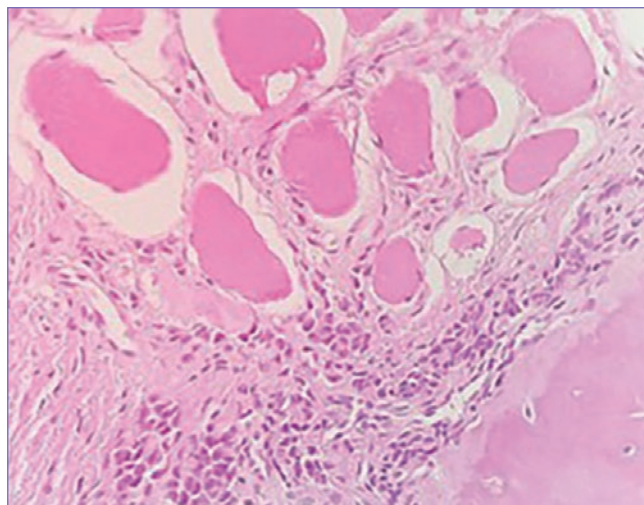


Figure 3. Biopsy (40x). Compact bone is identified at right lower corner of the image, and presence of active osteoblasts with atypia at the periphery and infiltrating adjacent muscle tissue (FM), as well as the presence of osteoid material deposits in between are also observed.

(Fig. 1) a cortical bone-dependent right middle-third ulnar diaphysis tumor with extension to soft tissues, radiologically consistent with parosteal osteosarcoma.

Chest CAT revealed multiple pulmonary nodules, probably related to deposits secondary to the known primary lesion, with the largest located at left lung base periphery, of 8 mm in size (Fig. 2). Middle third ulna diaphysis tumor incisional biopsy histopathological report was: dedifferentiated parosteal osteosarcoma of the right ulna diaphysis, Enneking III, AJCC IV A (Fig. 3).

The patient was started on neoadjuvant chemotherapy with cisplatin and doxorubicin until 3 cycles were completed. After neoadjuvant therapy, she underwent intercalary

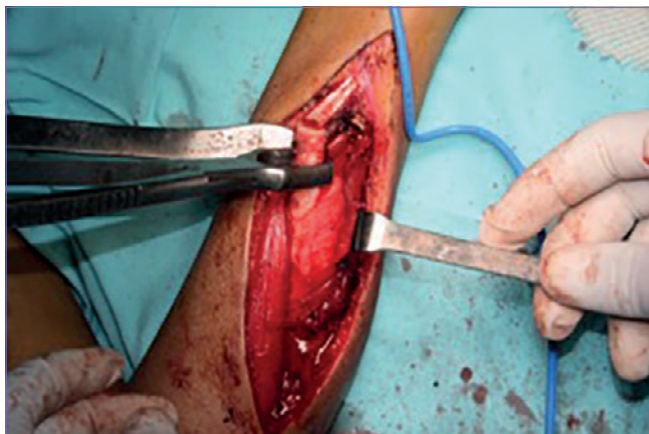


Figure 4. Right ulna diaphysis intercalary resection.

resection of the right ulna diaphysis plus microvascular autologous right fibula shaft reconstruction graft and graft stabilization by placing a 3.5-mm DCP plate and a one third tubular plate (Figs. 4 and 5). Subsequently, the thoracic surgery department carried out pulmonary metastasectomy by means of pulmonary wedge resection by thoracoscopy. Histopathology report indicated negative resection margins and 100% necrosis (Huvos grade IV), with metastasectomy product without evidence of viable neoplastic cells (complete response). Currently, at 12 months' follow-up, the patient is asymptomatic, with no evidence of local or distant tumor activity, and tolerating forearm and hand movement (Fig. 6).

DISCUSSION

Dedifferentiated parosteal osteosarcoma is a rare condition, but it should be considered when a parosteal osteosarcoma is found. In addition, the fact that this condition can generate metastasis owing to its dedifferentiated pattern should be taken into account.

Dedifferentiation in parosteal osteosarcoma is commonly reported in disease recurrences. Wold et al. described 11 cases of parosteal osteosarcoma dedifferentiation. In 10 of these cases, dedifferentiation was reported at first, second or third recurrence, whereas the coexistence of low and high-grade zones was only reported in one case⁷.

The presented case illustrates the importance of considering the possibility of dedifferentiated areas coexisting in parosteal osteosarcoma, as well as the possibility of these zones generating metastasis.

Several authors suggest that parosteal osteosarcoma should be considered a different entity to high-grade superficial osteosarcoma, since clinical evolution and mortality are different, with lower mortality rate for dedifferentiated osteosarcoma^{6,8,9}.

Invasion to the medullary canal is considered to be a prognostic factor for survival and disease-free interval in patients with dedifferentiated osteosarcoma^{6,10}.

Currently, survival rates of 80% are reported in patients with dedifferentiated parosteal osteosarcoma, with neoadjuvant chemotherapy plus surgical treatment with wide margins being associated^{5,6,10}.

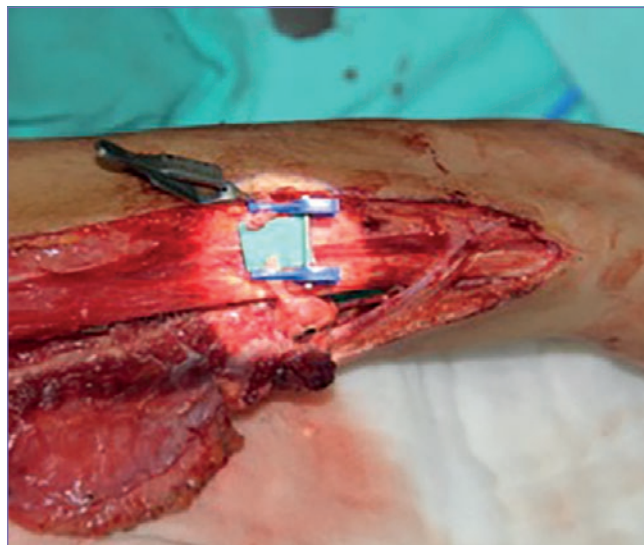


Figure 5. Vascularized fibular graft vascular anastomosis unto ulnar artery.

There is a strong correlation between prognosis and tumor necrosis volume in patients treated with neoadjuvant chemotherapy and surgery, with this percentage being assessed with Huvos classification¹¹, and with an excellent response being observed in this case (100% necrosis) after neoadjuvant treatment. In addition, it is important to consider that tumor debulking in response to neoadjuvant chemotherapy makes lesion excision much easier.

Reconstruction with vascularized fibula autologous graft enables better and faster integration of the graft in the



Figure 6. Postsurgical radiographic control, where osteosynthesis material and vascularized fibular graft adequate placement is observed.

recipient zone without causing donor area functional limitation¹². Taylor reported the first free vascularized fibular graft transfer in 1975¹³.

Weiland et al. have described the use of vascularized bone grafts for the treatment of several conditions, such as tumor resection, post-trauma bone defects and congenital pseudoarthrosis¹⁴⁻¹⁸. The role of this technique in long bone tumor reconstruction has been tried to be defined¹⁹; it has been proposed that it might be indicated in defects larger than 6 cm after tumor resection, and this technique enables the transfer of up to 24 cm of bone, with an 8-cm vascular pedicle on average²⁰.

In conclusion, dedifferentiated parosteal osteosarcoma is an osteosarcoma predominantly located on bone surface where a low-grade parosteal component is found associated with a high-grade sarcomatous component. Dedifferentiated parosteal osteosarcoma behavior is more aggressive than that of conventional parosteal osteosarcoma, and the presence of metastatic disease should be suspected in the presence of this pathology. It is important to plan a surgical treatment that enables appropriate functional reconstruction always taking into account the oncologic principle.

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