

CASE REPORT

DEDIFFERENTIATED LIPOSARCOMA OF THIGH WITH OSTEOSARCOMATOUS CHANGES: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT: Liposarcoma is the second most common type of soft-tissue sarcoma, accounting for 10%–35% of these lesions. Dedifferentiated liposarcoma represents a biphasic neoplasm, with one component being a well differentiated liposarcoma and the other a nonadipose cellular sarcoma. The risk of dedifferentiation depends substantially on location, and it occurs much more frequently in the retroperitoneum and less frequently in the extremities.^[1] Here we present a rare case of Dedifferentiated liposarcoma of left thigh with osteosarcomatous elements.

KEYWORDS: Liposarcoma, Dedifferentiation, Thigh.

INTRODUCTION: Dedifferentiated liposarcoma represents a malignant adipocytic neoplasm showing transition from atypical lipomatous tumor or well-differentiated liposarcoma to nonlipogenic sarcoma of variable histologic grade, usually at least several millimeters in diameter. It can originate in a primary lesion or occur in a recurrent tumor. In about 90% of cases, the dedifferentiated components are high-grade fibrosarcoma or malignant fibrous histiocytoma. Dedifferentiation most commonly occurs within well-differentiated liposarcoma in the retroperitoneum and inguinal regions, but it may also be encountered in the deep soft tissues of the extremities.^[2]

We present a case that demonstrates osteosarcomatous changes in a dedifferentiated liposarcoma in an extremity.

CASE REPORT: A 60-year-old female presented with a painless soft-tissue mass of the left thigh that had been progressively enlarging for about 8 months. At physical examination, the left proximal thigh was markedly larger than the right. A large firm mass was noted along the antero-medial aspect of the left thigh. The remainder of the physical examination findings were fairly unremarkable. CT scout tomogram shows a large soft-tissue mass in the anteromedial aspect of the thigh with focal calcifications within the proximal aspect of the mass. (Fig. 1)

The underlying femur was normal in appearance. Further evaluation by contrast enhanced CT was performed. Contiguous axial CT scans reveal the large soft tissue mass in the sub cutaneous and intramuscular planes on the medial aspect of proximal left thigh. The lesion demonstrates three components of differing attenuation - lipomatous region with thin septa (Fig. 2); region of low attenuation with Enhancing nodular lesions at the periphery (Fig. 3); and areas of calcification/ossification within the mass lesion (Fig. 4A and 4B). Underlying left femur appears normal. The patient underwent surgical debulking and Histopathology confirmed the diagnosis of liposarcoma with osteosarcomatous dedifferentiation.

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DISCUSSION: Liposarcoma is malignant tumor of mesenchymal origin. It is histologically divided into five types according to the most recent (2002) World Health Organization (WHO) classification:

- a) Well-differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes;
- b) Dedifferentiated;
- c) Myxoid;
- d) Round cell; and
- e) Pleomorphic.^[3]

The risk of dedifferentiation depends substantially on location. The estimated risk of dedifferentiation is 15% for retroperitoneal tumors and 5% for deep extremity lesions. Dedifferentiation in subcutaneous well-differentiated liposarcoma is extremely rare. It has been postulated that this predilection for dedifferentiation occurring more commonly in retroperitoneal lesions depends on time and size rather than site. Because retroperitoneal well-differentiated liposarcomas are present for a much longer time before diagnosis and are therefore larger at detection compared with extremity lesions, the prevalence of dedifferentiation is higher.^[1]

Dedifferentiation can occur “de novo” (in the original well-differentiated liposarcoma) or develop in local recurrent lesions and rarely following irradiation.

Histologically, the dedifferentiated tumor component resembles undifferentiated high-grade pleomorphic sarcoma in 90% of cases. In contrast, in about 10% of dedifferentiated liposarcoma, there is heterologous differentiation, often with rhabdomyosarcomatous, leiomyosarcomatous, or osteosarcomatous elements.^[4]

On imaging studies, the radiologic appearance of dedifferentiated liposarcoma reflects the histologic coexistence of a well-delineated fatty tumor and a closely apposed nonlipomatous mass. Because dedifferentiation usually occurs within a well-differentiated liposarcoma, a dedifferentiated liposarcoma shares radiologic features with the latter lesion. However, dedifferentiation is suggested by the additional presence of a focal, nodular nonlipomatous region greater than 1 cm in size. On CT images, these foci have attenuation similar to or slightly lower than that of skeletal muscle. These variations in imaging characteristics are likely related to histologic variations in myxoid (higher water content) and fibrous (with variable collagen content) content in the dedifferentiated region.

Calcification on imaging may suggest the presence of osteosarcomatous dedifferentiation.^[5]

The extent of calcification can vary from focal to diffuse ossification of the entire soft tissue mass. Radiography or computed tomography is superior to MR imaging in delineating the tumor mineralization or possible cortical bone erosion or destruction. The presence of osteosarcomatous elements is an adverse prognostic factor for dedifferentiated liposarcoma. It is important to recognize these foci, and biopsy must be directed at both the lipomatous and nonlipomatous components to ensure accurate pathologic diagnosis.^[6] Distant metastases from dedifferentiated liposarcomas occur in 15–20% of cases, and the most common sites of spread are the liver and lungs.

CONCLUSION: The presence of calcification or ossification in a liposarcoma indicates osteosarcomatous dedifferentiation. It's important to understand and recognize the imaging findings and direct biopsy of these regions for improved pathologic evaluation and a specific histologic diagnosis.

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FIG. 1: CT scout image showing a large soft-tissue mass in the anteromedial aspect of the thigh with focal calcifications (arrow) within the proximal aspect of the mass. The underlying femur normal in appearance.

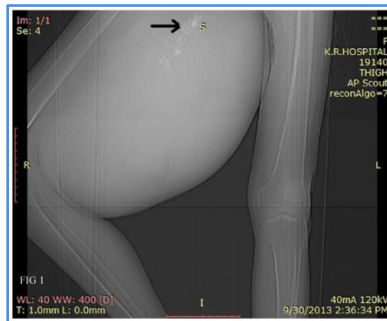


FIG. 1

FIG. 2: Axial contrast enhanced CT scan at the level of proximal thigh showing a large soft tissue mass in the sub cutaneous and intramuscular planes demonstrating a lipomatous region (asterisk) with thin enhancing septae within (arrowheads).



FIG. 2

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FIG. 3: Axial contrast enhanced CT scan showing region of low attenuation (asterisk) with enhancing nodular lesions at the periphery (black arrow).

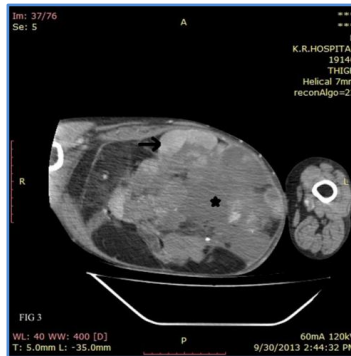


FIG. 3

FIG. 4A AND 4B: Axial and coronal reformatted CT images showing areas of calcification/ ossification within the mass lesion.



FIG. 4A AND 4B

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