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Case Report

# Dedifferentiated liposacrcoma with extensive calcification and ossification in pararenal region- A case report

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ABSTRACT

Sarcomas are the malignant tumour arising from mesenchymal tissue. Among the sarcomas, Liposarcoma has been one of the most common malignacy. Calcification and ossification can occur in liposarcoma; however, the presence of both ossification and calcification, a very rare entity. We present a case of a extensive calcification of dedifferentiated liposarcoma arising in the pararenal site in a 48 year old male. Retroperitoneal are more common among the sites. This present case presented as tumour arising the lower pole of right kidney as a solid calcified mass without infiltrating the kidney is very rare presentation.



## 1. Introduction

Sarcomas are the malignant tumours arising from the mesenchymal tissue. Liposarcoma is the most common malignant tumor in the retroperitoneal soft tissue, and this tumor is classified according to the amount of lipid inside the cells, the mucoid lipid and the degree of cell differentiation. This tumor is classified into the well differentiated, myxoid, round cell, pleomorphic and dedifferentiated types and the pleomorphic type is known to be the most common type. In dedifferentiated liposarcoma, the well and poorly differentiated liposarcoma and non-lipomatous sarcoma coexist.

## 2. Case Report

A 48 year old man presented with mild abdominal pain and pain during micturition. Further on examination, a mass is felt in the right hypogastric region and advised for . Patient doesn't complain of trauma or any accident or no history similar complaints in the family. On ultrasonography found pararenal mass with calcification Suspecting renal malignancy and advised X ray. Radiographs revealed a extensive calcified tumour abutting the kidney [Figure 1]. Followed by MDCT is done and given as ill-defined fat lesion and showed a densely calcified tumour in the paranephric space measuring 16x12x11cms. Lesion is adherent to outer cortex of the right kidney, superiorly extending to subhepatic region. Duodenum and pancreas are is displaced anteromedially [Figure 2]. Followed that case with multiteam disciplinary meeting and posted for excision of tumour with nephrectomy, during the surgery the tumour was abutting to right kidney and not infiltrating into the kidney cortex. Surgeon removed the tumour without nephrectomy and received the specimen for histopathological evaluation. On gross examination the tumour is well circumscribed and outer surface shows multiple nodular appearance weighting 1.9kgs [Figure 2]. Cut surface shows cystic spaces in the calcified areas, and at the periphery a pale yellow to grey in appearance measuring 5x3x2cm soft tissue seen [Figure 2].

On microscopy the tumour showed well differentiated and dedifferentiated areas having lipoblast with stomal cells, bizarre multinucleated cells, small round cells, large areas of collagenization, calcification, and chondromyxoid

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differentiation [Figure 3 (a-f)]. It's a difficult case to rule out extra-skeletal osteosarcoma as the tumour had large areas of calcification. Followed by MDM2 immunohistochemistry is done and confirmed the origin of tumour. The present case been followed up.

## 3. Discussion

Sarcomas are the malignant tumours arising from mesenchymal tissue. These tumours are classified based on the cell of origin and clinical behaviour. On the bases of cell of origin, sarcoma tumour arising from the adipocytes are the liposarcomas. Liposarcoma is defined as a malignant mesenchymal neoplasm that is composed of lipogenic tissue with a varying degree of cellular atypia, with lipoblast and possibly including nonlipogenic sarcoma cells and atypical mitotsis.[1] Based on clinical behaviour, tumour arising from adipocytes are classified as benign, intermediate/locally aggressive and malignant liposarcoma.[1,2] Accumulating evidence has demonstrated that the clinical outcomes can vary dramatically according to different histologic subtypes and location. Liposarcoma is prevailingly affecting adults with an equal distribution between males and females. Liposarcoma represents 12.8% of all sarcomas, specifically, 24% of all extremity and 45% of all retroperitoneal soft-tissue sarcomas. The peak age of onset are in the fifth to seventh decades. The present case is in 48 year old male is located retroperitoneally in the right pararenal region without infiltrating into the renal system and presented as dense calcified mass A retroperitoneal tumour often develops in people in their 40's to 50's, and especially in males when compared with females, but it is a very rare tumour that accounts for less than 0.2% of all types of malignant tumours.[2] Based on its histological features, liposarcoma was initially classified into four subtypes: the well differentiated, myxoid, round cell and pleomorphic types. Evans introduced dedifferentiated liposarcoma later in 1979, so now there are five subtypes.[3] A new update on 2013 WHO classification of tumours has been classified adipocytic tumour as intermediate (locally aggressive) and malignant tumour as dedifferentiated, myxoid pleomorphic and liposarcoma (not otherwise specified).[2–4] Approximately 90% of cases occur de novo, whereas 10% develop in recurrences.[3,4] The feature of dedifferentiated liposarcoma is the histological coexistence of well to poorly differentiated liposarcoma and non-lipomatous differentiated areas like cartilaginous and calcified areas.[2,5] Dedifferentiated liposarcoma is the least common type of liposarcoma accounting for about 5% of all liposarcoma cases.[1,5] Dedifferentiation occurs in up to 10% of well differentiated (WD) liposarcomas of any subtype, although the risk of dedifferentiation appears to be higher when dealing with deep seated retroperitoneal lesions[3,5], but more than 90% of lesion having calcified areas are extremely rare. Evans HL and Nassif N A et al suggested that Well-differentiated liposarcoma and/or atypical lipomatous tumour and dedifferentiated liposarcoma are considered to be along the same spectrum of disease with dedifferentiated liposarcoma arising from within well- differentiated liposarcoma and/or atypical lipomatous tumour with a mean time to dedifferentiation of 7-8years after diagnosis of well-differentiated liposarcoma and/or atypical lipomatous tumour.[3,4] Moon WS et al studied that dedifferentiated liposarcoma is inherently more aggressive and more likely to metastasize than well-differentiated liposarcoma and/or atypical lipomatous tumour with dedifferentiated liposarcoma having distant metastases in 15%-20% of cases with lungs and liver being most common.[6] Kim ES et al studied that the histological hallmark of dedifferentiated liposarcoma is represented by the transition from ALT/WD liposarcoma of any type to non-lipogenic sarcoma, which, in most cases, is high grade[5]. The extent of dedifferentiation is variable but most often this component is evident to the naked eye. The prognostic significance of microscopic foci of dedifferentiation is uncertain[2]. The transition usually occurs abruptly. However in some cases this can be more gradual and, exceptionally, low grade and high grade areas appear to be intermingled. Dedifferentiated liposarcoma may exhibit heterologous differentiation in about 5-10% of cases which apparently does not affect the clinical outcome. The present case is presented with dense calcified mass situated in the lower pole of kidney A peculiar "neural- like" or "meningothelial-like" whorling pattern of dedifferentiation has recently been described.[2] This pattern is often associated with ossification. Dedifferentiated liposarcoma appears to exhibit less aggressive clinical behaviour when compared with other high grade pleomorphic sarcomas. Careful and extensive sampling is therefore mandatory, particularly in large retroperitoneal lesions, as the well differentiated component may be overlooked. Additionally, it should be noted that local recurrences of dedifferentiated liposarcoma may be entirely well differentiated.[4] Dedifferentiated liposarcoma is characterized by a tendency to recur locally in at least 40% of cases. However, almost all retroperitoneal examples seem to recur locally if the patients are followed for 10-20 years.[4,6] Distant metastases are observed in 15-20% of cases with an overall mortality ranging between 28 and 30% at 5 years follow-up, although this figure is undoubtedly much higher at 10-20 years.[6–8] The most important prognostic factor is represented by anatomic location, with retroperitoneal lesions exhibiting the worst clinical behaviour. French Federation of Cancer Centers Grading System for Soft Tissue Sarcomas are used for grading these tumours.[2,3] The extent of dedifferentiated areas does not seem to predict the outcome. Interestingly, dedifferentiated liposarcoma, despite its high grade morphology, exhibits a less aggressive

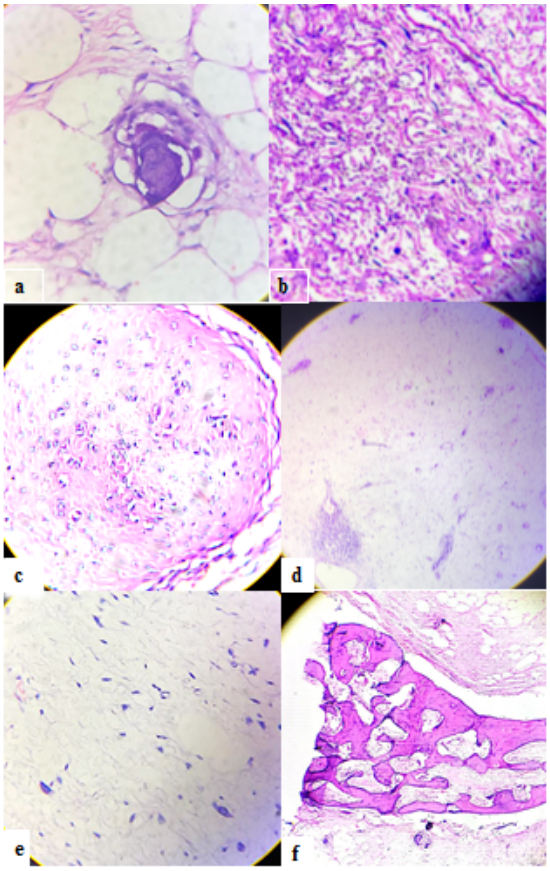


**Fig. 3:** Microscopic picture showing lipocytes andossification(a), spindle cell differentiation(b), cartilaginous differentiation(c), lipocytes(d), lipoblasts(e) and osteoid differentiation(f)

clinical course than other types of high grade pleomorphic sarcoma, although the basis for this difference is unknown.

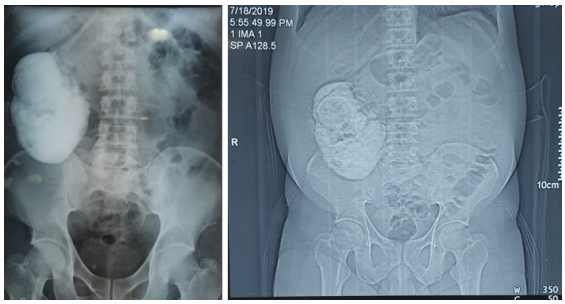


**Fig. 1:** X-ray and CT showing calcified tumour in the right perinephric region.

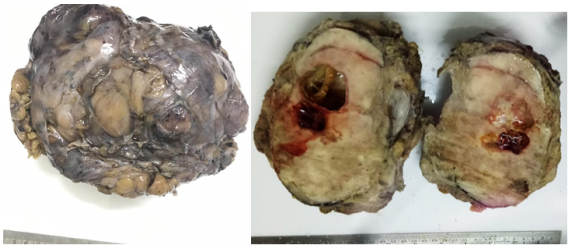

**Fig. 2:** Gross picture of dedifferentiated liposarcoma. Cut surface shows solid and cystic areas.

## 4. Ethics approval and consent to participate

Written informed consent was obtained from patients. Each author certifies that all investigations were conducted in conformity with the ethical principles.

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None.

## 6. Conflict of Interest

None.

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