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# The Blubbery Addle- Dedifferentiated Liposarcoma

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**Editorial**

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## Introduction

Dedifferentiated liposarcoma represents a transition within primary or reoccurring well differentiated liposarcoma typically into non lipogenic sarcoma. Although contemplated as a high grade neoplasm, dedifferentiated liposarcoma may demonstrate variable tumour grading.

Characteristically, dedifferentiated liposarcoma is configured of non lipogenic sarcoma which emerges from well differentiated liposarcoma. However, precursor element of well differentiated liposarcoma may or may not be discernible.

Dedifferentiated liposarcoma is categorized into low grade variant and high grade variant. Exceptionally, high grade neoplastic component may be lipogenic and simulates pleomorphic liposarcoma or focal homologous lipoblastic differentiation.

Typical molecular alterations are comprised of ring, giant marker or rod chromosomes constituted of genetic material of chromosome 12q13-15. Consequently, localized amplification of various neighbouring genes as MDM2 may occur [1,2].

Dedifferentiated liposarcoma exhibits chromosome 12q14 amplification on account of supernumerary ring or giant chromosome originating from chromosome 12q13-15 region. Proportionate copy number variation appears concurrent with progression into tumours of advanced grade. Neoplasm may exemplify genomic amplification of MDM2 gene.

Retroperitoneal high grade sarcoma occurring in adults may frequently be comprised of dedifferentiated liposarcoma. Retroperitoneal neoplasms configuring inflammatory subtype of undifferentiated pleomorphic sarcoma are contemplated to represent common morphological patterns of dedifferentiation within dedifferentiated liposarcoma [1,2]. Dedifferentiated liposarcoma articulating as a retroperitoneal sarcoma requires exclusion from diverse retroperitoneal sarcomas which may be obtained by cogent tissue sampling, especially obtained from peripheral tumour zones along with precise cytogenetic evaluation, immunostaining with MDM2 and CDK4 or molecular assay for discerning chromosomal 12q13-15 amplification [1,2].

Dedifferentiated liposarcoma is significantly heterogeneous and manifests multiple configurations of tumour differentiation and evolution. Ancillary investigations as confirmation of protein overexpression or genetic amplification may be beneficially adopted to categorize diagnostically challenging neoplasms.

Majority of dedifferentiated liposarcomas are configured of advanced grade tumours. However, low grade dedifferentiated liposarcoma is a contemporary and established entity [2,3]. Low grade liposarcoma with dedifferentiation may histologically mimic cellular variant of well differentiated liposarcoma. Generally, neoplasms with dedifferentiation are accompanied by adverse prognostic outcomes.

Although debatable, minimal dedifferentiation necessitates macroscopic evidence of dedifferentiation in > 1.0 centimetre magnitude of tumour parenchyma.

Tumefaction with minimal dedifferentiation < 1.0 centimetre diameter are associated with inferior prognostic outcomes, in contrast to well differentiated liposarcoma. Thus, extended clinical monitoring is recommended, irrespective of tumour magnitude.

Characteristically, dedifferentiated liposarcoma occurs in older adults. A mild male predilection is observed [2,3]. Dedifferentiation ensues in up to 10% of well differentiated liposarcomas. Primary retroperitoneal well differentiated liposarcoma is frequently associated with dedifferentiation. Generally, subcutaneous atypical lipomatous tumour is devoid of differentiation.

Dedifferentiated liposarcoma is commonly discerned within retroperitoneum, followed in frequency by extremities. Spermatic cord may be commonly incriminated. Neoplasm is exceptionally discerned within head and neck. Tumefaction encountered within subcutaneous tissue is extremely exceptional [2,3].

Upon cytological examination, hyper-cellular smears are composed of multinucleated, pleomorphic tumour giant cells incorporated with abundant cytoplasm. Spindle shaped cells with elongated nuclei or miniature clusters of neoplastic cells demonstrating enhanced nucleocytoplasmic ratio may be discerned. Occasionally, osteoclast-like giant cells may be delineated. Cytological samples may be appropriately subjected to molecular techniques of neoplastic confirmation [2,3].

Upon gross examination, an enlarged, firm, coarsely lobular tumefaction simulating fish flesh appearance is encountered. Peripherally, neoplasm demonstrates a well differentiated, adipose tissue rich component. Foci of dedifferentiation can be discrete, nodular or evolve gradually. Advanced grade tumefaction may delineate focal zonal necrosis [2,3]. Upon microscopy, advanced grade dedifferentiated liposarcoma demonstrates foci of well differentiated and dedifferentiated components with an abrupt or gradual transition between dual components. Dedifferentiated component of high grade neoplasm is cellular and characteristically manifests as a non-lipogenic sarcoma enunciating significant cellular and nuclear pleomorphism. Generally, proportionate mitotic activity of > 5 mitosis/10 high power fields is optimal for classifying high grade neoplasms [2,3]. Dedifferentiated liposarcoma may frequently simulate undifferentiated pleomorphic sarcoma and exemplify abridged fascicles of pleomorphic, spindle shaped cells intermingled with an acute and chronic inflammatory infiltrate. A peculiar 'whirling' neoplastic configuration appears reminiscent of meningothelial articulations. Around ~10% neoplasms depict heterologous elements as foci of neural differentiation, leiomyosarcoma, osteosarcoma, chondrosarcoma, rhabdomyosarcoma, pleomorphic liposarcoma or homologous zones of lipoblastic dedifferentiation. Besides, focal angiosarcomatous differentiation is documented. Neoplasms with focal rhabdomyoblastic differentiation are associated with inferior prognostic outcomes [2,3].

The infrequently discerned, low grade, dedifferentiated liposarcoma appears reminiscent of fibromatosis or well differentiated fibrosarcoma. Neoplasm may be non lipogenic, in contrast to well differentiated spindle cell liposarcoma constituted of atypical lipoblasts or adipose tissue cells. In contrast to conventional atypical lipomatous tumour, low grade dedifferentiated liposarcoma mandates a definitive neoplastic category as contemplated by contemporary World Health Organization (WHO) classification and manifests with inferior prognostic outcomes. Neoplastic perimeter of dedifferentiated liposarcoma may demonstrate foci of well differentiated liposarcoma which simulates aggregates of compressed mature adipose tissue delineating reactive alterations [3,4] (Figures 1 & 2).

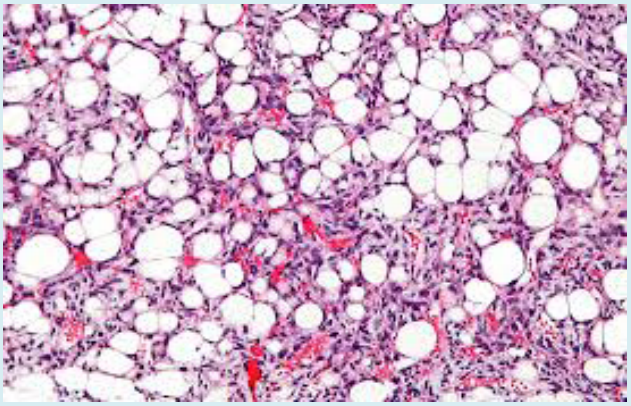

**Figure 1**: Dedifferentiated liposarcoma delineating abridged fascicles of spindle shaped cells intermingled with acute and chronic inflammatory cells and focal haemorrhage. Non lipogenic foci are intermixed with aggregates of mature adipose tissue cells [5].

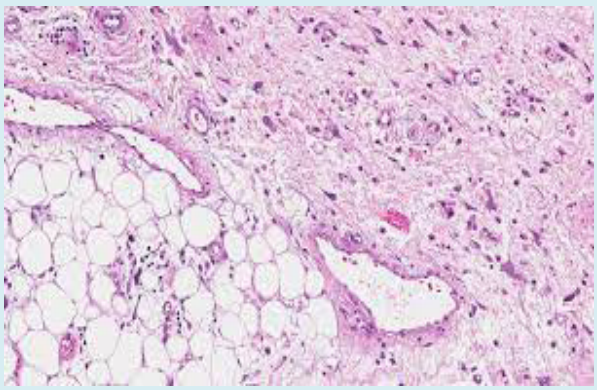

**Figure 2**: Dedifferentiated liposarcoma demonstrating a whirling pattern configured of short fascicles of spindle shaped cells commingled with acute and chronic inflammatory cells, patent capillaries and aggregates of mature adipose tissue cells [6].

## TNM Staging of Retroperitoneal Sarcoma

### Primary Tumour

- TX: Primary tumour cannot be assessed.
- T0: No evidence of primary tumour.
- T1: Tumour ≤ 5 centimetres in greatest dimension.
- T2: Tumour > 5 centimetres and ≤ 10 centimetres in greatest dimension.
- T3: Tumour >10 centimetres and ≤15 centimetres in greatest dimension.
- T4: Tumour > 15 centimetres in greatest dimension [3,4].

### Regional Lymph Nodes

- N0: Regional lymph node involvement absent.
- N1: Regional lymph node involvement present.

### Distant Metastasis

- M0: Distant metastasis absent.
- M1: Distant metastasis present.

### Histological Grading of Soft Tissue Sarcoma (G)

- G: Tumour grade cannot be assessed
- G1: Total differentiation, mitotic count and tumour necrosis score of 2 or 3.
- G2: Total differentiation, mitotic count and tumour necrosis score of 4 or 5.
- G3: Total differentiation, mitotic count and tumour necrosis score of 6, 7 or 8 [3,4].

## Staging of Retroperitoneal Sarcoma as per American Joint Committee on Cancer (8th edition)

- **Stage IA:** T1, N0, M0, GX or G1.
- **Stage IB:** T2, T3 or T4, N0, M0, GX or G1.
- **Stage II:** T1, N0, M0, G2 or G3.
- **Stage IIIA:** T2, N0, M0, G2 or G3.
- **Stage IIIB:** T3 or T4, N0, M0, G2 or G3 OR Any T, N1, M0, Any G.
- **Stage IV:** Any T, Any N, M1, Any G [3,4].

Dedifferentiated liposarcoma appears immune reactive to MDM2, CDK4, p16, vimentin, p53 or Rb gene. Focal immune reactivity to smooth muscle actin is encountered with peroxisome proliferator-activated receptor (PPAR) gamma [4,7].

Dedifferentiated liposarcoma requires segregation from neoplasms such as leiomyosarcoma, malignant peripheral nerve sheath tumour, malignant melanoma, pleomorphic liposarcoma, rhabdomyosarcoma, diverse subtypes of sarcoma infiltrating abutting adipose tissue, sarcomatoid carcinoma, sarcomatoid mesothelioma, undifferentiated pleomorphic sarcoma or malignant fibrous histiocytoma. Upon imaging, an adipose tissue and solid, non-adipose tissue component is observed [4,7].

In contrast to diverse high grade pleomorphic sarcomas, dedifferentiated liposarcoma is associated with superior prognostic outcomes. However, tumour reoccurrence may ensue within 40% to 75% neoplasms and distant metastasis ensues within 10% to 15% tumefaction. Neoplasm is associated with ~28% proportionate mortality [4,7]. Tumours of advanced grade as categorized with Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading and focal myogenic differentiation delineate adverse clinical outcomes. Distant metastasis can arise within low grade or high grade neoplasms with dedifferentiation. Distant metastasis is singularly comprised of dedifferentiated neoplastic component [4,7]. Minimal proportionate neoplastic dedifferentiation occurring within low grade or high grade neoplasms remains non concurrent with emergence of distant metastasis. Instances with myxofibrosarcoma-like morphological features appear especially aggressive. Retroperitoneal dedifferentiated liposarcoma demonstrates enhanced proportionate localized tumour reoccurrence or disease specific mortality. With extended monitoring, retroperitoneal tumours comprehensively (~100%) demonstrate tumour reoccurrence [4,7].

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5. Image 1 Courtesy: Wikimedia commons.

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