Case Report

Dedifferentiated Liposarcoma: A Rare Entity

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Abstract

Background: Dedifferentiated liposarcoma is defined as well-differentiated liposarcoma juxtaposed to areas of high-grade non-lipogenic sarcoma, usually resembling fibrosarcoma or undifferentiated pleomorphic sarcoma. The retroperitoneum is a common site; fewer than 20% of dedifferentiated liposarcomas occur in the head, neck, and rarely in the subcutis.

Case History: A 36-year-old male patient presented to the surgery department of our hospital with the chief complaint of a recurrence of a soft tissue mass over the left upper shoulder region. There is a history of a previous operation at the same site. In the MRI report, a well-defined lobulated lesion was noted in the subcutaneous plane over the left scapular region. The lesion is heterogeneously hyperintense on T2 and STIR and shows areas of cystic changes. A single globular skin-covered specimen measuring 10 x 6.0 x 5.0 cm. The outer surface of the skin is smooth, shiny, and varies in color from grey-white to grey-brown, with the lesion protruding from the skin (FIG 1). On the cut surface, a grayish, shiny, smooth, and cystic appearance is present. Multiple sections studied from the lesional tissue show both well-differentiated liposarcomatous and non-lipogenic (dedifferentiated) components. The liposarcomatous area consists of mature adipocytes and atypical spindle cells. Non-lipogenic components consist of highly cellular areas showing hemangiopericytoma-like vascular patterns, and in some places, malignant fibrous histiocytoma (MFH)-like areas are also noted.

Discussion: Dedifferentiated liposarcoma comprises less than 10% of all liposarcomas. It is found most often in the retroperitoneum and rarely in the extremities, head, and neck region. The incidence is approximately 1 in 330,000 persons per year.

Keywords:

Dedifferentiated Liposarcoma, Pleomorphic Liposarcoma, Lipogenic, Non-lipogenic

Introduction

Dedifferentiated liposarcoma is defined as well-differentiated liposarcoma juxtaposed to an area of high-grade non-lipogenic sarcoma, usually resembling high-grade fibrosarcoma, malignant fibrous histiocytoma, hemangiopericytoma, rhabdomyosarcoma, osteosarcomatous, and leiomyosarcomatous elements [1]. Dedifferentiated liposarcoma is found most often in the retroperitoneum and rarely in the extremities and head and neck region [2].

Well-differentiated liposarcoma does not metastasize but can dedifferentiate into dedifferentiated liposarcoma. Dedifferentiated liposarcoma is more aggressive in clinical behavior, with a greater likelihood of local recurrence and capacity for metastasis. It typically has the appearance of undifferentiated pleomorphic or spindle cell sarcoma. Both sarcomas are associated with high-level amplification in chromosome 12q13-15.

Case Report

A 36-year-old male patient presented to the surgical department of our hospital with the chief complaint of a large swelling over the left upper scapular region for one year. The swelling was initially small in size but later progressed to a larger size. A past history revealed an operation at the same site 3 years ago, where pleomorphic liposarcoma was diagnosed histopathologically. Due to poor compliance, the patient did not complete the postsurgical treatment. He now presented with recurrence of the swelling to the surgery department.

The MRI report showed a well-defined lobulated lesion in the subcutaneous plane over the left scapular region, which was heterogeneously hyperintense on T2 and STIR, showing areas of cystic changes with calcification. The patient underwent another operation, and the specimen was sent for histopathological examination.

On gross examination, a single globular skin-covered specimen measuring 10 x 6.0 x 5.0 cm was observed. The outer surface showed a smooth, shiny, grey-white to grey-brown lesion protruding from the skin (Fig 1). The cut surface was grayish, shiny, smooth, solid, and cystic, with a cyst measuring 0.4 cm in diameter filled with serous fluid (Fig 2-3).

Histopathological sections studied from the lesional tissue showed both well-differentiated liposarcomatous and non-lipogenic (dedifferentiated) components. The liposarcomatous area consisted of mature adipocytes and atypical spindle cells, with multivacuolated lipoblasts embedded in the loose stroma. The non-lipogenic components consisted of highly cellular areas showing hemangiopericytoma-like vascular patterns, and in some areas, malignant fibrous histiocytoma (MFH)-like features were also noted. The stroma was infiltrated by tumor-associated dense inflammatory cell infiltrates.

The immunohistochemistry test was positive for MDM2 (Fig 8) and VIMENTIN (Fig 9) in the non-lipogenic area, and negative for S-100 (Fig 10). After the second surgical intervention, the patient underwent 3 cycles of adjuvant radiotherapy, and no recurrence has been reported to date.

This case report has been written after obtaining written informed consent from the patient. Ethical approval is not applicable.

Discussion

Dedifferentiated liposarcomas are most commonly seen in the retroperitoneum (80%), extremities, spermatic cord, and other sites of the internal trunk [2], demonstrating the rarity of these lesions in the head and neck liposarcomas. They were originally thought to represent a distinct entity of high-grade disease within a well-differentiated liposarcoma [3, 4]. Dedifferentiated liposarcoma was described as a "tumor composed of areas of a non-lipogenic sarcoma associated with an atypical lipomatous tumor or well-differentiated liposarcoma." Current research concludes that dedifferentiated liposarcoma likely represents a progression of disease from well-differentiated liposarcoma to either a high or low-grade lesion. These tumors contain a large-scale amplification of sequences in the 12q13-15 region, which includes the MDM2 gene, and 90% contain an amplification of CDK4. These mutations work in concert to extend cell survival and underlie the neoplastic process, with MDM2 as a known strong inhibitor of p53, thereby decreasing apoptosis, and with CDK4 phosphorylating Rb gene products, thus helping the cell to proceed unchecked

through the G1–S checkpoint [3, 4].

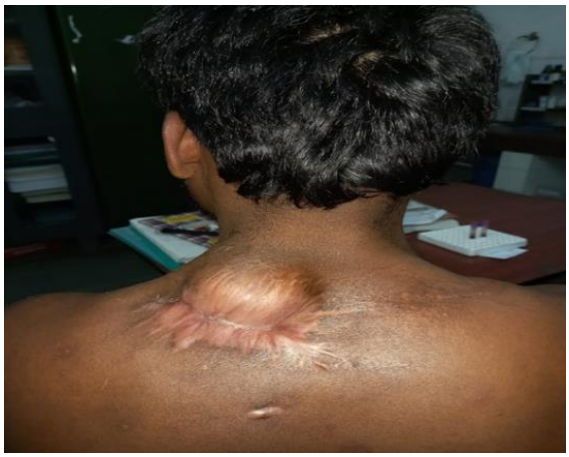

Figure 1: Swelling over left upper scapular region. Swelling was initially small in size then later progressed to larger size.

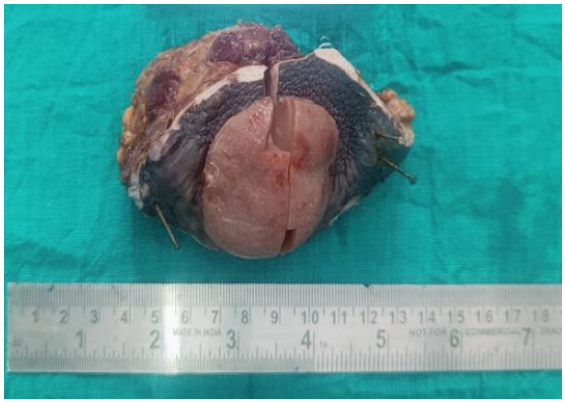

Figure 2: Single globular skin covered specimen measuring 10 x 6.0 x 5.0 cm, Outer surface skin covered shows smooth, shiny, grey white to grey brown lesion protruded from the skin.

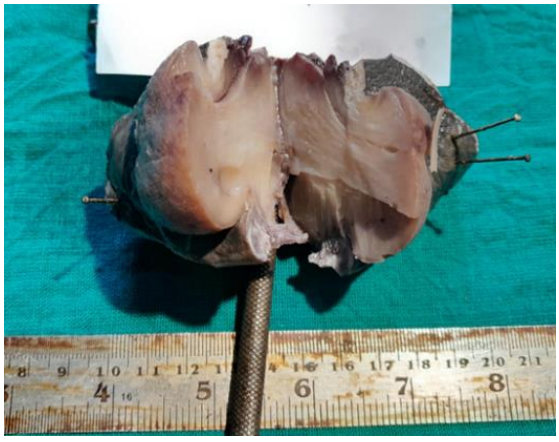

Figure 3: On cut surface grayish shiny, smooth, solid and cystic surface & cyst measuring 0.4 cm in diameter containing serous fluid.

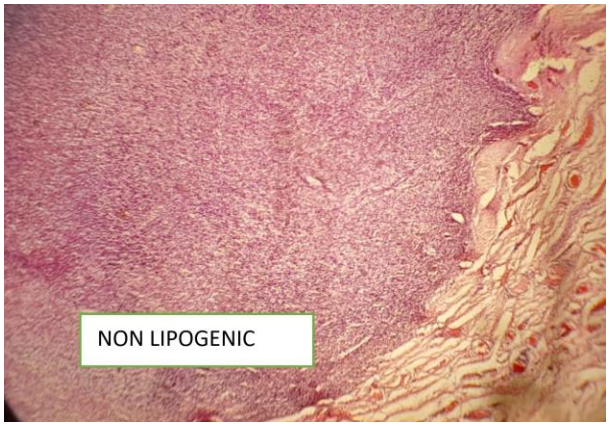

Figure 4: Nonlipogenic component consists of highly cellular area showing hemangiopericytoma-like vascular patterns and at places malignant fibrous histiocytoma (MFH) like areas are also noted. (H &E Stain, x10)

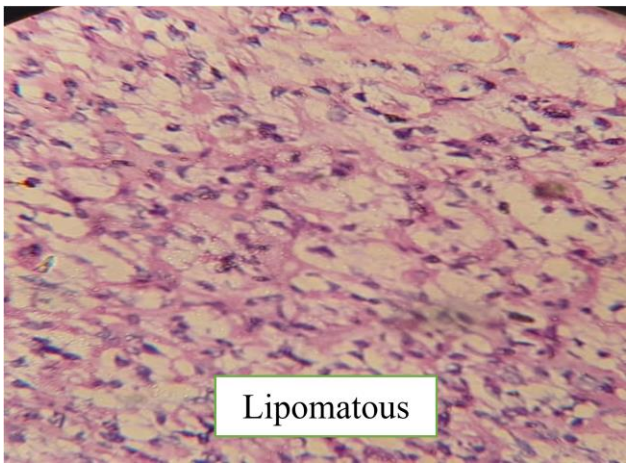

Figure 5: Liposarcomatous area consists of mature adipocytes and atypical spindle cells. Multivacuolated lipoblast embedded in the loose stroma. (H &E Stain, x40)

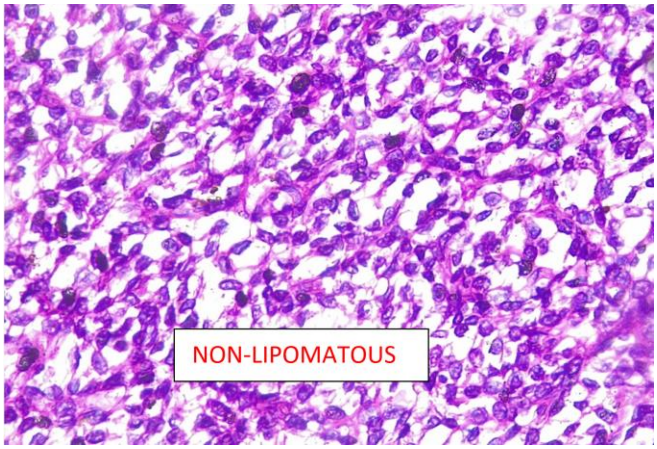

Figure 6: Stains the nucleus of non-lipomatous area of dedifferentiated liposarcoma (H &E Stain, x40)

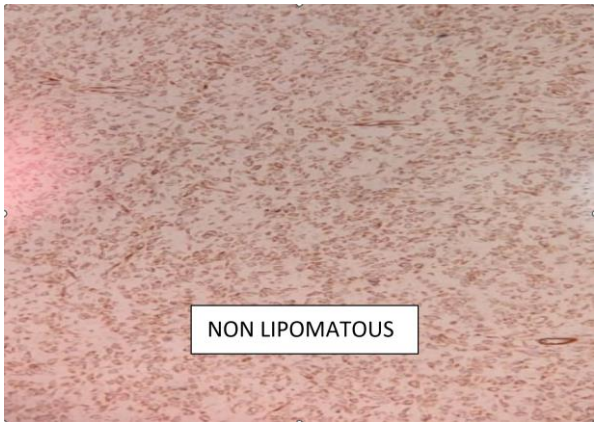

Figure 7: MDM2 positive for nuclear staining in nonlipogenic area. (x40)

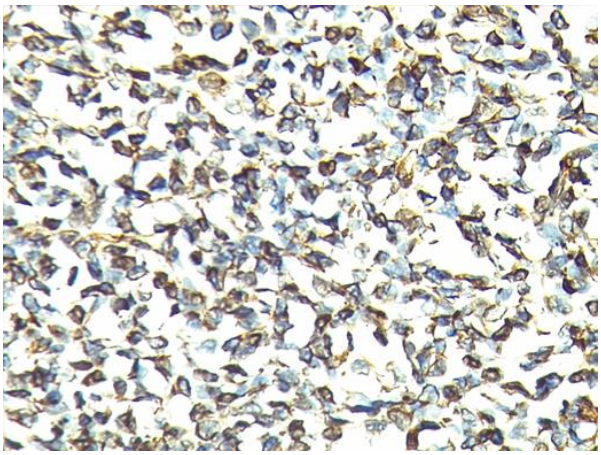

Figure 8: vimentin positive x40 for cytoplasmic staining in non-lipogenic area.

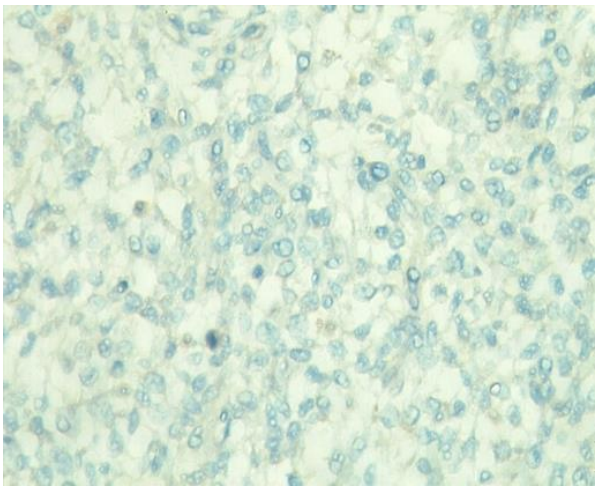

Figure 9: S-100 negative for nuclear staining

Surgical resection is traditionally the only potentially curative treatment. Histopathology is a diagnostic tool for dedifferentiated liposarcoma, and immunohistochemistry is mandatory for further confirmation and management of the tumor. Dedifferentiated liposarcomas that occur outside the mediastinum, trunk, and extremities are very rare. The rarity in this case is also due to the

heterologous component of dedifferentiated liposarcoma. There are only 10 previous papers available in the literature that address the radiological or pathological findings of dedifferentiated liposarcomas [5-8]. In our case, a 36-year-old male patient presented with a dedifferentiated liposarcoma with high-grade differentiation in the neck region, exhibiting local recurrence and morbidity due to its large size, which is one of the rare entities.

Conclusion

We are reporting a large dedifferentiated liposarcoma of the neck region with high-grade differentiation and local recurrence in a 36-year-old patient. This is a rare finding in the literature, as there are only a few case reports of dedifferentiated liposarcoma. In the present case study, histopathology supported the diagnosis of dedifferentiated liposarcoma, which is a focal outgrowth from the precursor of well-differentiated liposarcoma. The non-lipomatous area showed immunohistochemistry positivity for MDM2 and Vimentin and was negative for S100, which is very helpful for further confirmation and early detection and management of the tumor.

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Acknowledgements: None

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