

Clinical Outcome of Dedifferentiated Liposarcoma (DDLPS) in Extremities and Retroperitoneal Areas: A Case Series of 7 Patients

Salahuddin AL¹, Afif M. Yusof², Hau MA³

^{1,3}Department of Orthopaedic Hospital Raja Perempuan Zainab, Kota Bharu, Kelantan, Malaysia

²Department of Pathology Hospital Raja Perempuan Zainab, Kota Bharu, Kelantan, Malaysia

Abstract: This article explores the clinical outcomes and effectiveness of postoperative radiotherapy in the management of dedifferentiated liposarcoma (DDLPS) in extremities and retroperitoneal regions. DDLPS, a malignant adipocytic neoplasm, presents challenges due to its progression from atypical lipomatous tumour ALT / well - differentiated liposarcomas (WDLPS) to variable - histological grades. The study analyses 7 cases of DDLPS, 4 in extremities and 3 in retroperitoneal areas, focusing on factors such as tumour size, surgical margins, adjuvant therapy, and recurrence rates. In extremities DDLPS, postoperative radiotherapy shows promise for local control, especially with narrow surgical margins, while larger tumour sizes correlate with poorer outcomes. Retroperitoneal DDLPS cases with positive margins exhibit high recurrence rates and limited survival. These findings shed light on potential strategies for optimizing the management of DDLPS in different anatomical locations.

Keywords: dedifferentiated liposarcoma, extremities, retroperitoneal, postoperative radiotherapy, tumour size, surgical margin

1. Introduction

Dedifferentiated liposarcoma (DDLPS) is a rare, malignant adipocytic neoplasm characterized by progression from atypical lipomatous tumour (ALT) /well - differentiated liposarcoma (WDLPS) to a sarcoma of variable histological grades. DDLPS most commonly arises in the retroperitoneum, with extremity occurrence being rare. Wide surgical resection is the recommended primary treatment for DDLPS, aiming for complete excision with negative surgical margins. There are limited studies that have investigated the usefulness of adjuvant therapy (chemotherapy and radiotherapy) for DDLPS. This study's objective is to review the local recurrence rates of DDLPS in extremities and the retroperitoneum while examining their correlation with tumour size and surgical margin status, providing valuable insights to enhance treatment strategies for this challenging malignancy.

2. Methodology and Materials

All patients with liposarcoma of the extremities and retroperitoneal areas who had been treated between 2014 and 2020 were identified from the medical records. Out of the 34 cases of liposarcoma, 7 were diagnosed as DDLPS. The study assessed variables including the use of adjuvant therapy before and after surgery, tumour size, surgical margins, recurrence rates, metastatic rates, and oncological outcomes. Adjuvant therapy, both pre - and post - surgery,

was evaluated, tumour sizes were measured, surgical margins were recorded, and recurrence and metastatic rates were tracked. Subsequently, various oncological outcomes such as survival rates, progression - free survival, and overall quality of life were assessed.

3. Results

Details of 7 patients with DDLPS is listed in Table 1.

The present case series involved seven patients diagnosed with DDLPS, four patients with extremities DDLPS (two males and two females) had a mean age of 51.75 years and an average tumour size of 18 cm. Two of these patients had marginal margins and received post - operative radiotherapy, while the other two with wide inadequate margins did not experience recurrence or lung metastasis. One patient with local recurrence and lung metastasis passed away, and another underwent shoulder disarticulation. The remaining two patients with extremities DDLPS had no local recurrence or lung metastasis and remained disease - free after 4 years. On the other hand, the three male patients with retroperitoneal DDLPS had a mean age of 56 years and an average tumour size of 40 cm. All had intralesional margins, did not receive post - operative radiotherapy, and experienced recurrence within 6 months, with lung metastasis leading to their demise after 2 years of follow - up.

Table 1: Details of 7 patients with Dedifferentiated of Liposarcoma (DDLPS)

CN	Age	Sex	DOS	TOL	SM	TG	TS (cm)	L	Post Rx	Post Chem	LM	LR	Prognosis	F/Up
1	45	F	12	DDLPS	WIA	3	11x10	Right Leg	No	No	No	No	Disease free, 4 years follow - up	48
2	40	M	15	DDLPS	WIA	3	13x8	Right thigh	No	No	No	No	Disease free, 4 years follow - up	48
3	58	F	14	DDLPS	Mrg	3	20x15	Left Arm	Yes	Yes	Yes	Yes	Shoulder disarticulation after recurrence	48
4	64	M	18	DDLPS	Mrg	3	28x10	Left thigh	Yes	No	Yes	Yes	Patient died after 1 year of treatment	12
5	49	M	6	DDLPS	Intra	3	30x20	Abdomen	No	No	Yes	Yes	Patient died as disease progressed	48
6	57	M	6	DDLPS	Intra	3	60x40	Abdomen	No	No	Yes	Yes	Patient died as disease progressed	24
7	62	M	10	DDLPS	Intra	3	30x26	Abdomen	No	Yes	Yes	Yes	Patient died as disease progressed	24

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N: Case Number, Sex: M (Male), F (Female), DOS; Duration of symptoms (months), TOL: Types of Liposarcoma: Dedifferentiated Liposarcoma (DDLPS), TSM; Type of Surgical margin; (WIA: Wide Inadequate), (Intra: Intralesional), TS; Tumour Size, TG; Tumour Grade FNCLCC - Federation Nationale des Centres de Lutte Contre Le Cancer grade, L; Location, POST Rx: (radiotherapy after the operation), POST Chem (Chemotherapy after operation), LM; Lung Metastasis (detected with Ct Thorax performed after 6 months and later after diagnosed), LR: Local recurrence, F/Up; Total follow up months

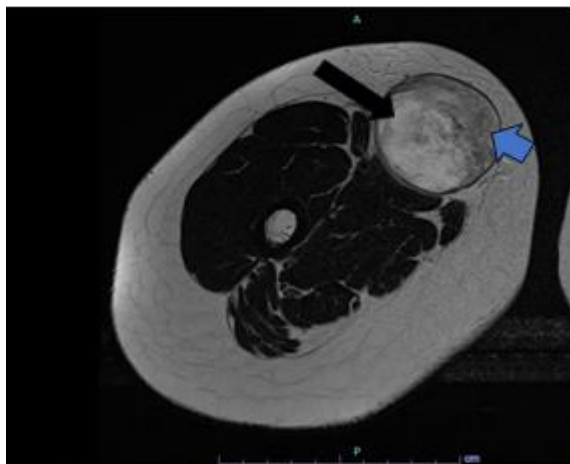


Figure 1: MRI T2 - weighted images of the right thigh. The ALT/WDLPS area shows high signal intensity (Black arrow). The dedifferentiated area shows low signal intensity (blue coloured arrow)

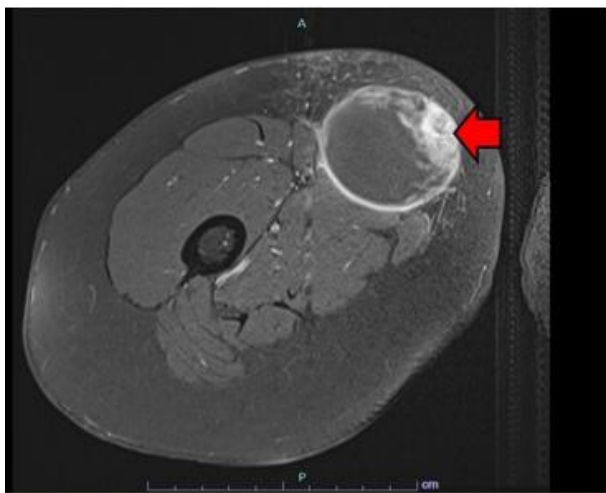


Figure 2: MRI T1 Fat suppressed postcontrast images of the right thigh. The dedifferentiated area is the area enhanced while the other area is fully suppressed (red coloured arrow)

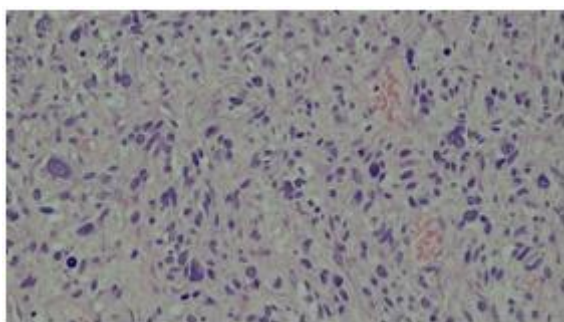


Figure 3: The histological slide shows cellular dedifferentiated area with cells having pleomorphic and enlarged bizarre nuclei in a myxocollagenous stroma. (Haematoxylin - eosin staining)

4. Discussions

In our case series, we administered postoperative radiotherapy to Case Number 3 and Case Number 4, as both of them had marginal margins in the surgical margin and experienced local recurrence. In this system, a resection margin is evaluated according to the distance of the margin from the tumour's reactive zone. We used the surgical margin as defined by JOA Japanese Orthopaedic Association (JOA). In the context of evaluating surgical margins, wide resection can be divided into wide adequate and wide inadequate. An adequate wide margin is characterized by a distance of 2 cm or more extending beyond the reactive zone, while an inadequate wide margin is defined as having only a 1 cm margin around the reactive zone. Additionally, a marginal margin is one that intersects the reactive zone itself. The minimum margin observed in the resected specimen is considered the surgical margin for the surgery, providing a crucial framework for determining the extent of excision in surgical procedures. (Kawaguchi, Ahmed et al.2004) . Many studies have mentioned that postoperative radiotherapy in extremities may provide effective local control for cases with a narrow surgical margin. (Gronchi, Lo Vullo et al.2009) . In our centre, we did not administer preoperative chemotherapy for DDLPS because its reported benefits are minimal, with response rates of $\leq 12\%$, and systemic therapy is infrequently used in both the primary and recurrent settings. (Jones, Fisher et al.2005, Italiano, Toulmonde et al.2012) . Our study reveals that the mean size of DDLPS is larger in retroperitoneal DDLPS cases compared to those occurring in the extremities. This finding is consistent with previous research, which has also suggested that larger tumour size is associated with poorer clinical outcomes. (Ghadimi, Al - Zaid et al.2011, Okada, Hasegawa et al.2011, Knebel, Lenze et al.2017) . The ALT/WDLPS and dedifferentiated areas within the tumour margin should be evaluated independently when determining the optimal extent of resection for DDLPS in the extremities. For the dedifferentiated area, it is recommended to perform resection with a wide margin. In situations where resection with an insufficiently wide margin or a marginal margin is unavoidable, additional postoperative radiotherapy may be considered to enhance local control. (Kito, Yoshimura et al.2016)

5. Conclusions

Postoperative radiotherapy in extremities may provide good local control for cases of DDLPS with a narrow surgical margin. The larger the size of the DDLPS tumor, the poorer the outcome.

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