

An Inflammatory Variant of Well-differentiated Liposarcoma of back - A Case Report on the Rare Entity

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Abstract

Lipoma is a slow-growing and usually harmless swelling of the subcutaneous tissue. They are rarely cancerous. They commonly occur in the neck, shoulder, back, abdomen, arms and thighs.

Liposarcoma is a rare tumour derived from fat tissue, which accounts for 2% of all tumours. The severity of Liposarcoma is based on the subtype and the presenting stage of the primary tumour. It can arise from any location of the body, but occurring in the back is rare.

This case report describes a case of an inflammatory variant of well-differentiated liposarcoma of the back presenting as Lipoma.

Keywords: liposarcoma; inflammatory well-differentiated liposarcoma; MDM2; Sarcoma

Abbreviations

MDM2 - Murine double minute 2.

LPS - Liposarcoma.

IWDLPS - Inflammatory Well-differentiated Liposarcoma.

OPD - Out-Patient Department.

FUS - Fused in sarcoma.

CHOP - C/EBP Homology Protein.

ALP - Atypical Lipomatous tumour.

DDLPS - Dedifferentiated Liposarcoma.

Fig - Figure.

Introduction

Liposarcomas are malignant tumours of mesenchymal origin (adipose tissue). The major sites of liposarcomas are the extremities, retroperitoneum and inguinal region. Liposarcomas are remarkable because of their frequently large sizes [17]. Virchow was the first to describe liposarcoma in the year 1860.

The development of liposarcoma from a preexisting benign lipoma is rare. Most cases arise de novo. Liposarcomas most frequently arise from the deep-seated stroma rather than the submucosal or subcutaneous fat. Dermal lesions are rare and may resemble pleomorphic fibroma [18].

The most recent World Health Organization classification of soft tissue tumours recognizes 5 categories of liposarcomas:

1. *well differentiated*, which includes the adipocytic, sclerosing, and inflammatory subtypes;
2. *dedifferentiated*;
3. *myxoid*;
4. *round cell*; (5) *pleomorphic*.

Case Report

A 44-Year-old homemaker came to General Surgery OPD at Sree Balaji Medical College & Hospital with midline swelling in the mid and lower back for the past 8 years. The patient was apparently normal before 8 years, following which she noticed a small swelling in the midline in the mid and lower back, which was insidious in onset and progressive in nature to attain the current size. Initially, the swelling was not associated with pain or restriction in the range of movement, but past 2 months patient complaints of pain in the lower back and being unable to bend forward. The patient noticed a sudden increase in size in the past 2 years.

No history of trauma, fever, cough with expectoration, evening rise of temperature or similar swelling elsewhere in the body.

The patient does not have a known case of Type 2 Diabetes Mellitus / Systemic Hypertension / Bronchial Asthma / Tuberculosis.

No significant spinal surgeries/surgeries for pilonidal sinus in the past.

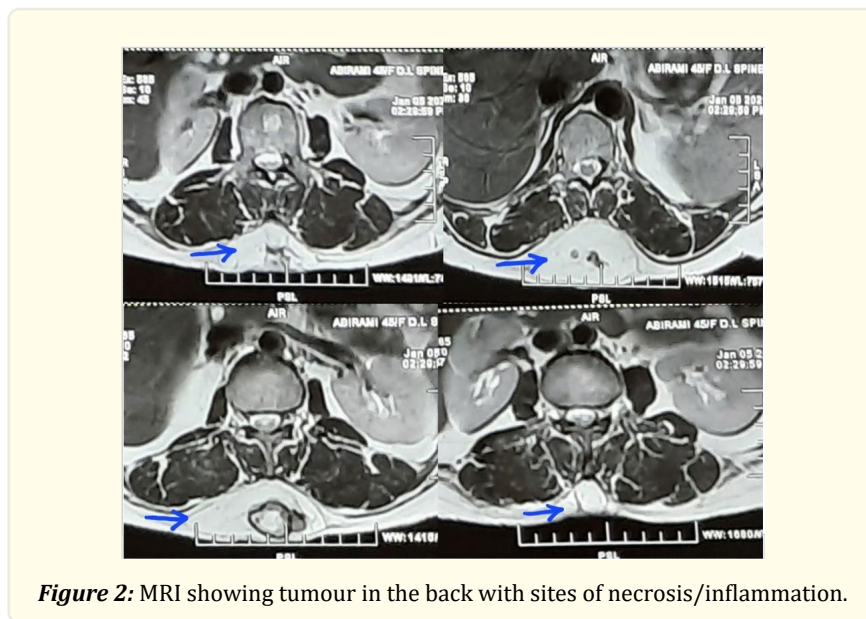
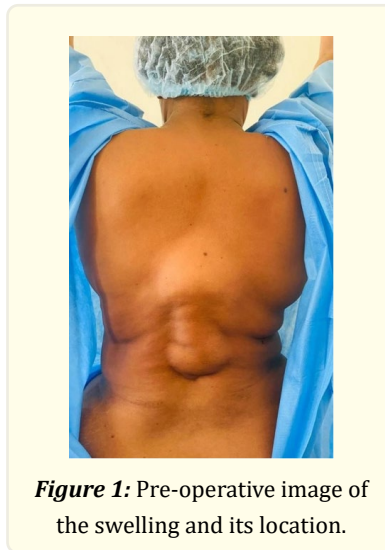
On Examination

A Large irregular-shaped swelling of size approximately 45*10cm is present over the midline in the mid and lower back. No transmitted pulsations, No superficial dilated or engorged veins, No hyperpigmentation or hypopigmentation, no scar or sinus over the swelling, and No signs of infection or inflammation over the swelling. Fluctuation is present. The surface is smooth, margins are not well defined, and variable in consistency. Preoperative image of the patient is given in Figure 1.

Meticulous radiological investigations were done to narrow down the possibilities. Ultra-sonogram of the superficial swelling showed a large hypoechoic swelling with minimal internal septations with internal vascularity. In the ultrasonogram, we could not rule out the spinal invasion.

MRI opted to rule out spinal cord involvement and the same was ruled out (Fig 2, 4).

With all these blind ends, we had Giant Lipoma and a variant of sarcomatous change in Lipoma as our working diagnosis. With this, we proceeded with a Trucut Biopsy of the swelling taking 5 biopsy bites and sending them for Histopathological evaluation. The result came out as Lipoma with Atypia. Keeping sarcoma in hindsight we planned and proceeded with the Wide Excision of the Swelling. Post-operative specimen image of our patient are given in Figure 5.



An encapsulated mass of size 48*15cm weighing 21kg without invasion of adjacent neurovascular structures with a pseudo capsule was found. The lesion was meticulously dissected from the closely associated neurovascular structure without injuring the adjacent neurovascular bundle. The specimen was removed in toto with a 3cm margin in all directions. The defect was closed with a Split skin graft.

Diagnosis

The specimen was sent for Histopathological examination to conclude the diagnostic dilemma. It came out as *Inflammatory well differentiated Liposarcoma*.

Discussion

IWDLPS shows a low metastatic potential, but it is a locally aggressive mesenchymal neoplasm. Tumour growth is slow, but if local excision is not complete, recurrence is likely to occur.

Inflammatory well-differentiated liposarcoma is high-grade, slow-growing tumour malignant neoplasms arising from adipocytic tissues characterized by a variable number of pleomorphic lipoblasts and the absence of areas of well-differentiated liposarcoma and other lines of differentiation. Inflammatory well-differentiated liposarcoma (IWDLPS) present in unusual locations; can cause diagnostic dilemmas [19].

There are three major subtypes of liposarcoma, each of which is characterized by its clinicopathologic and cytogenetic features. Well-differentiated/dedifferentiated liposarcoma can be considered to lie within the same spectrum because dedifferentiated liposarcoma represents a progression from low-grade, well-differentiated liposarcoma [2, 3]. Both are characterized cytogenetically by a ring or giant marker chromosome derived from 12q13-15 [4, 5], although additional cytogenetic aberrations are often detected in dedifferentiated variants [6]. Similarly, myxoid/round-cell liposarcoma can be considered along a histologic spectrum; round-cell liposarcoma is the poorly differentiated form of myxoid liposarcoma [7], and they share the characteristic t (12;16) [8].

IWDLPS is the most frequent subtype, in our patient the HPE is composed of mature adipocytes with a variation in cell size, focal nuclear atypia, and hyperchromatic, whereas sclerosing LPS exhibits a typical severe nuclear hyperchromasia and rare multivacuolated lipoblasts in an extensive collagenous stroma making it a differentiating feature. Finally, inflammatory ALT/WDLPS is a rare entity characterized by chronic inflammatory infiltration in which the adipocytic nature can be easily overlooked [14]. The HPE of IWDLPS of our patient is given in Figure 3.

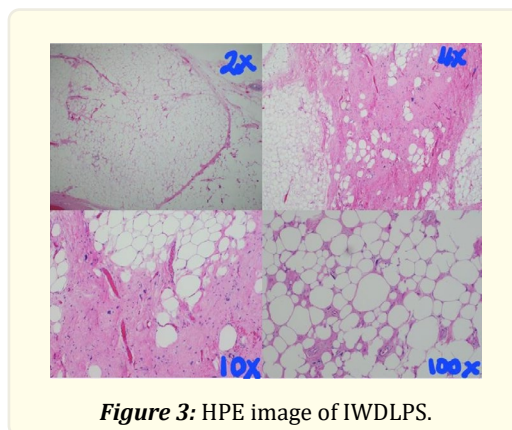


Figure 3: HPE image of IWDLPS.

In the case of IWDLPS overexpressing MDM2, cancer cells have another means to block p53. The sarcomas in which MDM2 amplification is a hallmark are well-differentiated liposarcoma/atypical lipomatous tumour, dedifferentiated liposarcoma, intimal sarcoma, and low-grade osteosarcoma [11]. MDM2 expression is detected by IHC and is a very sensitive tool in recognizing inflammatory well-differentiated liposarcoma [10].

MDM2 is the most frequent amplified oncogene in ALT/WDLPS and DDLPS, which also acts as a ubiquitin ligase, binding the transactivation domain of the tumour suppressor p53, thus promoting its degradation. This is confirmed by the fluorescence in situ hybridization analysis of MDM2 gene amplification, representing the standard differential diagnosis of ALT/WDLPS and DDLPS [15].

Liposarcoma is a lipogenic tumour of large deep-seated connective tissue spaces. Fusion proteins created by chromosomal abnormalities are key components of mesenchymal cancer development. An abnormality of band 12q13 has been associated with the development of liposarcomas. The most common chromosomal translocation is the *FUS-CHOP* fusion gene, which encodes a transcription factor necessary for adipocyte differentiation. These and other distinct genetic aberrations may aid in the diagnosis of particular liposarcoma subtypes, and they can potentially be targets that can be exploited therapeutically [13].

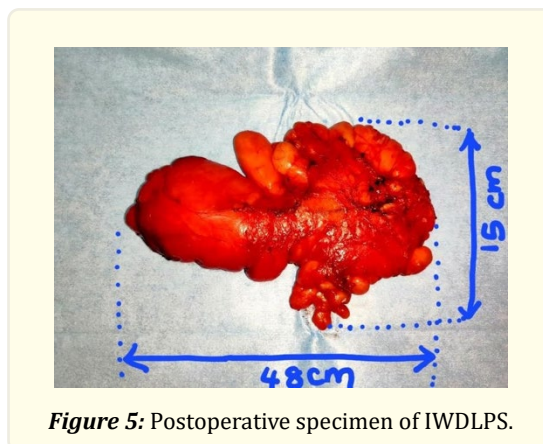
This case was followed up with IHC to narrow down the diagnosis and it came as MDM2 overexpression. The patient was started with Radiotherapy with 7mCu daily for 21 days with a period of rest for 7 days for 6 cycles. A 5-FDG PET CT was done to rule out metastasis.

The patient was currently in 6 Months follow-up for the past 1 year.



Conclusion

Since there are no studies to date on the role of surgery and RT focused specifically on the LPS histotype, In this case, report we have published the treatment protocol followed for our patient with an Inflammatory well-differentiated histotype of LPS.



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