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### Taking abdominal pain seriously: a case of aggressive dedifferentiated liposarcoma

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## INTRODUCTION

Liposarcomas (LPS) are malignant tumors of adipocytic differentiation. **Well-differentiated liposarcoma (WDL)**/atypical lipomatous tumor and **dedifferentiated liposarcoma (DDL)** together comprise the largest subgroup of liposarcomas. They occur most commonly in middle-aged to older adults typically within the retroperitoneum or proximal extremities. (1) Symptoms suspicious for liposarcoma include weight gain up to 10-20 lbs. Tumors on the arms or thighs might easily be noticeable, however, abdominal tumors can often go unnoticed. (2)

- **WDL**: does not metastasize, but can dedifferentiate to DDL
- **DDL**: more aggressive clinical behavior, with greater likelihood for 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 amplifications in chromosome 12q13-15, which includes *CDK4* and *MDM2* cell cycle oncogenes (3)

## CASE DESCRIPTION

64-year-old female with PMH of PE on Eliquis, s/p IVC filter presents with **abdominal discomfort, subjective weight gain** and **bilateral leg swelling**. Examination showed a distended abdomen and bilaterally tender edematous legs.

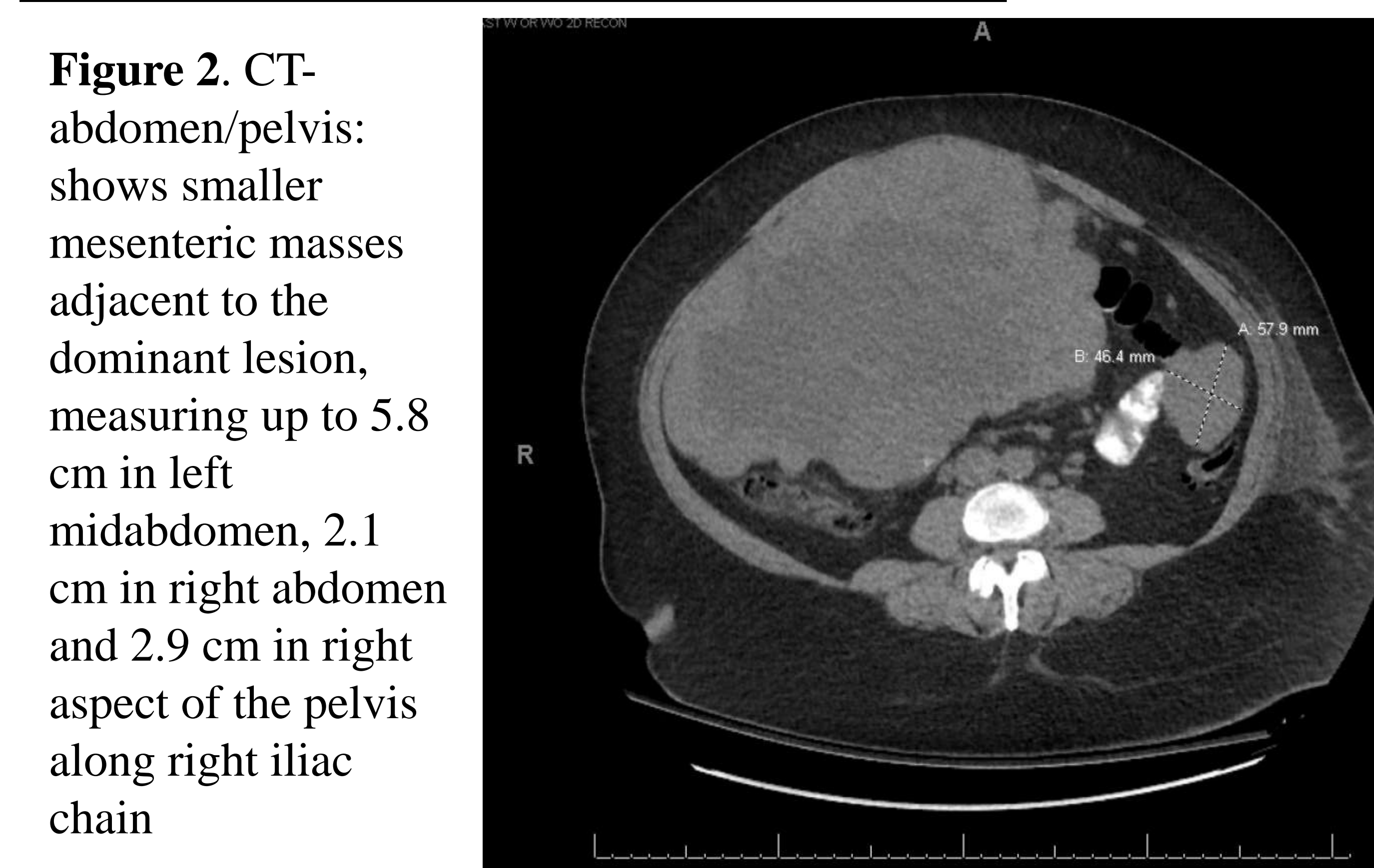
Doppler ultrasound of lower extremities showed extensive DVTs. Patient developed oliguria and AKI, ultrasound of kidneys was obtained, showing two large masses in abdomen and pelvis. CT abdomen/pelvis demonstrated **mesenteric mass measuring 26 cm** in right mid-abdomen with multiple smaller masses. IR biopsy showed malignant cells of **spindled and epithelioid features**. Molecular studies were positive for **MDM2 amplification**, with focal weak staining for CD10 and inhibin. Features were indicative of **dedifferentiated liposarcoma** given presence of *MDM2*.

Patient was **not a surgical candidate** but was a candidate for systemic chemotherapy and received one cycle of **Adriamycin, Ifosfamide, and Mesna (AIM)** for treatment of dedifferentiated liposarcoma; however, course was complicated by ifosfamide-induced encephalopathy. Patient received **palliative radiation**, and unfortunately, expired secondary to cardiac arrest from suspected pulmonary embolism.

## IMAGING



**Figure 1.** CT-abdomen/pelvis without contrast: demonstrated a 26 x 16 x 20 cm lobulated heterogeneous mesenteric mass centered in the right of midline in the mid-abdomen, confluent with small bowel loops

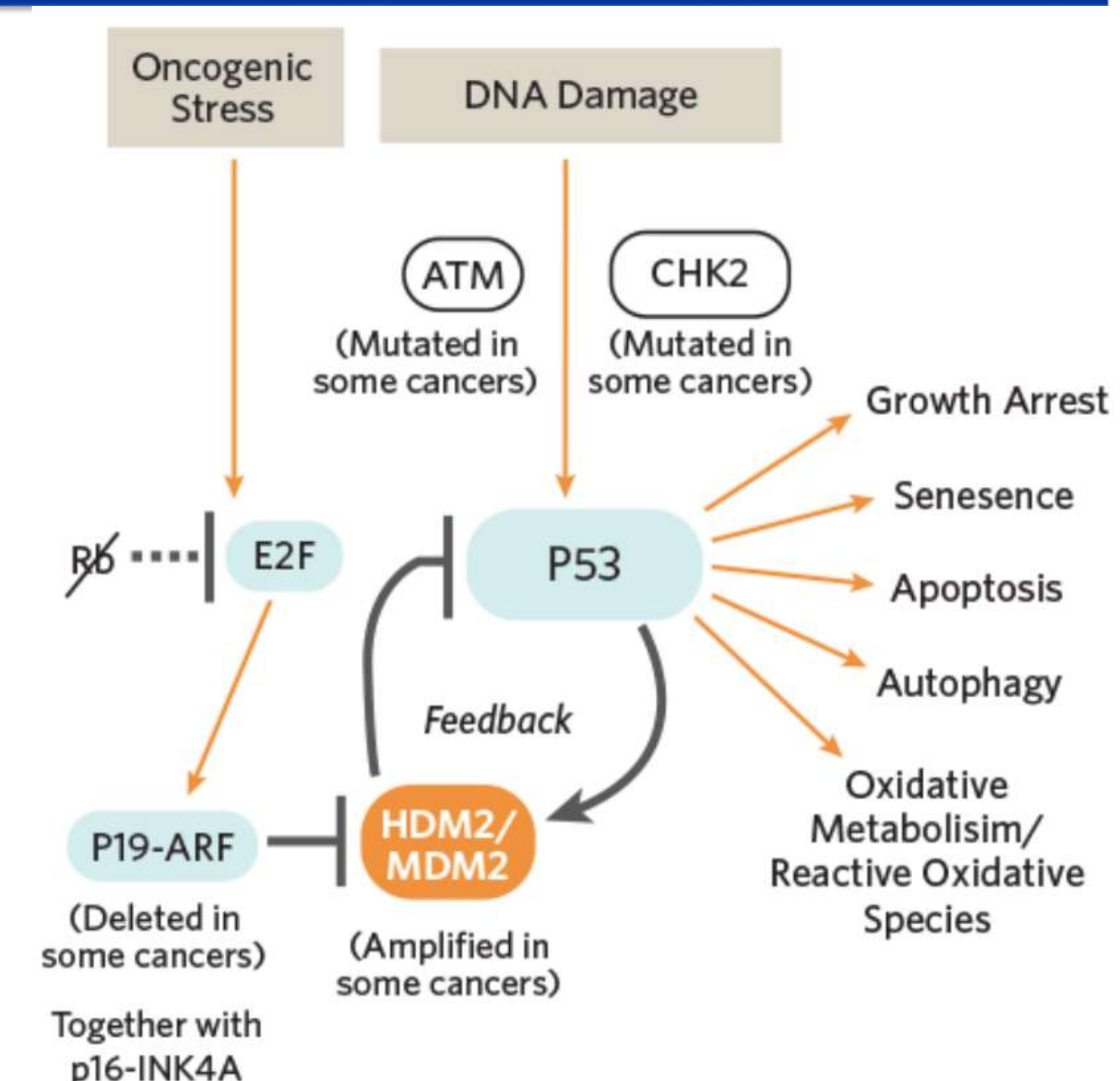


**Figure 2.** CT-abdomen/pelvis: shows smaller mesenteric masses adjacent to the dominant lesion, measuring up to 5.8 cm in left midabdomen, 2.1 cm in right abdomen and 2.9 cm in right aspect of the pelvis along right iliac chain

## PHYSIOLOGY

Gene amplification of *MDM2*, which binds and inactivates *TP53*, results in uncontrolled cell growth in response to cell damage and signals that typically trigger *TP53* to cause cell cycle arrest. (4)

*MDM2* is amplified in nearly 100% of patients, and *CDK4* is co-amplified in over 90% of patients.



## DISCUSSION

DDL tumors have six-fold the rate of local and metastatic recurrence and disease-specific mortality than that of WDL tumors (2)

Due to the rarity of these tumors and complexity of their treatment, approach to treatment of liposarcoma is multidisciplinary.

- Surgical resection is traditionally the only potentially curative option, typically indicated for small, low-grade and potentially resectable WDL tumors
- Perioperative radiotherapy has been reported to reduce local recurrence in retroperitoneal LPS (3)
- RT and/or chemotherapy is used to relieve symptoms in patients with unresectable local and/or metastatic LPS. Anthracycline-based therapy including doxorubicin with ifosfamide, given with mesna is preferred treatment (3,5).
- Investigational therapies inhibiting *MDM2* and *CKD4* (palbociclib, ribociclib), as well as immunotherapy (pembrolizumab) are currently being researched (2,3).

## CONCLUSIONS

- Signs and symptoms that can point a clinician towards a malignant liposarcoma include **abdominal discomfort** and **weight gain**; however, they are typically asymptomatic until they are large enough to compress or invade surrounding structures
- DDL is a focal outgrowth from precursor WDL lesions; therefore, **early detection of these tumors is key** to prompt diagnosis and management.

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