



HELLENIC REPUBLIC
**National and Kapodistrian
University of Athens**
———EST. 1837———

SCHOOL OF MEDICINE

**Sarcoma and Rare Cancers Unit
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**Monthly TARPSWG
International Tumor
Board**

Monthly TARPSWG International Tumor Board Case Presentation 27-10-2023

Update

61 yo male patient with retroperitoneal myxofibrosarcoma

- Back pain 2 months ago
- US: Soft tissue mass dislocating the right kidney
- CT CAP: Right retroperitoneal mass 12 x10.4 x10.1 cm in contact with psoas muscle and IVC dislocating the right kidney and the duodenum
- CT guided biopsy: spindle cell neoplasm with myxoid features CD34(+), Vimentin(+), EMA (+), CD99(+)
- Negative for IHC CDK4 and MDM2/FISH (-)
- MRI scan Right retroperitoneal tumour as previously described with fascia involvement of the quadratus lumborum.
- Recommendation: Treat it as myxoid fibrosarcoma

Treated as high grade myxofibrosarcoma with preop RT and perioperative CTX

02/2024 Partial response-tumour shrinkage

Plan for surgery

Monthly TARPSWG International Tumor Board Case Presentation 22-09-2023

Update

45 yo female patient with metastatic myxoid LPS

- **2014** Primary Right gluteal liposarcoma treated with pre-op radiotherapy and surgery R0
- **2017**: 2cm intraosseous lesion at the left iliac blade-biopsy Histology: Metastatic myxoid liposarcoma-Stereotactic RT
- **2017**: 3 month interval: New 1.5 cm bony lesion L5 surveillance
- **08/2018**: 3months increase of the lumbar lesion and 3 cm Breast lesion
2018: Chemotherapy DOXO +olaratumab
- **04/2019**: SD lumbar lesion+ PR breast lesion->Mastectomy +recon
- **08/2019**: local recurrence right gluteus-treated with surgery WLE
- **2020**:3month interval soft tissue metastasis at the Left thigh-> Surgery WLE
- **2021**: Adjuvant trabectedine 24 months
- **02/2022**: Distant metastasis right retroperitoneal space+right acetabulum ->Right nephrectomy + RT right thigh
- **06/2022**: adjuvant chemo 6months epirubicine stop due to neurotoxicity 09/23
- **06/2023**: Distant metastasis upper abdomen with retrohepatic vena cava involvement and presence of ascites at the lower abdomen → preop RT of the lesion at the upper abdomen
- **08/2023**: Partial Response of the large mass at the upper abdomen

Recommendation: Not for surgery due to inability to preform R0 resection

February 2023: Further response of the treated lesion at the upper abdomen(25 cm to 5.6 cms). New mass at the lower abdomen within the area of the previously reported ascites. Plan for RT

Monthly TARPSWG International Tumor Board Case Presentation 28-07-2023

Update

34 yo male patient with recurrent dedifferentiated myxoid LPS

- **2019** Primary Right Sided Retroperitoneal DDLPS-Surgery: **R2 resection**
- **Interval scans:** Residual disease increasing in size from 3 cms to 11 cms during an 8 months period and new lesions in the area of the surgical bed.
- **May 2020:** Surgery: Debulking removal of one mass 11 cms **R2 resection**
- Interval scan **July 2020** multiple lesions in the Right retroperitoneal area max diameter 11cms
- **August 2020 :Chemotherapy Doxorubicin/Ifosfamide**
- Partial response until PD in **April 2021**
- **July 2021** Debulking surgery **R2 resection**
- **Residual disease progressing until April 2022**
- **April 2022** Debulking surgery **R2 resection**
- **June2022: Chemotherapy Trabectedin**
- **June2022- March 2023: Partial response**
- **May 2023 RT: SD**
- **Scan 28/6/2023:** Right retroperitoneal DDLPS infiltrating the psoas muscle, obstructing the right kidney, dislocating the vena cava and the right iliac vessels with a WDLPS extending on the pelvis. Another WDLPS component on the contralateral side of the vena cava and another WDLPS on the left iliac fossa.
- **Recommendation:.** Discuss with the patient the option of clinical trial or palliative chemotherapy or surgery with the morbidity and mortality risks and not with curative intend.
- No clinical trial with anti-MDM2 regimen in Greece was running. In a new scan at 11/2023 PD of the DDLPS. Discussed surgery

December 2023: Compartmental surgical resection of the DD-WD right sided mass and R1 resection of the WD pelvic mass and WD left paracaval mass

3month interval scan March 2024 scan: No evidence of local or distant recurrence