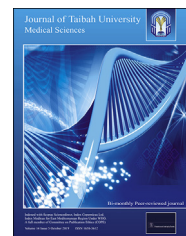




# Taibah University

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### Case Report

## Giant retroperitoneal liposarcoma: A case report and literature review



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### المخلص

الساركومات الشحمية خلف الصفاق مجموعة غير متجانسة من أورام اللحمية المتوسطة. لهذه الأورام تنوع واسع من الأنصاف الفرعية النسيجية التي تظهر بأعراض غامضة. نستعرض رجلاً يبلغ من العمر ٧٥ عاماً مصاباً بفقدان الشهية وفقدان الوزن بلغ ٧ كجم مع محيط بطني متزايد خلال الأسابيع الستة الماضية. أظهر التصوير المقطعي المحوسب للبطن والحوض كتلة كبيرة ملأت كامل البطن تقريباً. بعد التشاور مع مجلس الأورام متعدد التخصصات، تم إجراء عملية استئصال كاملة للورم. بالإضافة إلى ذلك، تمت إزالة الكلية اليسرى وجزء من الحجاب الحاجز الأيسر. كانت قياسات الورم ٢٠.٥٣٥ x ٢٩ x ١١.٦ بسم ووزنه ١١.٦ كجم. أكد التقرير النسيجي أنها ساركومة شحمية ذات درجة تباين متدنية مع قلة من الخلايا الدهنية غير الطبيعية والخلايا المنتجة للدهون، مع وجود مكونات الخلايا المغزلية والمتعددة الأشكال والغضروفية. تعافى المريض دون مضاعفات وبقي مستقراً أثناء زيارة المتابعة. نبغ عن هذه الحالة كي يتخذ غير المبالي تدابير جراحية استثنائية في التعامل مع مثل هذه الأورام الصلبة أثناء العملية الجراحية، حيث يصعب التعامل مع هذه الأورام الصلبة بسبب الغزو الموضعي والمجاور الذي يستلزم عادة الاستئصال الكامل.

**الكلمات المفتاحية:** ساركومة دهنية؛ الساركومة العملاقة خلف الصفاق؛ كتلة بطنية؛ فريق متعدد التخصصات؛ جراحة

### Abstract

Retroperitoneal liposarcomas are a heterogeneous group of mesenchymal tumours that have a wide spectrum of histological subtypes and vague clinical presentations.

Herein, we present the case of a 75-year-old man with anorexia, weight loss of 7 kg, and a growing abdominal circumference within a span of 6 weeks. Computed tomography of the abdomen and pelvis showed a large mass that filled almost the entire abdominal cavity. After consultation with a multidisciplinary tumour board, en bloc resection of the tumour was performed. In addition, the left kidney and a part of the left diaphragm were removed. The tumour measured 35 × 29 × 20.5 cm and weighed 11.6 kg. The histological report confirmed low-grade dedifferentiated liposarcoma with scarce atypical adipocytes, lipoblasts containing spindle cell, pleomorphic, and chondroid components. The patient had uneventful recovery and remained stable during the follow-up period. We report this case to highlight the need for customized surgical oncological measures in the treatment of solid abdominal tumours due to locoregional invasion that usually necessitates en bloc resection.

**Keywords:** Abdominal mass; Giant retroperitoneal sarcoma; Liposarcoma; Multidisciplinary team; Surgery

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### Introduction

Liposarcomas are rare malignant tumours that commonly originate in the thigh or retroperitoneum.<sup>1</sup> Their worldwide incidence varies between 0.07% and 0.2% of all retroperitoneal tumours.<sup>2</sup> The estimated incidence of liposarcoma in Europe is 4–5/100,000/year; however, in

Germany, these are amongst the most common malignant soft tissue tumours in adults.<sup>3,4</sup> Retroperitoneal liposarcomas (RPSs) present at a median age of 56 years,<sup>5</sup> but they have been reported to occur at all ages (2–98 years).<sup>6</sup> The gender distribution of RPSs is approximately equal, although some studies have reported female predominance.<sup>7</sup>

The European Society for Medical Oncology (ESMO) has identified certain predisposing factors for RPS: genetic aberrations, ionizing radiation exposure, particularly in children undergoing repeated computed tomography (CT) and those with diabetes mellitus.<sup>8</sup> Approximately 20%–25% of the patients with RPS have been reported to have diabetes mellitus; the co-existence of these two diseases has been associated with adverse prognosis.<sup>9</sup> Interestingly, most RPSs rarely metastasise, even when they grow to a large size.<sup>10</sup> According to the ESMO, although they are malignant lesions, only about 10% of these tumours are found to have metastasised to distant organs, particularly the lungs and liver, through haematogenous spread.<sup>11</sup> The pathological diagnosis of RPSs should include morphology, molecular pathology, and immunohistochemistry. RPSs are broadly classified into four histologic subtypes: well-differentiated, myxoid, pleomorphic, and dedifferentiated. Dalal et al. have found that the well-differentiated subtype is the most common variant of RPS (46%), followed by the myxoid (28%), dedifferentiated (18%), and pleomorphic subtypes (8%).<sup>12</sup>

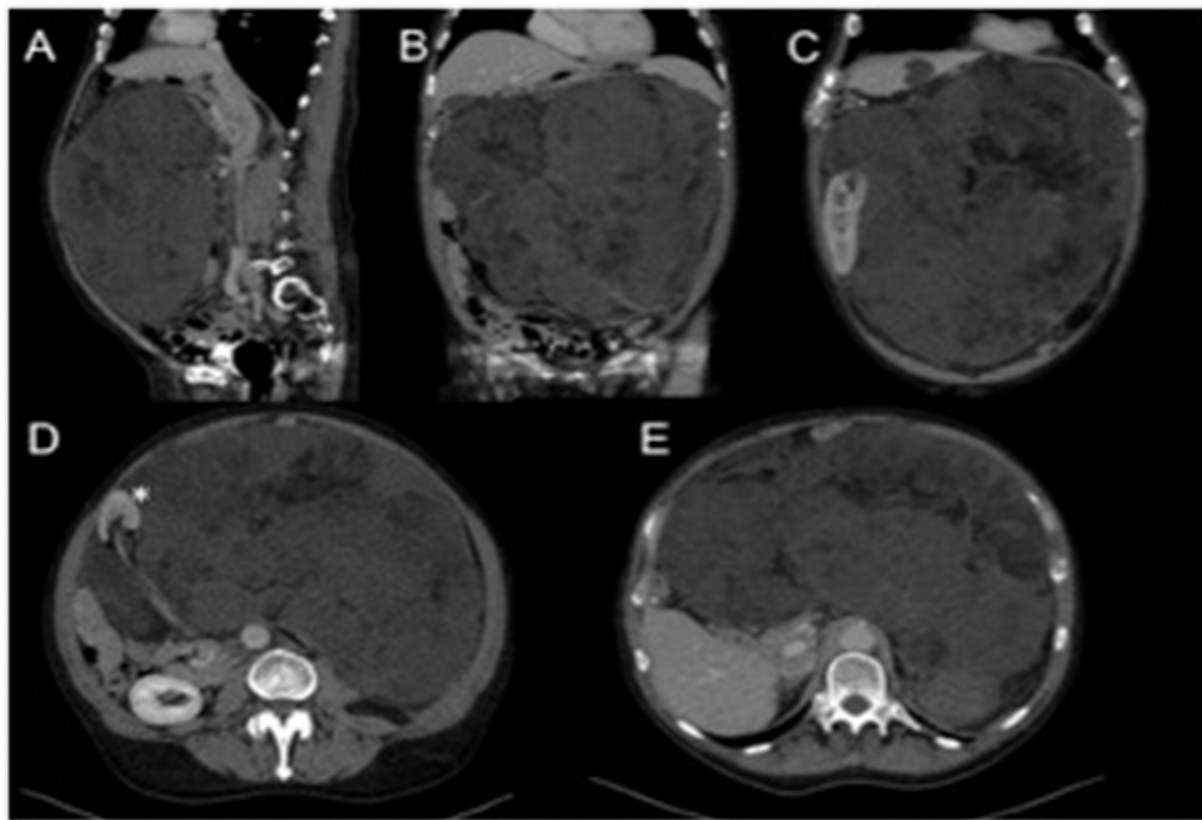
As RPSs grow in an expandable space, they attain a large size before being discovered as a palpable lump or as the

cause of increasing abdominal girth or gastrointestinal, urologic, or neurological compression symptoms. This is in sharp contrast with hollow viscus malignancies which manifest relatively during to luminal obstruction or intra-luminal bleeding.<sup>13,14</sup> Approximately 60%–80% of the patients present with a palpable abdominal mass and half of the patients have vague abdominal pain at presentation.<sup>15</sup> RPS is probably one of the largest neoplasms in the human body, invariably measuring >5 cm when diagnosed.<sup>5</sup>

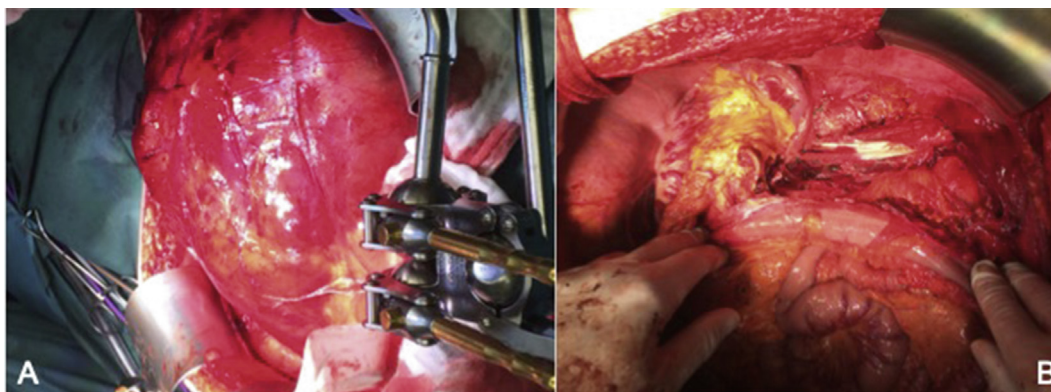
Herein, we report the successful management of a giant RPS, weighing 11.6 kg, in a 75-year-old man, at the Department of Surgery of the Hospital Reinbek St. Adolf-Stift, Germany. We aim to convey an important message to physicians to keep a high index of suspicion for RPS in patients presenting with vague abdominal symptoms. We also provide an update on the diagnostic and management guidelines for RPS.

### Case report

A 75-year-old man presented to the Surgical Clinic of the Department of Surgery of the Hospital Reinbek St. Adolf-Stift, Germany, with complaints of rapid weight loss of 7 kg, reduced appetite, and a growing abdominal circumference within the previous 6 weeks. The patient also experienced intermittent constipation. He had a history of bilateral hip arthroplasty due to arthrosis and a left-sided inguinal hernia repair. Physical examination revealed a non-tender, distended abdomen with a palpable mass extending from the epigastrium to the pelvic region. Routine



**Figure 1:** Preoperative CT scan of the giant liposarcoma. Sagittal (A), coronal (B, C), and axial images (D, E); \*left kidney.



**Figure 2:** Perioperative illustrations showing the tumour's gross characteristics at initial surgical exposure (A) and deeper dissection (B).

biochemical parameters were within normal limits. Abdominal ultrasound revealed a heterogeneous soft tissue mass that filled nearly the entire abdominal cavity. CT of the abdomen showed a giant heterogeneous mass, measuring  $31 \times 23 \times 30$  cm, leading to the displacement of the left kidney, descending colon, the small bowel loops, and the mesenteric axis (Figure 1). No ascites, locoregional or distant metastases, or calcifications were observed. Staging was performed using endoscopy and the findings were unremarkable.

In consultation with our multidisciplinary tumour board, it was decided that exploration and surgical resection of the tumour be performed. Laparotomy was performed using the midline approach; an enormous, lobulated, retroperitoneal mass presented (Figure 2). After resection of the tumour mass, the left kidney was also removed as it appeared to be infiltrated by the tumour. Furthermore, multiple nodular lesions were observed on the left hemi-diaphragm. Partial

diaphragmatic resection was performed with subsequent reconstruction.

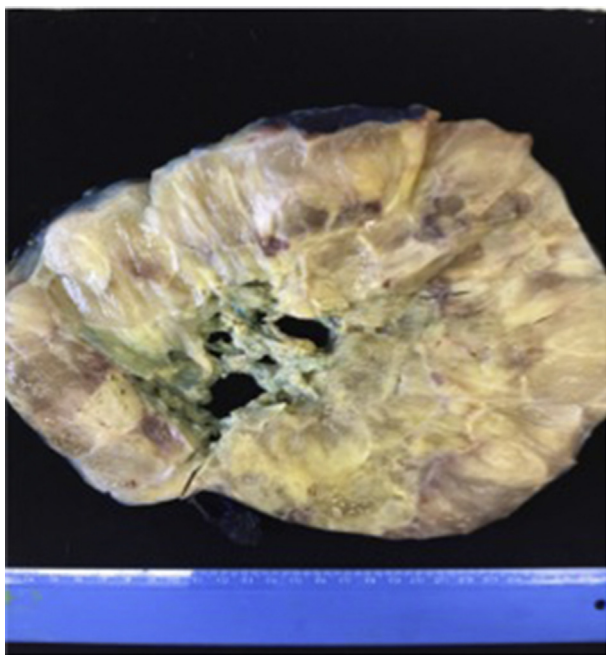
On macroscopic examination, the tumour's dimensions were  $35 \times 29 \times 20.5$  cm and it weighed 11,685 g. The cut section showed a yellow, lobulated tumour with a large, central necrotic area of  $19 \times 10 \times 25$  cm, comprising approximately 70% of the tumour mass (Figure 3). The attached diaphragm and muscles were infiltrated by the tumour. Examination of the left kidney revealed infiltration of the capsule without any infiltration into the parenchyma. Histopathological examination revealed a lipomatous tumour with a low-grade, dedifferentiated liposarcoma. Apart from well-differentiated areas with only scarce atypical adipocytes and lipoblasts, regions with spindle cell, pleomorphic, or myxoid/chondroid components were also observed (Figure 4). Mitotic activity was low, with a Ki67 proliferating index of 5%–10%. The tumour showed typical *MDM2* gene amplification.

The patient had an uneventful recovery and was discharged on the twelfth postoperative day. His follow-up plan consisted of visits every 3 months for 2 years. During the short-term follow-up, the patient remained free of pain, and his physical condition improved. Additionally, the physical examination and abdominal ultrasound were unremarkable.

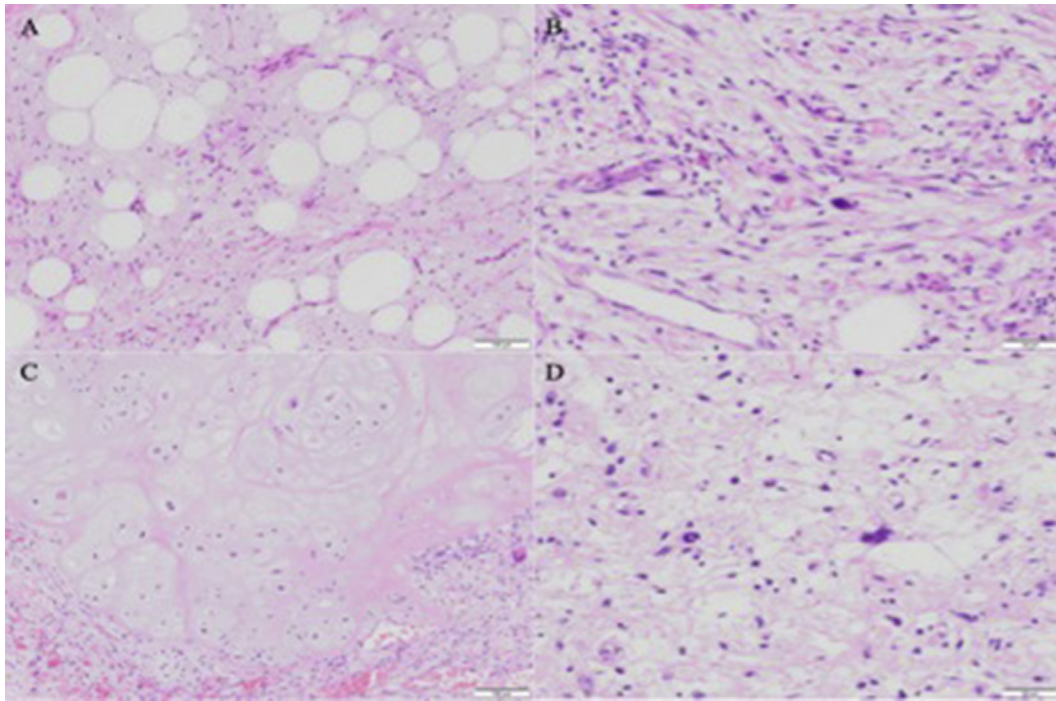
## Discussion

We present a case of an exceptionally large RPS which was diagnosed by abdominal CT and was successfully removed. The tumour had infiltrated into the adjacent left kidney and hemi-diaphragm, which necessitated nephrectomy and partial excision of the diaphragm. Postoperatively, the patient had uneventful recovery. Our case is similar to other reported cases in terms of non-specific presentation<sup>16</sup>; however, an RPS with such enormous weight and dimensions has not been previously reported.

According to the Transatlantic Retroperitoneal Sarcoma Working Group (TARPSWG), abdominal ultrasound can be the initial imaging test, but CT or magnetic resonance imaging should be performed in all cases.<sup>17</sup> The TARPSWG has emphasized the value of collaboration among various disciplines in the treatment of RPSs. Thus, multidisciplinary team work involving surgeons, pathologists, imaging specialists, radiation therapists, and



**Figure 3:** Macroscopic examination of the tumour with a central necrotic area.



**Figure 4:** Histological examination of the liposarcoma. Atypical lipomatous areas (A); spindle cell and pleomorphic areas (B); chondroid differentiation (C); pleomorphic cells with Touton giant cells and lipoblasts (D).

medical oncologists is recommended. In our case, CT of the abdomen and pelvis was helpful in diagnosing RPS. Thereafter, the case was managed under the guidance of the multidisciplinary tumour board of the hospital.

Percutaneous biopsy is recommended prior to surgery for suspected RPS; it can also guide the surgical approach and follow-up protocol.<sup>18</sup> According to the ESMO recommendations, following an appropriate imaging evaluation, the standard approach to RPS diagnosis includes multiple core-needle biopsies using  $\geq 14$ –16 G needles. However, such biopsies could also jeopardize the accurate estimation of malignancy in large RPSs. In our case, neither fine-needle nor core-needle biopsy was performed prior to surgery.

Complete surgical resection is the gold standard in the treatment of RPSs.<sup>19</sup> There are no size restrictions that would contraindicate a surgical resection. Our case also validates the observations that even enormous RPSs may be surgically resected under the guidance of a multidisciplinary team, which is consistent with the consensus management of liposarcomas in adults.<sup>20</sup> The significance of an R0 resection was first demonstrated by Lewis et al. in 1988.<sup>21</sup> In their cohort of 500 patients with RPS, the median survival after an R0 resection was 103 months compared to 18 months after an R1 or R2 resection. In a review by Zeng et al., the long-term 5- and 10-year survival rates without complete resection were described as 16.7% and 8.0%, respectively.<sup>22</sup> This reaffirms the curative role of surgical excision of RPS. In the era of evolving surgical innovations and technologies, surgical education and training courses,<sup>23,24</sup> particularly on live animals and high-fidelity simulation models,<sup>25,26</sup>

provide a valuable support in upskilling the trainees and established surgeons.

In cases where the RPS is associated with non-resectable vital organs or increased morbidity and mortality is anticipated in association with total resection, preoperative radiotherapy and/or chemotherapy may be considered to downstage the tumour.<sup>27</sup> In cases of massive tumour dimensions, due to the resulting complexity of the operation, an open approach is recommended.

Some studies have advocated the value of radiotherapy in addition to surgical resection. Lee et al. reported a local recurrence rate of 38% after additional radiotherapy versus 48% without additional radiotherapy in a cohort of 77 patients.<sup>28</sup> However, there was no statistically significant benefit with the addition of radiotherapy. Conversely, in a retrospective analysis, Ecker et al. described an improved survival in patients undergoing neoadjuvant radiotherapy.<sup>29</sup> The authors reported an improved overall survival of 129.2 months versus 84.3 months in patients treated with radiotherapy, particularly in cases of tumours with organ invasion. There is currently no randomized controlled study for the usefulness of radiotherapy or chemotherapy in RPSs that would aid the decision of management. In our case, the patient did not undergo any multimodal therapy.

Due to the wide spectrum of histological subtypes with different biological behaviours, a standard recurrence rate of RPSs cannot be determined. In a review by McCallum et al., the researchers investigated the correlation of the local recurrence and distant metastasis rates with tumour size.<sup>30</sup> The estimated 5-year metastasis rate in tumours  $< 2.5$  cm

was approximately 3%, whereas in tumours  $\geq 20$  cm, the rate was up to 60%. Recently, neoadjuvant chemotherapy as a radiosensitizer in combination with radiotherapy has been described. In the recently reported phase I/II clinical trial, researchers investigated the long-term outcomes of 83 patients with RPS who underwent three cycles of ifosfamide along with radiotherapy before surgical excision of the tumour.<sup>31</sup> This trial endorsed the effectiveness of a combination therapy; however, the long-term outcomes of the treated patients remained poor. Local recurrence of RPSs, particularly within the first 6 months to 2 years, was found to be more common than distant metastasis.<sup>32</sup>

The consensus guidelines by the TARPSWG for the management of recurrent RPS recommend a careful review of the preoperative imaging, operative notes, and histological findings by a multidisciplinary team.<sup>20</sup> Unfortunately, the outcome and prognosis deteriorate with each surgical resection with a concomitant increase in morbidity.<sup>33</sup> Thus, oncologists should weigh the risks and benefits when managing recurrent RPSs. The prognosis of dedifferentiated liposarcomas is better than that of the pleomorphic type, but worse than that of the atypical lipomatous tumours/well-differentiated liposarcoma.<sup>16</sup> These variations are influenced by the different biological behaviours of liposarcomas.

## Conclusion

Our case suggests that dedifferentiated liposarcomas should be treated by radical surgical excision, with multi-visceral resection if required. A high index of suspicion for RPS should be maintained in patients presenting with non-specific abdominal symptoms. A multidisciplinary tumour board should be consulted before embarking on any intervention. Surgery remains the cornerstone of curative therapy for intraabdominal liposarcomas.

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## Conflict of interest

The authors have no conflict of interest to declare.

## Ethical approval

The authors declare, that the involved patient gave his informed consent for participation in research. The study was done according to the declaration of Helsinki.

## Authors contributions

JH conceived and designed the study, conducted research, provided research materials, and collected and organized, analyzed and interpreted data. KN and HH performed surgery and postoperative follow-up and collected data. KHU performed histopathology examination and provided research materials. SYG and TS wrote final draft of article, and provided logistic support. All authors have critically

reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

## References

1. Lechner M, Borhanian K, Mitterwallner S, Bittner R, Klieser E, Köhler G, et al. Retroperitoneal liposarcoma: a concern in inguinal hernia repair. *JSLs J Soc Laparoendosc Surg* 2019; 23(1).
2. Ioannidis A, Koutserimpas C, Konstantinidis M, Drikos I, Voulgaris P, Economou N. Dyspnea caused by a giant retroperitoneal liposarcoma: a case report. *Oncol Lett* 2018; 16(2): 1539–1542.
3. Matthyssens LE, Creytens D, Ceelen WP. Retroperitoneal liposarcoma: current insights in diagnosis and treatment. *Front Surg* 2015; 2: 4.
4. Rensing M, Wardelmann E, Hohenberger P, Jakob J, Kasper B, Emrich K, et al. Strengthening health data on a rare and heterogeneous disease: sarcoma incidence and histological subtypes in Germany. *BMC Public Health* 2018; 18(1): 235.
5. Stoeckle E, Coindre JM, Bonvalot S, Kantor G, Terrier P, Bonichon F, et al. Prognostic factors in retroperitoneal sarcoma: a multivariate analysis of a series of 165 patients of the French Cancer Center Federation Sarcoma Group. *Cancer* 2001; 92(2): 359–368.
6. Bremjit PJ, Jones RL, Chai X, Kane G, Rodler ET, Loggers ET, et al. A contemporary large single-institution evaluation of resected retroperitoneal sarcoma. *Ann Surg Oncol* 2014; 21(7): 2150–2158.
7. Gronchi A, Miceli R, Shurell E, Eilber FC, Eilber FR, Anaya DA, et al. Outcome prediction in primary resected retroperitoneal soft tissue sarcoma: histology-specific overall survival and disease-free survival nomograms built on major sarcoma center data sets. *J Clin Oncol* 2013; 31(13): 1649–1655.
8. Krille L, Zeeb H, Jahnen A, Mildnerberger P, Seidenbusch M, Schneider K, et al. Computed tomographies and cancer risk in children: a literature overview of CT practices, risk estimations and an epidemiologic cohort study proposal. *Radiat Environ Biophys* 2012; 51(2): 103–111.
9. Casali P, Abecassis N, Bauer S, Biagini R, Bielack S, Bonvalot S, et al. Soft tissue and visceral sarcomas: ESMO–EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2018; 29(Supplement\_4): iv51–iv67.
10. Liles JS, Tzeng C-WD, Short JJ, Kulesza P, Heslin MJ. Retroperitoneal and intra-abdominal sarcoma. *Curr Probl Surg* 2009; 6(46): 445–503.
11. Group EESNW. Soft tissue and visceral sarcomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2014; 25(suppl\_3): iii102–iii12.
12. Dalal KM, Kattan MW, Antonescu CR, Brennan MF, Singer S. Subtype specific prognostic nomogram for patients with primary liposarcoma of the retroperitoneum, extremity, or trunk. *Ann Surg* 2006; 244(3): 381.
13. Guraya SY, Al Naami M, Al Tuwaijri T, Arafah MMI. Malignant melanoma of the small bowel with unknown primary: a case report. *J Ayub Med Coll Abbottabad* 2007; 19(1): 63–65.
14. Guraya SY. Malignant duodeno-colic fistula: a rare complication of colorectal cancer. *J Clin Diagn Res JCDR* 2015; 9(11): PL01.
15. Lehnert T, Cardona S, Hinz U, Willeke F, Mechttersheimer G, Treiber M, et al. Primary and locally recurrent retroperitoneal soft-tissue sarcoma: local control and survival. *Eur J Surg Oncol* 2009; 35(9): 986–993.
16. Nizri E, Fiore M, Barretta F, Colombo C, Radaelli S, Callegaro D, et al. Intraperitoneal invasion of retroperitoneal

- sarcomas: a risk factor for dismal prognosis. *Ann Surg Oncol* **2019**; 1–7.
17. van Houdt WJ, Raut CP, Bonvalot S, Swallow CJ, Haas R, Gronchi A. New research strategies in retroperitoneal sarcoma. The case of TARPSWG, STRASS and RESAR: making progress through collaboration. *Curr Opin Oncol* **2019**; 31(4): 310–316.
  18. Almond L, Tirotta F, Tattersall H, Hodson J, Cascella T, Barisella M, et al. Diagnostic accuracy of percutaneous biopsy in retroperitoneal sarcoma. *Br J Surg* **2019**; 106(4): 395–403.
  19. Singer S, Antonescu CR, Riedel E, Brennan MF. Histologic subtype and margin of resection predict pattern of recurrence and survival for retroperitoneal liposarcoma. *Ann Surg* **2003**; 238(3): 358.
  20. Group T-ARW. Management of primary retroperitoneal sarcoma (RPS) in the adult: a consensus approach from the Trans-Atlantic RPS Working Group. *Ann Surg Oncol* **2015**; 22(1): 256–263.
  21. Lewis JJ, Leung D, Woodruff JM, Brennan MF. Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution. *Ann Surg* **1998**; 228(3): 355.
  22. Zeng X, Liu W, Wu X, Gao J, Zhang P, Shuai X, et al. Clinicopathological characteristics and experience in the treatment of giant retroperitoneal liposarcoma: a case report and review of the literature. *Cancer Biol Ther* **2017**; 18(9): 660–665.
  23. Guraya SY. The changing landscape of surgical education and training. *J Musculoskelet Surg Res* **2018**; 2(1): 1.
  24. Khalid M. Educational leadership and choice of postgraduate surgical training. *J Musculoskelet Surg Res* **2019**; 3(2): 177.
  25. Forgione A, Kislov V, Guraya SY, Kasakevich E, Pugliese R. Safe introduction of laparoscopic colorectal surgery even in remote areas of the world: the value of a comprehensive tele-mentoring training program. *J Laparoendosc Adv Surg Tech* **2015**; 25(1): 37–42.
  26. Guraya SY, Forgione A, Sampogna G, Pugliese R. The mapping of preferred resources for surgical education: perceptions of surgical trainees at the advanced international minimally invasive surgery academy (AIMS), Milan, Italy. *J Taibah Univ Med Sci* **2015**; 10(4): 396–404.
  27. Singer S, Corson JM, Demetri GD, Healey EA, Marcus K, Eberlein TJ. Prognostic factors predictive of survival for truncal and retroperitoneal soft-tissue sarcoma. *Ann Surg* **1995**; 221(2): 185.
  28. Lee HS, Yu JI, Do Hoon, Lim SJK. Retroperitoneal liposarcoma: the role of adjuvant radiation therapy and the prognostic factors. *Radiat Oncol J* **2016**; 34(3): 216.
  29. Ecker B, Peters M, McMillan M, Sinnamon A, Zhang P, Fraker D, et al. Preoperative radiotherapy in the management of retroperitoneal liposarcoma. *Br J Surg* **2016**; 103(13): 1839–1846.
  30. McCallum OJ, Burke II JJ, Childs AJ, Ferro A, Gallup DG. Retroperitoneal liposarcoma weighing over one hundred pounds with review of the literature. *Gynecol Oncol* **2006**; 103(3): 1152–1154.
  31. Gronchi A, De Paoli A, Dani C, Merlo DF, Quagliuolo V, Grignani G, et al. Preoperative chemo-radiation therapy for localised retroperitoneal sarcoma: a phase I–II study from the Italian Sarcoma Group. *Eur J Cancer* **2014**; 50(4): 784–792.
  32. Raut CP, Callegaro D, Miceli R, Barretta F, Rutkowski P, Blay J-Y, et al. Predicting survival in patients undergoing resection for locally recurrent retroperitoneal sarcoma: a study and novel nomogram from TARPSWG. *Clin Cancer Res* **2019**; 25(8): 2664–2671.
  33. Neuhaus S, Barry P, Clark M, Hayes A, Fisher C, Thomas J. Surgical management of primary and recurrent retroperitoneal liposarcoma. *Br J Surg* **2005**; 92(2): 246–252.

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