

A Huge Pleomorphic Liposarcoma of the Retroperitoneum

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ABSTRACT Retroperitoneal liposarcoma (RLS) is a rare, highly malignant tumor with poor prognosis and high recurrence rate. Pleomorphic liposarcoma is the rarest subtype and is a high grade tumor with cells with very different appearance from normal cells. Herein we report a case of a 68 year-old man with a 35 cm mass adjacent to kidney with synchronous inguinal 7 cm mass. The patient was operated in our clinic and pathology revealed pleomorphic liposarcoma. The case was discussed because of its rarity and two mass in the two distinct region.

Keywords: Retroperitoneal liposarcoma; liposarcoma; nephrectomy

Soft tissue sarcomas (STS) are rare and locally invasive tumors. The most common type of soft tissue sarcomas is liposarcomas. It consists of <1% of all malignant tumors in adulthood.^{1,2} Retroperitoneal liposarcoma (RLS), is a rare, biologically heterogeneous tumor that presents considerable challenges due to its size and deep location.¹ 85% of RLS are malignant tumors that remain occult for long periods and grow quite large. Their typical symptoms are discomfort, pain or a palpable mass due to large size. These tumors occur most frequently in men, generally in the fifth or sixth decade of life. Magnetic resonance imaging (MRI) provide reliable data about localization of mass and relationship between vascular structures. Although complete surgical resection is mandatory for extending survival, the majority of patients with huge RLS will develop locally recurrent disease following surgery.¹⁻³ So that patients should be followed routinely in every 3 months during first two years.⁴ Pleomorphic liposarcoma is the rarest subtype and is a high grade tumor. The risk of recurrence and metastasis with liposarcoma increases with higher grade. Herein, we report a case of a 68 year-old man with a 35 cm mass adjacent to kidney with synchronous inguinal 7 cm mass.

CASE REPORT

A 68 year-old man, who had been suffering from progressive enlargement and discomfort of abdomen with weight loss during last 3 months. Other past medical and family histories were unremarkable. On physical examination, a palpable mass on the right flank was found. The abdomen was extremely swollen, tense and dilated veins were apparent over abdomen.

Other examinations were normal. The routine laboratory tests were normal. The computed tomography scan showed a large lobulated, heterogeneous perirenal mass without preservation of perirenal fat (13x18x24.5cm) (Figure 1) and another round solid mass on the right side of bladder (4.4x4.5x4.0 cm) (Figure 1 b). The biopsy of this mass revealed sarcoma. FDG-PET CT scan for staging showed no metastasis. After informed consent was obtained from the patient, retroperitoneal exploration was performed. Large perirenal mass with the intraoperative impression of renal involvement was present. Ascending colon was adherent to it and displaced anteriorly. Right ureter was pushed across the midline towards left side. Right radical nephrectomy was completed (Figure 2). At the same operative session, the mass on the right

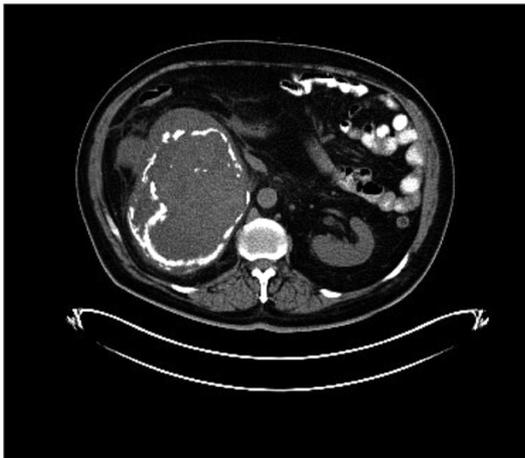


FIGURE 1: a) A Heterogeneous retroperitoneal mass with calcification (13x18x24.5cm) (SUV max 6.5).

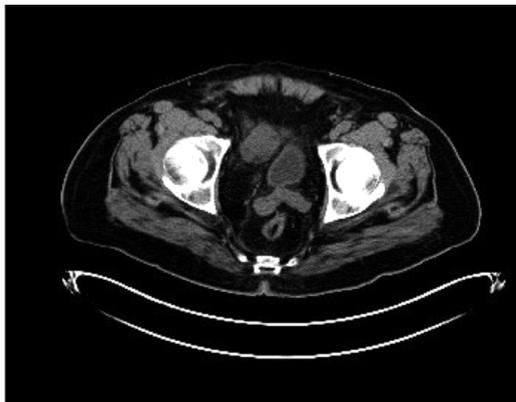


FIGURE 1: b) A mass on right side of the bladder (4.4x4.5x4.0 cm) (SUV max 4.2).

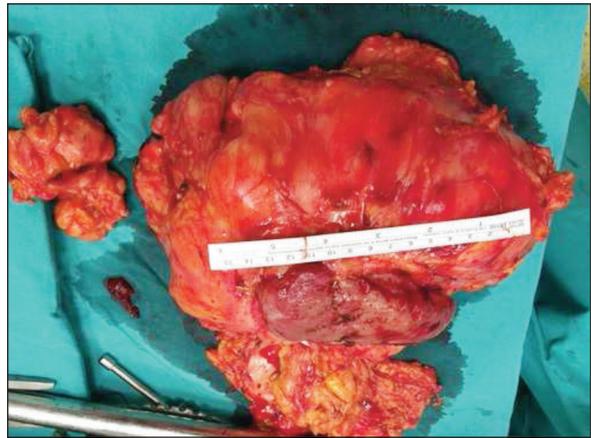


FIGURE 2: Right retroperitoneal mass and synchronous mass on the right side of bladder.

side of bladder was also removed with another incision. The postoperative period was uneventful and the patient was discharged after 3 days. The histopathology was pleomorphic liposarcoma with the invasion of renal capsule.

RT was not thought in our patient because of the separated two masses in different locations. Chemotherapy protocol known as AIM (doxorubicin, mesna) schema was started. During the chemotherapy, febrile neutropenia and life-threatening toxicities happened. So, the chemotherapy withheld. At the nine months follow-up, the patient was asymptomatic without any evidence of recurrence or metastasis.

DISCUSSION

Soft tissue sarcoma account for <1% of all malignant tumors in adults.^{1,2} About 10-15% of adults STS are located in the retroperitoneum (RP). Liposarcoma is the most common variant and accounts for more than 50% of RP sarcomas. Retroperitoneal liposarcoma is a rare tumor and exhibits considerable histological heterogeneity.¹ The natural behavior of and outcome of STS are dependent of the age of the patient, anatomical site and depth, size, and resectability of the tumor, as well as of histology, grade, nodal disease and distant metastasis.⁶ Most RLS, even of important size, rarely metastasize.¹ They occur generally in the 4th and 6th decades of life. The tumors are clinically asymptomatic for a long periods of time, but symptoms can develop

when the tumors become large enough. Pain, abdominal mass and weight loss are usually presenting symptoms. Hematuria is rare even when the tumor reaches a large size.⁵ Pleomorphic liposarcoma is the rarest subgroup with higher grade.^{7,8}

The diagnosis and treatment of RLS mandates a multidisciplinary approach. Different imaging studies can be used in the evaluation of retroperitoneal tumors. Ultrasound is useful as a quick first test with limited value for in-depth evaluation. The diagnostic investigation of choice is contrast-enhanced CT scanning or MRI of the abdomen. Liposarcoma demonstrate a characteristic appearance on CT and MRI with a predominantly fatty component. In most patients with RLS, preoperative radiological findings are nearly diagnostic and pre-treatment biopsy therefore unnecessary.¹⁰ In some patients, however, radiology may suggest a different pathology that may not require surgery as the first approach as in our case (Lymphoma etc.).

CT scan of the chest and abdomen is choice of investigation for staging. FDG positron emission tomography scan may provide additional biological information about the retroperitoneal tumor and may possibly differentiate a high grade from a low grade tumor apart from staging (detection of metastasis).¹

Surgery is the mainstay of treatment of non-metastatic retroperitoneal liposarcoma. Total surgical resection with free margins offers good prognosis.⁹ Even after complete resection of RLS, local recurrence remains common and constitutes the most frequent cause of death. Therefore, adjuvant radiation therapy (RT) may constitute a treatment option. Preoperative RT is also regarded as the treatment of choice. It helps to avoid damage to radiation sensitive structures and organs that usually fill in the resection bed.¹¹ RT was not considered as a treatment option in our patient because of separated two masses in different locations.

Chemotherapy has an established role in the palliative management of advanced or metastatic soft tissue sarcomas. In a meta-analysis it's shown that adjuvant doxorubicin-based chemotherapy combined with ifosfamide decreases both the local recurrence and the distant metastases significantly in soft tissue sarcomas.¹² Our case was considered as a stage 4 and extensive primary tumor. So that, chemotherapy was thought in our case as considered stage 4 and extensive primary tumor, even though both of the masses were resected completely.

In conclusion, because of the rarity of the disease, patients with suspected retroperitoneal liposarcomas should be referred to a high-volume center where they can be diagnosed and treated using a multidisciplinary team approach of surgeons, oncologists, radiologists, pathologists.

Informed Consent

Informed consent was obtained from the patient.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Fatih Kurtuluş; **Design:** Oğuzhan Parlakkılıç; **Control/Supervision:** Fatih Kurtuluş, Mustafa Teoman Yanmaz; **Data Collection and/or Processing:** Fatih Kurtuluş, Mustafa Teoman Yanmaz; **Analysis and/or Interpretation:** Fatih Kurtuluş, Oğuzhan Parlakkılıç; **Literature Review:** Fatih Kurtuluş, Oğuzhan Parlakkılıç; **Writing the Article:** Fatih Kurtuluş; **Critical Review:** Mustafa Teoman Yanmaz.

REFERENCES

1. Matthysens LE, Creyten D, Ceelen WP. Retroperitoneal liposarcoma: current insights in diagnosis and treatment. *Front Surg* 2015;2:4.
2. Giray E, Akay EO, Ebioloğlu T, Köprü B, Tahmaz L. [Retroperitoneal liposarcoma: case report]. *Bulletin of Urooncology* 2015;14:74-6.
3. Raza M, Reddy S, Dileep D. Huge retroperitoneal liposarcoma: case report. *Paripex-Indian Journal of Research* 2014;3(8):125-6.
4. Osmanağaoğlu MA, Bozkaya H, Ozeren M, Cobanoğlu U. Primary retroperitoneal liposarcoma. *Eur J Obstet Gynecol Reprod Biol* 2003;109(2):228-30.
5. Kim MJ, Koo HL, Jun SY, Ro JY. Pleomorphic liposarcoma of the kidney. A case report. *Korean Journal of Pathology* 2003;37:210-3.
6. Bader DA, Peres LA, Bader SL. Renal liposarcoma. *Int Braz J Urol* 2004;30(3):214-5.
7. Hornick JL, Bosenberg MW, Mentzel T, McMenamin ME, Oliveira AM, Fletcher CD. Pleomorphic liposarcoma: clinicopathologic analysis of 57 cases. *Am J Surg Pathol* 2004;28(10):1257-67.
8. Wang L, Ren W, Zhou X, Sheng W, Wang J. Pleomorphic liposarcoma: a clinicopathological, immunohistochemical and molecular cytogenetic study of 32 additional cases. *Pathol Int* 2013;63(11):523-31.
9. Mayes DC, Fechner RE, Gillenwater JY. Liposarcoma renal. *Am J Surg Pathol* 1990;14(3):268-73.
10. Chew C, Reid R, O'Dwyer PJ. Value of biopsy in the assessment of a retroperitoneal mass. *Surgeon* 2006;4(2):79-81.
11. Mohindra P, Neuman HB, Kozak KR. The role of radiation in retroperitoneal sarcomas. *Curr Treat Options Oncol* 2013;14(3):425-41.
12. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. *Cancer* 2008;113(3):573-81.