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Management of Giant Retroperitoneal Liposarcoma: A Case Report

Dev Retroperitoneal Liposarkomun Yönetimi: Bir Olgu Sunumu

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Abstract

Retroperitoneal liposarcoma (RPLS) is a rare tumor. Early diagnosis and treatment are difficult due to absence of specific clinical presentations. We report a case of a 66-years-old woman who succesfully underwent complete surgical resection for a giant retroperitoneal liposarcoma. The complete surgical resection is the most important predictor of local recurrence and overall survival. We believe that complete surgical resection involving adjacent organs is a curative treatment to increase overall survival, especially in the presence of invasion of large tumors.

Keywords: retroperitonel, liposarcoma, large tumor, surgery

Öz

Retroperitoneal liposarkom (RPLS) nadir görülen bir tümördür. Belirgin klinik bulguların olmaması nedeniyle erken tanı ve tedavi zordur. Biz 66 yaşında kadın hastada dev bir retroperitoneal liposarkomun başarılı bir şekilde tam cerrahi rezeksiyon olgusunu sunuyoruz. Tam cerrahi rezeksiyon, lokal nüksün ve genel sağkalımın en önemli prediktörüdür. Komşu organları içeren tam cerrahi rezeksiyonun, özellikle büyük tümörlerin invazyonu varlığında genel sağ kalımı artırmak için önemli bir tedavi olduğuna inanıyoruz.

Anahtar Kelimeler: retroperitoneal, liposarkom, büyük tümör, cerrahi

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Inroduction

Retroperitoneal sarcomas represent 10-15% of all soft tissue sarcomas. The most common histological type of sarcomas is liposarcoma, accounting for 20-45% of cases [1]. Retroperitoneal liposarcoma (RPLS) usually occurs in 40-60 year-old patients, with a male /female ratio of 1:1 [2]. Because of the largeness of retroperitoneal area, liposarcomas are usually asymptomatic. When initially diagnosed, the sarcoma has reached a large size and often invades adjacent organs [3]. If needed a negative surgical margin should be provided by resection of adjacent organs to improve survival. However, the 5-year survival rate is 20% in the well-differentiated and 83% in the undifferentiated subtypes [4]. We report the management of a rare case of a giant 25 cm retroperitoneal liposarcoma.

Case Presentation

A 66-year-old female was admitted to our clinic with abdominal pain. On physical examination a hard, non-mobile mass extending from the bottom of the left costal margin towards the inguinal region was palpated. Contrast-enhanced abdominal computed tomography (CT) was performed. A 25x16 cm sarcoma mass surrounding the left kidney was detected on CT, and the operation was planned (**Figure 1**). The patient was operated through a left semi-chevron incision. It was observed that the left kidney was completely surrounded by the mass. The mass and the left kidney were removed together (**Figure 2**). The operation time was 170 min and the estimated blood loss was 250 ml. No perioperative complication occured. The patient was discharged on the 4th postoperative day. The histopathologocical evaluation revealed a well-differentiated liposarcoma with a size of 25x16x12 cm (**Figure 3**). Neoadjuvant and/or adjuvant radiotherapy and chemotherapy were not given to the patient. No recurrence or distant metastasis was detected in the postoperative 12 months of follow-up.

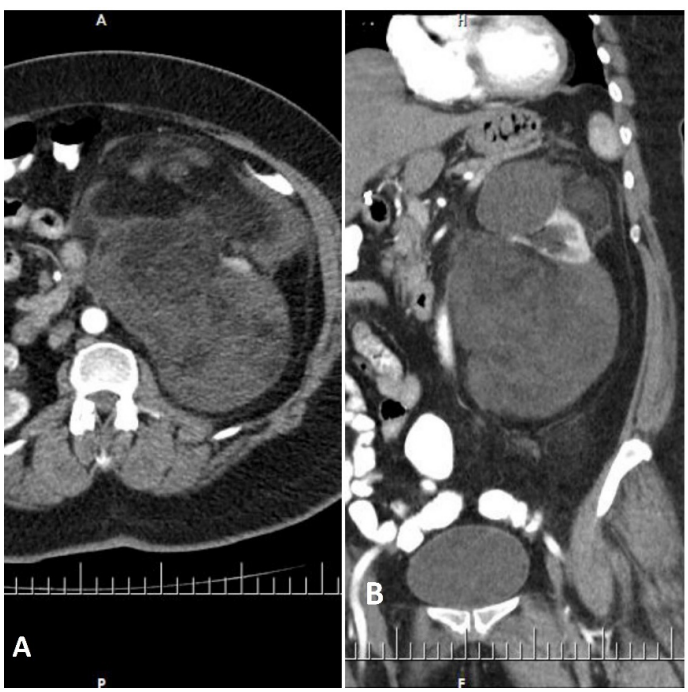

Figure 1. A- Contrast-enhanced abdominal computed tomography (CT) in the axial plane showing a large tumor in the abdomen B- Contrast enhanced CT scan in the coronal plane showing large tumor surrounding the left kidney

Discussion

RPLS is a rare tumor accounting for less than 0.2% of all malignancies. However, it is the most common type of retroperitoneal tumor. RPLS does not manifest typical symptoms, so it is difficult to make a diagnosis at an early stage. When clinical symptoms occur, the liposarcoma usually becomes large in size and compresses the adjacent organs [5]. Liposarcomas can be of different sizes and weights. Retroperitoneal liposarcomas

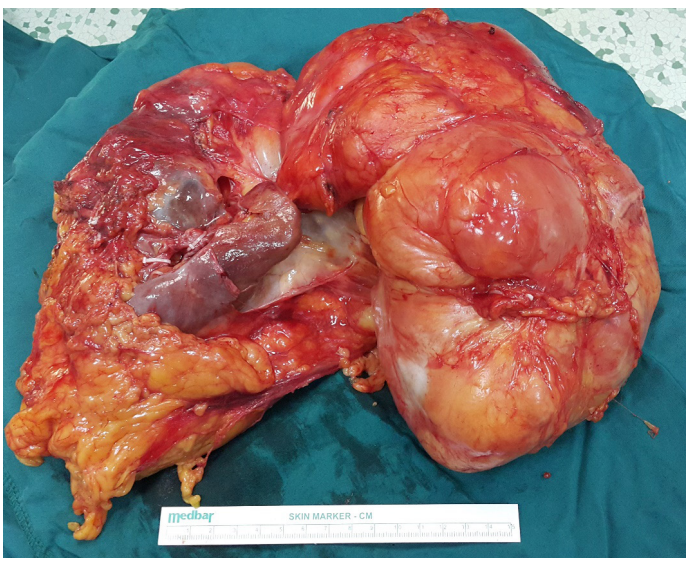

Figure 2. Gross appearance of the liposarcoma, 25x16x12 cm in size

that weigh more than 20 kg are called giant liposarcomas and are extremely rare [2].

The most common diagnostic method for RPLS is CT. On CT imaging, liposarcomas appear as a large encapsulated mass containing different amounts of fat and soft tissue components. CT also helps in detecting tumor size, adjacent organ invasion, and distant metastases [6]. In our case, we used contrast-enhanced abdominal CT as a diagnostic test.

The definitive diagnosis of RPLS is established by histopathological examination. RPLS histologically (undifferentiated, pleomorphic, well- differentiated, myxoid/ round cell RPLS) is divided into 4 groups [7]. Undifferentiated, pleomorphic types are high-grade carcinomas, and the rate of metastasis and recurrence is high. On the other hand, well -differentiated and myxoid / round cell types are low- grade carcinomas with a good prognosis [4]. Gronchi et al. reported overall 5-year survival rates for well-differentiated, myxoid/ round cell, undifferentiated and pleomorphic, as 90%, 60 - 90%, 75% and 30 - 50%, respectively [8].

Complete surgical resection is the most important point of treatment. Successful complete resection of retroperitoneal liposarcoma has been shown to increase the 5-year survival rate from 16.7% to 58% [9]. It has been shown that approximately 80% of RPLS patients are eligible for complete surgical

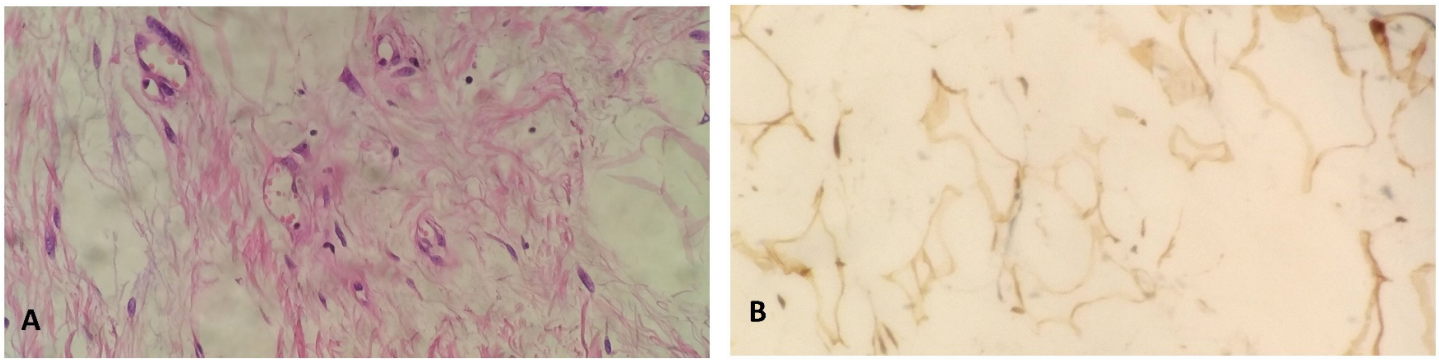

Figure 3. Histopathologic findings A- Well- differentiated liposarcoma (original magnification X100, H&E staining) B- Atypical lipocytes with strong s-100 positivity are observed

resection and that this treatment prolongs the median survival time by 83 months and these patients have a 5-year disease-free survival rate of 60%. In another study, 3 and 5-year survival rates of patients who underwent complete resection were reported as 73% and 60%, respectively [10]. Long-term prognosis is poor in patients without complete resection and 5-, and 10-year survival rates are 16.7% and 8%, respectively [11].

To achieve complete removal of the tumor, 57.0 - 83.0% of patients with RPLS require resection of adjacent organs such as kidney, adrenal gland , ureter, colon, small intestine, omentum and spleen [10]. In addition, complete resection involving resection of organs adjacent to the tumor has been shown to be useful for the prevention of local recurrence. In RPLS cases, the most common resected organ during complete resection is the kidney [12]. Radical nephrectomy performed in cases of RPLS located near the kidney has a beneficial effect on disease-free survival [13]. In our case, a giant 25 cm liposarcoma located in the left retroperitoneal region was found to surround the left kidney. The mass and the kidney were removed all together to increase survival rates and achieve a negative surgical margin. Indeed, in our case surgical margin negativity was obtained. No local recurrence or distant metastasis was detected in CT at postoperative 3th, 6th, and 12th months.

The benefits of using adjuvant chemotherapy and radiotherapy to improve survival in RPLS patients are controversial [10]. Adjuvant chemotherapy has been shown to benefit very few patients in a limited number of studies [13,14]. However, chemotherapy has been suggested, but it has worsened patient's prognosis [10]. The effectiveness of postoperative radiotherapy has been inquired in local control rather than overall survival [11]. Ballo et al. also stated that radiotherapy was ineffective in RPLS cases and radiotherapy applied may cause neuropathy, hydronephrosis, fistula and ileus [15]. It should not be ignored that radiotherapy to be applied to the retroperitoneal area may damage the visceral organs such as kidney, liver and intestines [15]. In our case of RPLS, we performed complete resection. Adjuvant radiotherapy and chemotreapy were not performed because the surgical margin was negative and histological subtype was well-differentiated liposarcoma.

In conclusion, RPLS is a rare tumor with a high rate of relapse without any typical symptoms. The large size of the mass at the time of diagnosis can make surgery difficult. Complete resection of the mass (resection of adjacent organs may also be required) is the most important step for treatment. In addition, histopathologic subtypes are important in survival. RPLS should be treated with a multidisciplinary approach and a long-term follow-up examination should be performed.

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