CASE REPORT

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Review of the Literature of Retroperitoneal Liposarcoma with Two Cases

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ABSTRACT Retroperitoneal liposarcoma is a very rare tumor with an incidence of one per 2.5 million and usually presents between ages of 40 and 70. The retroperitoneal space is a large area which delays the symptoms of liposarcoma. Here we present 2 cases with retroperitoneal liposarcoma. The first one is about huge retroperitoneal liposarcoma about 22 cm that was excised with open surgery and the second one is an incidentally diagnosed small liposarcoma about 3 cm that was excised with laparoscopic surgery.

Keywords: Liposarcoma; retroperitoneal liposarcoma; laparoscopy

Retroperitoneal liposarcoma is seen very rarely and has a very low incidence of approximately 1/2,500.000.[1] The retroperitoneal space is a large area which delays the symptoms of liposarcoma. So liposarcoma can grow asymptomtomatically until compresses the surrounding organs.[2] Early diagnosis is very difficult because of disease nature and the absence of specific clinical presentations. The most common symptoms are flank pain and constipation due to mass compression.

Here we present 2 cases with retroperitoneal liposarcoma. The first one is about huge retroperitoneal liposarcoma that was excised with open surgery and the second one is an incidentally diagnosed small liposarcoma that was excised with laparoscopic surgery.

CASE REPORTS

CASE 1: HUGE RETROPERITONEAL LIPOSARCOMA

A 55-years old man was admitted to the our polyclinic with right flank pain. He had no surgical and renal disease history. With physical examination, a palpable large mass filling the right abdomen was palpated. Complete blood count (CBC), renal function test (RFT) and urine analysis were normal. In computed tomography, a huge mass (22x16x12 cm) that pushing the right kidney to anterior was revealed in right retroperitoneal area (Figure 1). At laparotomy with a right transverse incision showed a giant soft yellow mass that originated in the right retroperitoneal area. This mass involved the right renal vascular structures. We performed retroperitoneal mass excision and right nephroureterectomy due to kidney involvement. The tumor was excised with two parts-nephro-ureterectomy specimen and retroperitoneal mass (Figure 2). There was no complication during surgery.

On postoperative second day, he had severe pain and dyspnea. A chest X-ray revealed right pneumothorax and a thorax tube was inserted. The thorax tube was removed at postoperative fifth day. The patient was discharged at postoperative seventh day without any symptoms.

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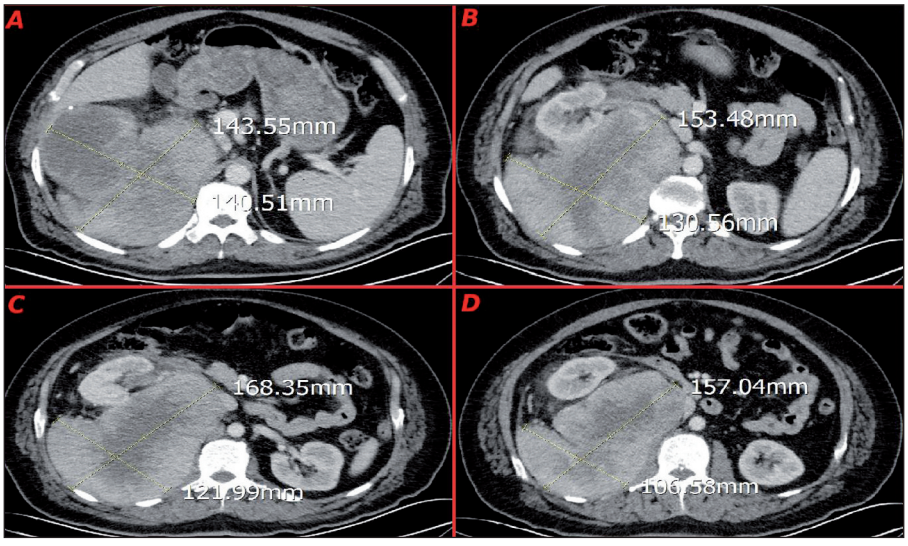

FIGURE 1: Radiologic imaging of liposarcoma in the first case.

Histopathological examination of the specimen revealed retroperitoneal dedifferentiated liposarcoma that infiltrated right ureter and right renal parenchyma. On postoperative 3. month, radiologic imaging revealed a local recurrence and radiotherapy (25x200 cGy) was planned. Radiotherapy treatment continues. Informed consent was obtained from the patient.

CASE 2: SMALL RETROPERITONEAL LIPOSARCOMA

A 42 years-old man was admitted to our polyclinic with a 3 cm aortocaval mass that was incidentally detected (Figure 3). He had left open partial nephrectomy with a flank incision 6 years ago due to renal cell carcinoma. The patient underwent laparoscopic retroperitoneal mass excision (Figure 4, Figure 5). The patient was discharged at postoperative second day without any symptoms.Histopathologic examinations of the specimen revealed dedifferentiated liposarcoma. The sixth month follow-up was uneventful and abdomen magnetic resonance imaging (MRI) revealed no recurrence. Informed consent was obtained from the patient.

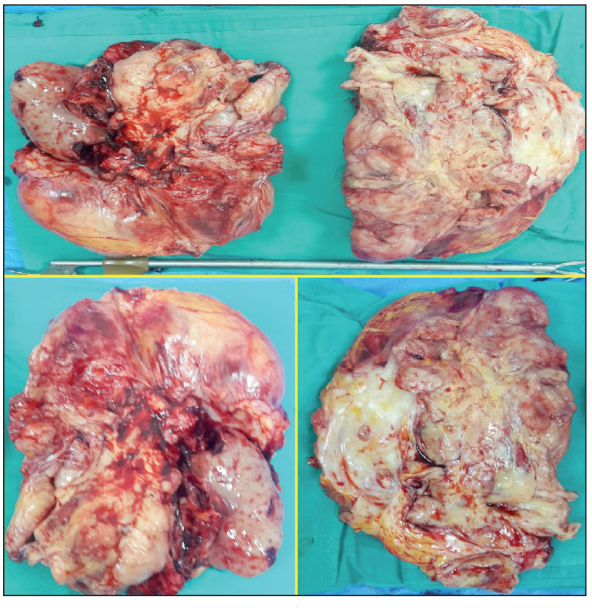

FIGURE 2: Macroscopic view of nephrectomy and liposarcoma in the first case.

DISCUSSION

Liposarcoma is one of the most common soft-tissue sarcoma that is usually seen between the fourth-to-seventh decades.[3] Retroperitoneal liposarcoma usually originates from fat, loose connective tissue, fascia, muscles, lymphatic tissue or residual embryonic tissue, that mostly malignant. Retroperitoneal liposarcoma is seen very rarely and may give symptoms when it reaches huge dimension. Because of the large retroperitoneal space, liposarcoma can grow without no compressing symptoms.

Liposarcomas are malign tumors consist of lipoblasts. Liposarcoma has 5 subtypes; myxoid, well-differentiated, round cell (poorly differentiated

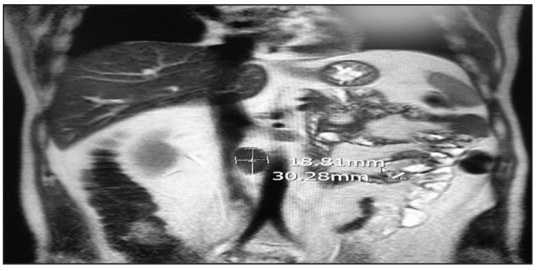

FIGURE 3: Radiologic imaging of liposarcoma in the second case.

myxoid liposarcoma), pleomorphic and dedifferentiated liposarcoma.[4] The clinical characterization is related to histological grade of this tumors. Dedifferentiated liposarcomas are distinguished by a giant chromosome derived from the 12q region, a finding identical to that displayed by well-differentiated liposarcomas.[4] Singer and coworkers reported that dedifferentiated liposarcomas show different patterns than differentiated liposarcomas in a study of 177 cases.[5] This study revealed the risk of local recurrence is approximately four-times.[5]

Survival rates are extensively affected by histologic pattern of subtype for the patients diagnosed with liposarcomas. Well-differentiated subtype's five-year survival rate is 90% but pleomorphic subtype's is only 30-50%. Rates for de-differentiated and myxoid/round cell subtypes are in series of %75 and 60-90%.[5] De-differentiation, grade II-III, stage II,III, size >20 cm, and positive surgical margins are other negative prospective determinants that affect survival.[6]

Retroperitoneal sarcoma is also seen in aorta-caval areas. Depending on the size of mass and location, the clinician can decide on surgical approach; laparoscopic or open surgery. In the first case, we reported that a giant retroperitoneal liposarcoma involved the left kidney pedicle and adherent to left ureter. R0 (microscopically margin-negative resection) resection was completed with open surgery. In the second case, a small 3 cm mass was detected in aortacaval area and R0 resection was completed with laparoscopic surgery. With open surgery or laparoscopic surgery, complete surgical resection is the gold standard in the treatment of retroperitoneal liposarcomas.[5]

The significance of an R0 resection was first represented by Lewis et al. in 1988.[7] The authors reported that the median survival rate of liposarcoma was approximately 103 month with R0 resection. R2 (all of the tumor was not resected macroscopically) resection of retroperitoneal liposarcomas have poor outcomes compared with R0 and R1 (The tumor was fully resected macroscopically, but margins are positive microscopically) resection.[8] Despite of them, there is no consensus about differences of R0 and R1 resection in literature.

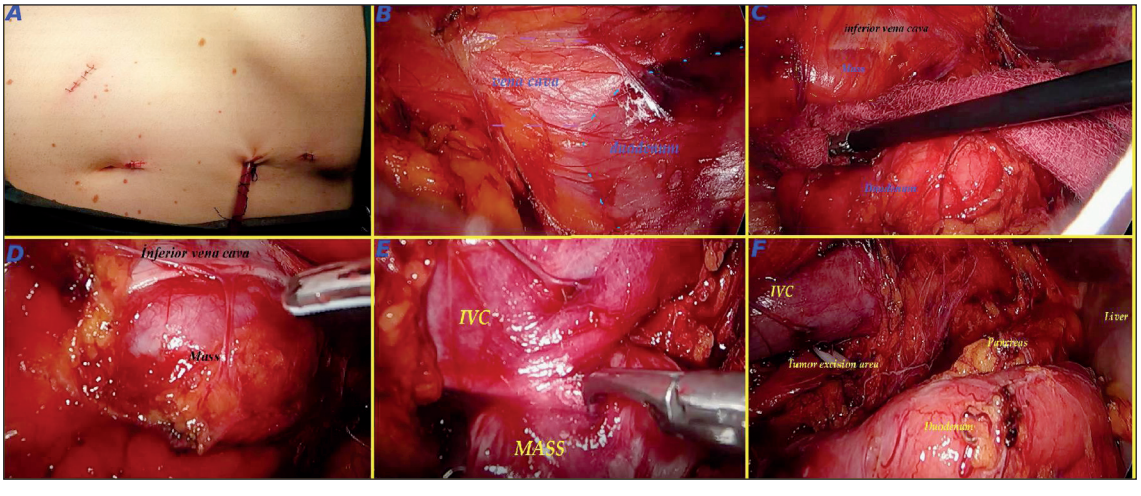

FIGURE 4: Images during surgery in the second case. **A)** Points where ports were placed, **B)** After colon was medialised, **C)** Dissection of the duodenum and IVC, **D)** view of the mass in posteior of IVC after dissection from duodenum, **E)** Excision of the mass from IVC, **F)** After excision).

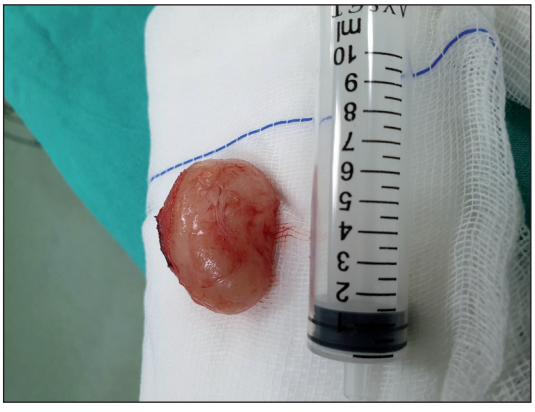

FIGURE 5: Macroscopic view of sarcoma in the second case.

Local recurrence usually occurs after surgery of retroperitoneal liposarcoma in R1 and R2 resection. In local recurrence, gold standart therapy is surgery like the treatment of primary tumor because of resistance of systemic therapy.[9] Re-surgery has lots of complications so patient selection is very important. Besides surgery, radiotherapy is considered in local recurrence.[9]

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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: *Erdinç Dinçer, Ahmet Sahan;* ***Design:*** *Erdinç Dinçer, Ahmet Sahan;* ***Control/Supervision:*** *Oktay Akça;* ***Data Collection and/or Processing:*** *Cengiz Çanakçı;* ***Analysis and/or Interpretation:*** *Ahmet Sahan, Burcu Hancı;* ***Literature Review:*** *Erdinç Dinçer, Burcu Hancı;* ***Writing the Article:*** *Erdinç Dinçer, Ahmet Sahan;* ***Critical Review:*** *Oktay Akça.*

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