

# Giant retroperitoneal liposarcoma

## Report of 5 cases



*Ann. Ital. Chir.*, 2012 83: 161-166

Amin Makni, Aymen Triki, Fadhel Fetirich, Rachid Ksantini, Faouzi Chebbi, Mohamed Jouini, Montassar Kacem, Zoubeir Ben Safta

*Department of General Surgery 'A', La Rabta Hospital, Tunis, Tunisia*

### Giant retroperitoneal liposarcoma. Report of 5 cases

**BACKGROUND:** *Liposarcoma is the most frequent histopathological variety of the retroperitoneum, surgery is the gold standard for treatment.*

**CLINICAL SERIES:** *We report, retrospectively over 7 years (2000-2006), 5 cases (four men and one woman, the mean age was 48 years) of giant retroperitoneal liposarcomas and study their clinical characteristics intra operatively findings and course evolution. None of our patients had a percutaneous biopsy of his tumour. Surgery was indicated to all patients. Resection was performed in 4 cases (we performed a bloc resection in three cases and a simple tumour resection in one case), and in one case, abstention was decided. None of patients had an adjuvant therapy. Three of the patients still alive after the follow up of 2, 3 and 4 years.*

**CONCLUSION:** *Retroperitoneal liposarcomas represent a unique situation and require a more aggressive surgical approach including multiple resections for recurrences. Based on the ability of the patient to tolerate the procedure, surgery is suggested to evaluate resectability of the tumor. We must take into consideration whether prolonged survival will be attained and tumor removal will result in palliation of symptoms.*

**KEY WORDS:** Giant, Liposarcoma, Retroperitoneum.

### Introduction

Liposarcomas are neoplasms of mesodermic origin derived from adipose tissue and correspond to 10-14% of all soft tissue sarcomas. They represent < 1% of all malignant tumors<sup>1,2</sup>. The most frequent subtypes are liposarcoma (41%)<sup>3</sup>. Retroperitoneal liposarcomas alone comprise 0.07–0.2% of all neoplasias<sup>4</sup>. Approximately 85% of these are malignant, with soft-tissue sarcomas

representing 35% of this group. Liposarcoma is the most frequent histopathological variety of the retroperitoneum<sup>2</sup>. It presents with inherent characteristics in relation to its deep localization and slow expansive growth. Average diameter of the tumor is 20-25 cm with a weight of 15-20 kg<sup>4</sup>. There is compromise of the adjacent organs in up to 80% of the cases.<sup>4,5</sup> Surgery is the gold standard for treatment of liposarcoma. There is a low incidence of distance metastasis (7%) compared to other histological subtypes that range from 15 to 34%<sup>6</sup>.

### Clinical Series

#### CASE REPORT N. 1

A 60-year-old male presented with a 6-month evolution of his disease with abdominal pain and constipation without weight loss. He also presented with respiratory

*Pervenuto in Redazione Settembre 2011. Accettato per la pubblicazione Novembre 2011*

*Correspondence to: Amin Makni MD, Department of General Surgery 'A', La Rabta hospital, Jabbari 1007, Tunis, Tunis El Manar University, Faculty of Medicine of Tunis, 15 Rue Djebel Akhdhar, Tunis, Tunisia (E-mail: aminmakni@msn.com)*

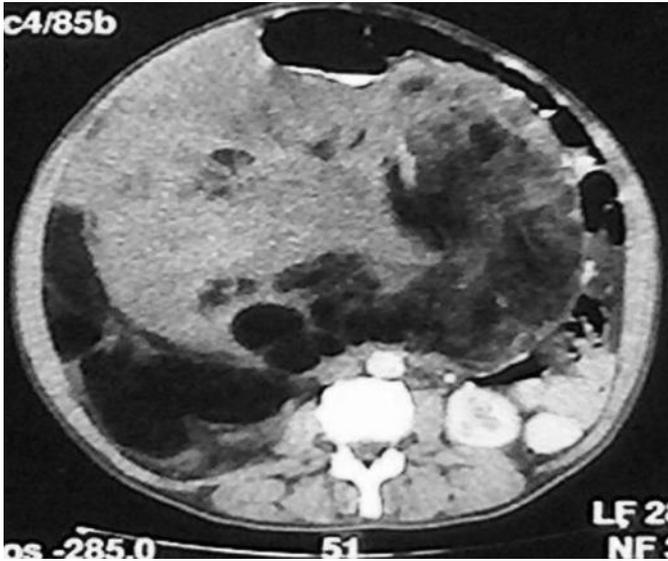


Fig. 1: Abdominal CT scan. Heterogeneous lesion is observed with zones of fat and solid density that entirely occupy the abdominal cavity, displacing retroperitoneal structures dorsally.

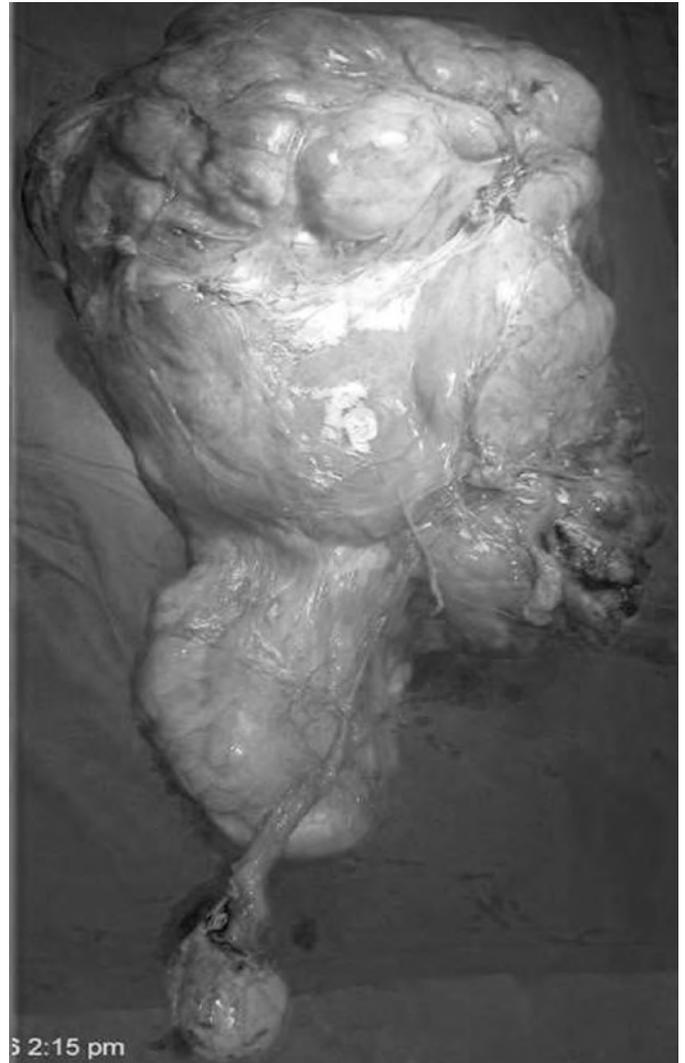


Fig. 3: Photo of the specimen: Retroperitoneal tumor (80 x 60 cm), weighing 18 kg. Complete resection with right orchiectomy.



Fig. 2: Intraoperative photo: Giant right retroperitoneal tumor after mobilization of the right colon.

difficulty. During the physical examination, we were able to palpate the large abdominal mass that extended from the epigastrium to the pelvic region. For this reason, abdominal CT-scan was performed, demonstrating a heterogeneous lesion with zones of fat and solid density that entirely occupied the abdominal cavity, displacing retroperitoneal structures dorsally (Fig. 1). Surgery was performed with the patient in dorsal decubitus and a midline incision was made from the xiphoid to the pubis, revealing a 40 x 30-cm tumor that encompassed the entire retroperitoneal cavity, with loose adhesions to the ascending colon and right ureter, without multiorgan resection and with macroscopic free margins (Fig. 2). Complete resection of the tumor was performed (Fig. 3). The patient had a satisfactory evolution and was discharged 72 h postsurgery without adjuvant treatment.

The final histopathological report showed undifferentiated liposarcoma of the retroperitoneum weighing 20 kg, and the resection margins were tumor-free. The follow-up imaging study showed an intraperitoneal recurrence unresectable after 1.5 years and the patient died 2 years after the operation.

#### CASE REPORT N. 2

A 49-year-old male presented with a 7-month evolution of his disease with abdominal pain without weight loss. He had noted the absence of bowel dysfunction. During the physical examination, we were able to palpate the large abdominal mass that extended from the epigastrium to the pelvic region. It gave the left lumbar contact. The abdominal CT scan confirmed the presence of a large mass of 23 cm occupying the left lumbar fossa, the left flank and hypochondrium, This tumor was of two com-

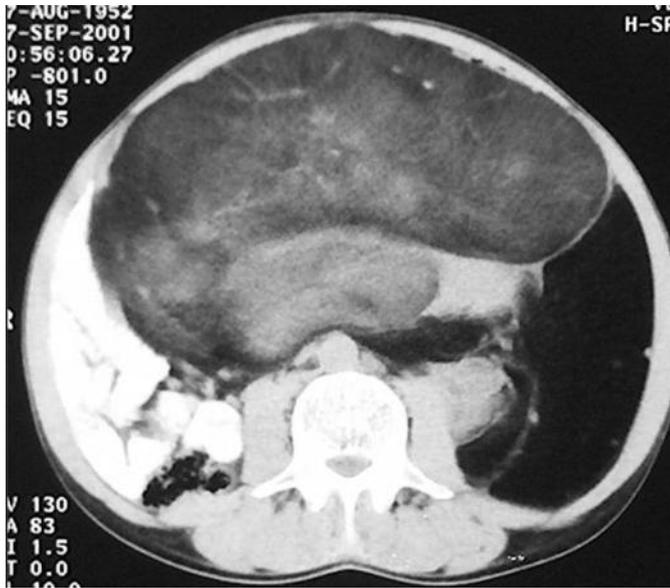


Fig. 4: Abdominal CT scan. Heterogeneous lesion is observed with zones of fat and solid density that entirely occupy the abdominal cavity, displacing digestive structures to the right.



Fig. 5: Abdominal CT scan. Heterogeneous lesion is observed with zones of fat and solid density that entirely occupy the abdominal cavity, displacing medially the left kidney.

ponent, predominantly fatty tissue and taking a moderate contrast (Fig. 4). Moreover, the left kidney and ureter are subsumed and repressed forward and medially by the mass. Surgery was performed with the patient in dorsal decubitus and a midline incision was made from the xyphoid to the pubis, revealing a 25 x 20-cm tumor. The exploration had found a tumor occupying the entire abdominal cavity, adhering to the left renal pedicle and seemed overwhelmed by its lower pole left ureter. It was realized intracapsular enucleation of the mass including the kidney and left adrenal. The patient had a satisfactory evolution and was discharged 72 h post-surgery without adjuvant treatment. The final histopathological report showed undifferentiated liposarcoma of the retroperitoneum weighing 18 kg. The follow-up imaging study showed no evidence of tumour recurrence after 3 years.

#### CASE REPORT N. 3

A 45-year-old male presented with a 12-month evolution of his disease with increased of abdominal girth, without bowel dysfunction. The physical examination found a large mass extended from the left lower quadrant to the left flank, fixed to the deep plane. This mass gave the contact on the left lumbar. In addition we noted the absence of signs of venous compression. The abdominal CT scan confirmed the presence of a large mass of 30 cm occupying the epigastrium, the left hypochondrium and the left flank. This tumor was a two component, predominantly fatty tissue and taking a moderate contrast (Fig. 5). This mass repressed the left

colon, bowel loops, left kidney, spleen and tail of the pancreas. The patient was operated on by median xypho-pubic, exploration had found a large left retroperitoneal mass which adheres closely to the aorta and iliac vessels. Abstentions with simple biopsy of the tumor was decided. The postoperative course was uneventful. The pathologic examination of intraoperative biopsy had found a retroperitoneal liposarcoma. Patient died 1 month later.

#### CASE REPORT N. 4

A 47-year-old woman presented with a 7-month evolution of his disease with abdominal pain without bowel dysfunction. During the physical examination, we were able to palpate the large abdominal mass growing at the left flank. It gave the left lumbar contact. The abdominal CT scan confirmed the presence of a large mass of 20 cm occupying the left lumbar fossa, the left flank and hypochondrium, This tumor was a two component, predominantly fatty tissue and taking a moderate contrast (Fig. 6). Moreover, the mass infiltrated the left ureter. Surgery was performed with the patient in dorsal decubitus and a midline incision was made from the xyphoid to the pubis, revealing a 25 x 20-cm tumor wich adhering to the left kidney, the splenic flexure of the colon, the spleen and the tail of the pancreas. It was conducted a monobloc resection of the tumour mass, associated to the left pancreatectomy, splenectomy, nephrectomy and left colectomy. The patient had a satisfactory evolution and was discharged 9<sup>th</sup> day postsurgery without adjuvant treatment. The final histopathological report



Fig. 6: Abdominal CT scan. Heterogeneous lesion is observed with zones of fat and solid density that entirely occupy the left abdominal cavity.

showed well-differentiated liposarcoma of the retroperitoneum weighing 16 kg. The follow-up imaging study showed no evidence of of tumour recurrence after 2 years.

#### CASE REPORT N. 5

A 40-year-old man presented with a 2-month evolution of his disease with thrombophlebitis of the right lower. The various investigations had found a giant right retroperitoneal liposarcoma of 20 cm, compressing the right common iliac vein. After turning on effective anticoagulation with heparin, the patient was operated on by median laparotomy. It was performed after separation of the right colon, resection of the tumor associated with a right nephrectomy. The postoperative course was marked by the appearance of a hematoma at the renal space that has evolved under medical treatment. The patient had a satisfactory evolution and was discharged 9<sup>th</sup> day postsurgery without adjuvant treatment. The final histopathological report showed well-differentiated liposarcoma of the retroperitoneum weighing 14 kg. The follow-up imaging study showed no evidence of of tumour recurrence after 4 years.

#### Discussion

Liposarcoma is the most frequent histological type of retroperitoneal sarcoma, corresponding to 41% of these tumors<sup>4,5</sup>. It has been reported that 20% of the tumors are greater than 10 cm at the time of diagnosis<sup>3</sup>; however, few cases of retroperitoneal liposarcomas exist that can be considered as giant<sup>4,6-9</sup>. The cases we present

may be considered among the largest series reported for this histology. Retroperitoneal liposarcomas are then further classified by their histologic grades of cellular differentiation: well-differentiated and de-differentiated. Clinically, these tumors tend to present with diffuse abdominal pain accompanied by anorexia and weight loss and increase in abdominal girth which was present in all cases (100%). The most characteristic sign is an abdominal pain detected in 78% of the cases<sup>3</sup>, while this sign was present in 3 of our 5 cases (60%). Abdominal symptomatology as well as thrombophlebitis of right lower limb were due to compression of the organs or vessels, similar to that reported with the present cases<sup>3,4</sup>. In most patients, definite symptoms are lacking, and the diagnosis is routinely made on the basis of CT scans of the abdomen and pelvis.<sup>10</sup> Abdominal CT scan alone can make the diagnosis of retroperitoneal liposarcoma in case of predominantly fatty tissue mass. This eliminates the need for a percutaneous biopsy. In any case, even for a tumor unresectable, no neoadjuvant therapy in the objective of a tumor shrinkage has demonstrated its effectiveness. In our cases, diagnosis was made by abdominal CT scan alone.

Only surgical intervention has been shown to improve overall survival, and the most important variable in outcome is the ability to completely resect the tumor, which often requires removal of the adjacent organs. In a previous study, the median survival of patients who underwent complete resection was 103 months. Conversely, the median survival of patients undergoing incomplete resection was 18 months, which was no different from performing no resection at all<sup>3</sup>. Patrik et al.<sup>6</sup> demonstrated that in liposarcomas > 10 cm, complete resection can be carried out in up to 70% of cases; however, in up to 50% of these cases, multiorgan resection is necessary in order to reach this goal<sup>2</sup>. The most frequent organ resected is the kidney (30%), 60% in the present study. Various chemotherapy regimens have been described based on mesna, doxorubicin, ifosfamide, dacarbazine and paclitaxel. However, their use is limited for recurrent metastatic disease or palliation. Survival benefits have not been demonstrated<sup>11</sup>. With regard to radiotherapy (RT) as complementary treatment, there is agreement for its palliative use in non-operable tumors or in cases of incomplete resection<sup>7,8</sup>. Although mesodermic tumors are radioresistant, liposarcoma is more radiosensitive<sup>6</sup>. Although it has been noted that RT may increase survival and disease-free interval<sup>3,7,8</sup>, other authors reported that this treatment has not demonstrated long-term improvement in survival or specific disease in cases of complete macroscopic resections<sup>6,8</sup>. This occurs despite using intraoperative RT with the goal of increasing efficacy of the local dose with 50-60 Gy<sup>7</sup> and of minimizing toxicity to adjacent organs. The resectability of the tumor and histopathologic grades are associated with disease prognosis, and both factors are hard to elucidate before undergoing surgical inter-

TABLE I - The summary of clinical and therapeutic features of our five observations of giant retroperitoneal liposarcomas.

Case N.	Age (sex)	Symptoms	Side	Size (cm) (Invasion, metastasis)	Therapy	Follow up
1	60 (male)	Abdominal pain, constipation, respiratory difficulty	Right	40 (None)	Resection with orchiectomy	2 years (died)
2	49 (male)	Abdominal pain, constipation	Left	23 (Left kidney)	Resection with nephrectomy	3 years (alive)
3	45 (male)	Increased of abdominal girth	Left	30 (Aorta, iliac vessels)	Abstention	1 month (died)
4	47 (Female)	Abdominal pain	Left	30 (Left kidney, spleen, pancreas, colon)	Bloc tumour resection (with nephrectomy, colectomy, splenectomy and pancreatectomy)	2 years (alive)
5	40 (male)	Thrombophlebitis of the right lower	Right	20 (Right kidney)	Resection with nephrectomy	4 years (alive)

vention. Histopathologic grades of differentiation and tumor subtype also affect overall survival rates. Of the subtypes, the 5-year survival rate of well-differentiated liposarcoma reaches 90%. By contrast, the survival rate of pleomorphic subtypes has been reported to be 50%. In addition, the survival rate has been reported to be 75% in de-differentiated cases and 60% in cases of the myxoid/round cell subtype<sup>1,5</sup>. Resections of contiguous organs are common and include en bloc resection of retroperitoneal organs, including the kidney, adrenal gland, pancreas, spleen, and even vascular resection (inferior vena cava) if indicated<sup>3</sup>. Bloc resection was performed in 3 of our 5 cases, these patients still alive.

## Conclusion

Retroperitoneal liposarcomas are a unique situation and require a more aggressive surgical approach including, when necessary, multiorgan resection or multiple resections with recurrences. In accordance with the ability of the patient to tolerate the procedure, surgery is suggested to evaluate tumor resectability, taking into consideration prolonged survival. After tumor removal, palliation of symptoms will be accomplished.

## Riassunto

Il liposarcoma rappresenta la più frequente varietà istopatologica del retroperitoneo e la chirurgia rappresenta il gold standard del trattamento.

Nell'articolo viene condotta una revisione retrospettiva su sette anni (2000-2006) di 5 pazienti (quattro uomini ed

una donna, dell'età media di 48 anni) portatori di liposarcomi retroperitoneali giganti, studiando le loro caratteristiche cliniche, i reperti intraoperatori e l'evoluzione successiva. Nessuno dei nostri paziente è stato sottoposto a biopsia percutanea del tumore, ed in tutti è stata posta l'indicazione operatoria. In quattro casi si è provveduto ad una resezione (in tre casi una resezione in blocco ed una resezione semplice in un caso), mentre in un altro caso è stata decisa un'astensione intraoperatoria. Nessuno di questi pazienti ha ricevuto un trattamento adiuvante. Tre pazienti erano ancora vivi dopo un follow up di 2, 3 e 4 anni.

In conclusione il liposarcoma retroperitoneale rappresenta una situazione unica e richiede un approccio chirurgico molto aggressivo, comprensivo all'occorrenza di resezioni multiple. Contando sulla capacità del paziente di tollerare la procedura, la chirurgia viene affrontata anche per valutare la reseccabilità del tumore, e bisogna prendere in considerazione se si può ottenere una sopravvivenza prolungata e se l'asportazione del tumore comporta una palliazione della sintomatologia.

## References

1. Kilkenny JW 3rd, Bland KI, Copeland EM 3rd: *Retroperitoneal Sarcoma. The University of Florida Experience*. J Am Coll Surg, 1996; 182:329-39.
2. Hassan I, Park SZ, Donohue JH, et al.: *Operative management of primary retroperitoneal sarcomas. A reappraisal of an institute experience*. Ann Surg, 2004; 239:244-50.
3. 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; 228:355-65.

4. Echenique-Elizondo M, Amodarain-Arratibel JA: *Liposarcoma retroperitoneal gigante*. Cir Esp, 2005; 77:293-95.
5. Jaques DP, Coit DG, Hajdu SI, Bennan MF: *Management of primary and recurrent soft-tissue sarcoma of the retroperitoneum*. Ann Surg, 1990; 212:51-59.
6. McGrath PC, Neifeld, Lawrence W Jr, DeMay RM, Kay S, Horsley JS, Parker DA: *Improved survival following complete excision of retroperitoneal sarcomas*. Ann Surg, 1984; 200:200-204.
7. Azpiazu Arnaiz P, Muro Bidaurre I, De Frutos Gomero A, et al.: *Tumores retroperitoneales. Liposarcoma mixoide retroperitoneal. Presentación de un nuevo caso*. Arch Esp de Urol, 2000; 53:170-73.
8. Romero Pérez P, Rafie Mazketli W, Amat Cecilia M, Merenciano Cortina FJ, Gonzalez Devesa M: *Tumores adiposos retroperitoneales. A propósito de un liposarcoma mixoide gigante*. Actas Urol Esp, 1996; 20:79-84.
9. Guzman Martinez-Valls PI, Ferrero Doria R, López Alba J, et al.: *Liposarcoma retroperitoneal. A propósito de tres casos*. Arch Esp de Urol, 1997; 50:529-31.
10. McCallum OJ, Burke JJ 2nd, Childs AJ, Ferro A, Gallup DG: *Retroperitoneal liposarcoma weighing over one hundred pounds with review of the literature*. Gynecol Oncol, 2006; 103:1152-154.
11. Yoshida Y, Inoue K, Ohsaco T, Nagamoto N, Tanaka E, Tsuruzoe S: *Weekly paclitaxel therapy is curative for patients with retroperitoneal liposarcoma*. Gan To Kagaku Ryoho, 2007, 34:465-67.