

CLINICAL CASE

**A RARE CASE OF A GIANT RETROPERITONEAL
RECURRENT LIPOSARCOMA****Petronela Eugenia Mateoiu¹, F. Dumitrache¹, O. Andronic¹, Alexandra Bolocan^{1,2}, D. Ion^{1,2}**¹The University of Medicine and Pharmacy “Carol Davila”, Bucharest, Romania²The Clinical Emergency Hospital of Bucharest, Romania³The Clinical Nephrology Hospital “Carol Davila”, Bucharest, Romania

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Abstract

We report the case of a giant retroperitoneal liposarcoma, with two rare histological subtypes: pleomorphic subtype at initial surgical resection, and dedifferentiated subtype at the recurrence. A 61-year-old male patient, presented at the University Emergency Hospital Bucharest, for progressive enlargement of the abdomen. The exploratory laparotomy found a deforming mass in the retroperitoneal area, with medial displacement of the left colon, from the splenic flexure to the recto-sigmoid junction. After assessing the resectability of the tumor, it was decided and performed an en-bloc excision of the tumor, without multiorgan resection and with macroscopic free safety margins. Due to the intersection with the genital vessels, their resection was imposed. The tumor was trefoil-shaped, with a diameter of 35 cm. After 7 months of chemotherapy, tumor recurrence occurred, within the same location, confirmed as a dedifferentiated subtype. The same surgical procedure was performed, and the oncologist decided to continue with a more aggressive chemotherapy. Pleomorphic liposarcoma has a high rate of recurrence and has a high resistance at chemotherapy. Due to its deep retroperitoneal location, the relations with the inferior vena cava, the aorta and the genital vessels are essential, in some cases multiorgan resections being necessary. The case presents a rare malignant tumor, a 35 cm diameter retroperitoneal liposarcoma, highly resistant to chemotherapy and with a high recurrence rate.

Keywords: liposarcoma, pleomorphic subtype, en-block excision**Introduction**

Liposarcoma, corresponding to 10–14% of all soft tissue sarcomas [1], is a malignant tumor that arises in the fat cells, such as those located in the thigh or in the retroperitoneum. The pleomorphic subtype of liposarcoma is a rare entity with less than 40 cases diagnosed each year. The dedifferentiated subtype is also rare; it comprises about 9% of liposarcomas and has a high local recurrence rate [8]. Surgery is the

gold standard for treatment of retroperitoneal liposarcoma. It was demonstrated that in liposarcomas larger than 10 cm, complete resection can be carried out in up to 70% of cases [4]. Few cases of giant retroperitoneal liposarcomas have been reported [3]. The most characteristic clinical sign is a painless abdominal mass [1].

Case presentation

A 61-year-old male patient, presented at the University Emergency Hospital, Bucharest, for progressive enlargement of the abdominal volume, without changes in bowel transit, pain or weight loss. As for the clinical examination, the abdomen was enlarged, with a circumference of 110 cm, due to a giant, painless, mobile on superficial plans, fixed posteriorly tumor mass that was occupying the left part of the abdomen. His medical history revealed: diabetes mellitus type 2, for 6 years, under control with oral medication, arterial hypertension stage II, right inguinal hernia, and secondary anemia.

On ultrasound, the following aspects were noticed: intensely echogenic tumoral mass, increased in size, with heterogeneous structure, occupying the left hypochondriac region, the left lateral flank, the left iliac region and the hypogastric area.

The abdominopelvic CT examination reveals voluminous intraperitoneal and retroperitoneal tumoral masses, developed in two-thirds of the left abdominal cavity, the upper pole reaching the infrasplic level, caudally extending anterior to the urinary bladder, with mixed structure containing fluid, fat and solids areas. The left kidney was anteriorly and superiorly displaced, as well as the body and tail of the pancreas (Figure 1-4).

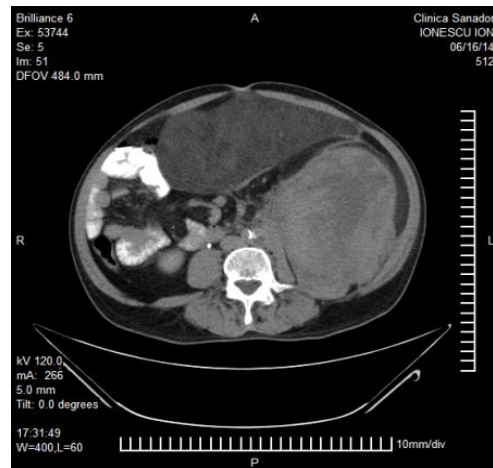


Figure 2 - CT aspect - The giant tumor seen in transverse section



Figure 3 - The descending colon displaced to the right

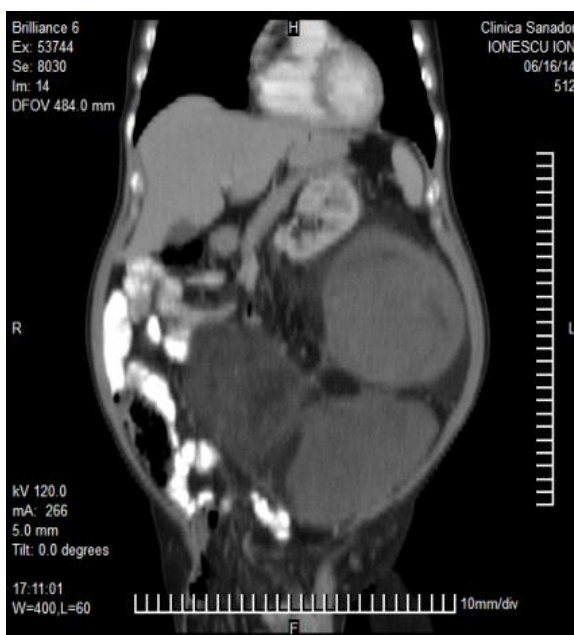


Figure 1 - CT aspect - Three-lobed tumoral mass seen in frontal section



Figure 4 - 3D aspects of the tumor relations. Left kidney pushed antero-medially

Exploratory laparotomy was decided. Preoperative preparation of the digestive tract was applied in the case that hemicolectomy was required.

Surgical procedure: Midline xyphopubic incision was performed extended to the left side at the level of the umbilicus for better access. Skin, fat, linea alba and peritoneum were divided in this order. Intraoperatively a deforming mass that was occupying the left part of the abdomen, starting from the retroperitoneum and pushing to the right the descending colon was found. After assessing the resectability of the tumor, it was decided and performed an en-bloc excision of the tumor. The genital vessels cross almost transversally the tumor and impose their resection. No macroscopic adjacent organ invasion was found. The tumor was trefoil-shaped, with a maximum diameter of 35cm, presenting lipomatous consistency and peritumoral pseudocapsule (Figure 5).



Figure 5 - The larger lobe of the tumor, having a diameter of about 15 cm

Postoperative evolution is favorable. The patient was discharged, surgically cured with the following recommendations: oncological treatment, no strenuous exercises for 90 days post operation, treatment for comorbidities. The anatomopathological outcome sustains the diagnosis of pleomorphic liposarcoma. After 7 months of chemotherapy, tumor recurred.

CT reveals an expansive process presenting irregular contour in the left part of the retroperitoneum, posterior to the left kidney, keeping a demarcation limit with the kidney, with the cranial pole located sub-

diaphragmatically. The cranio-caudal diameter was of 11 cm (Figure 6-7).

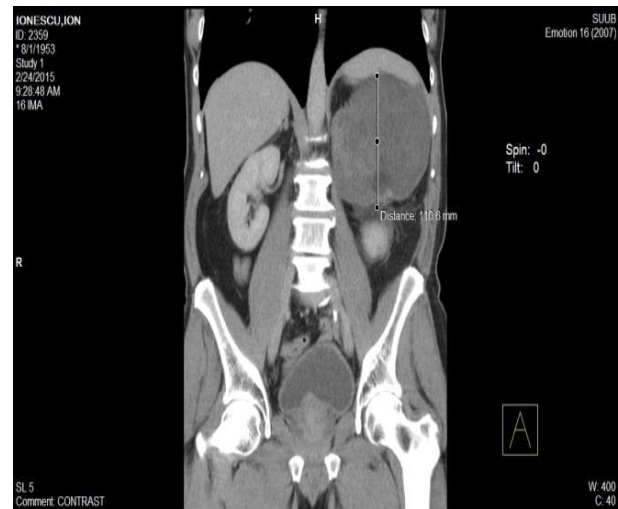


Figure 6 - CT aspect - Tumoral recurrence seen in frontal section

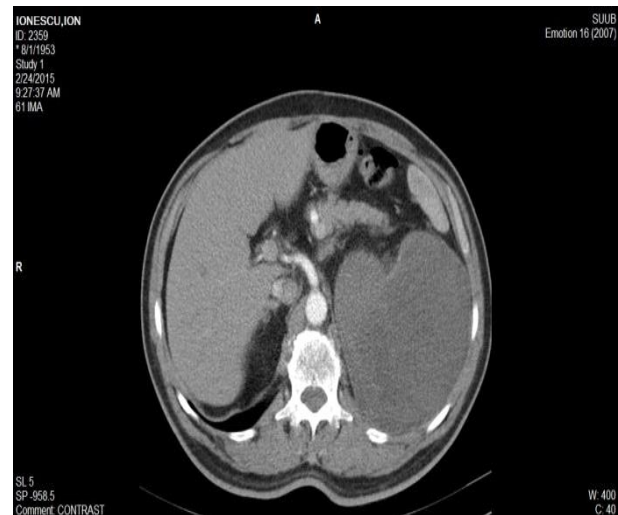


Figure 7 - CT aspect - Tumoral recurrence seen in transverse section

The same surgical procedure was performed, i.e. the en-block excision. No macroscopic adjacent organ invasion was found, therefore no organs were excised. The oncologist decided to continue with a more aggressive chemotherapy. This time, immunohistochemistry analysis revealed dedifferentiated subtype of liposarcoma. Currently, at 11 months after diagnosis, the patient is on chemotherapy treatment.

Discussions

Liposarcoma often becomes symptomatic only when it reaches larger dimensions [5] by

compressing neighboring tissues. Thus, patients usually present to the hospital because of abdominal volume distension, or for nonspecific symptoms, when the tumor has reached impressive dimensions.

In the surgical excision of the retroperitoneal tumors, there is an increased risk of bleeding due to important vascular relations with the inferior vena cava, the aorta, the superior mesenteric pedicle and the genital vessels [2], and the possibility of anatomical variants of vascularization. In certain circumstances, the resection of the genital vessels may be imposed. In some cases, the lower polar artery, a renal supernumerary vessel, may arise from testicular artery [7]. Also, the genital veins may be completely or partially duplicated [6].

If the tumor is retro- or pre-renal, and the kidney is not invaded, the cleavage plane can be used for tumor mobilization. If it becomes necessary, nephrectomy should be proceeded. In case of invasion of the distal ureter, but not the renal parenchyma, ureter resection followed by re-implantation of ureteral segment remaining in the bladder [2] can be practiced. If the retroperitoneal tumor interferes with the colon, removal of the colonic segments may be required. Thus, a preoperative preparation of the colon is necessary. Adjuvant chemotherapy is recommended after surgical excision.

Conclusions

In conclusion, we present a rare tumor, a pleomorphic liposarcoma at initial resection and a dedifferentiated subtype at recurrence. Retroperitoneal liposarcomas are rare tumors which require special medical approach. Because of their increased dimensions, they may affect the neighboring tissues. The en-block excision of the tumor is the gold standard treatment. However, the surgical treatment

should be individualized, depending on the location, the position and the dimensions of the tumor. Lifelong follow-up is recommended in order to monitor for recurrence at the initial site, as well as distant metastasis.

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