

CLINICAL CASE

PRIMITIVE RETROPERITONEAL TUMORS – GIANT LIPOSARCOMA

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Abstract

Primitive retroperitoneal tumors are formations with varying degrees of severity. They may occur locally, being impressive due to their generous diameters, but they can also invade the entire body. The latest genetic studies have somewhat changed the vision about the etiology of soft tissue tumors, while the immunohistochemical studies offer customized results on recurrence, metastasis and survival rate. At the same time, the importance of the imaging findings is crucial, especially when the localization poses problems regarding the surgical technique and a means of adjuvant therapy must be chosen accordingly. The peculiarity of this case is that we managed to resect the impressively large retroperitoneal malignant tumor with oncological safety margins. We present the case of a patient admitted in 2012 in the Department of General Surgery II of the "Carol Davila" Central Military Emergency Hospital, because of the development of a bulky formation in the right hemiabdomen and subocclusive symptoms. A thorough investigation was performed. The initial suspicion was of a primitive retroperitoneal tumor, the CT aspect indicating a liposarcoma. The total tumoral excision was decided and performed, with the previous checking of the specific areas of metastases. The diagnosis certitude was obtained after sending the 5.5 kg piece to the anatomical pathology examination. Post-operatively, the patient was referred to the oncological service for appropriate adjuvant treatment. So far, the surgical treatment combined with the oncological one has had favorable results.

Keywords: liposarcoma, retroperitoneal tumor**Introduction**

Primitive retroperitoneal tumors (PRT) are characterized by the development and localization in the retroperitoneal space, they exclude the metastases or invading tumors from the vicinity and are independent from the structures in the retroperitoneal space.

In adults, the PRT develops on adipose-connective, vascular, nervous tissue (especially in children), on fascia, lymphatic vessels and lymph nodes. The development of primitive

retroperitoneal tumors such as lipomata, liposarcoma, fibromata and paragangliomata is possible as isolated or confluent blocks that become palpable transabdominally. Although the growth of primitive retroperitoneal tumors is slow, they protrude anteriorly and laterally. The supportive connective tissue tumors (lipomata, liposarcoma) can reach impressive diameters. These tumors displace and compress the intra-abdominal viscera, but their invasion is rare.

The liposarcoma are PRT with a mesodermal origin which are formed from supportive

connective tissue. These are the most common malignant tumors of the retroperitoneal space [1]. The predominantly affected segment of the population is represented by males after their fifth decade of life. They generally have a large volume, a polylobed shape, being frequently delimited by a pseudocapsule, with a tendency to displace the mesenteries and the viscera. It is precisely this impressive dimension which leads to their discovery, in the absence of a clear symptomatology. They are divided into four histological types: well-differentiated and myxoid with a low degree of malignancy and the types with round cells, respectively the pleomorphic ones with a high degree of malignancy, with recurrences occurring very fast [2,3]. The dissemination occurs hematogenously the first organs to be affected being the lungs, subsequently metastases developing at the level of the liver.

Case report

O.I., a 48-year-old patient, was admitted in Department of General Surgery II of the "Carol Davila" Central Military Emergency Hospital for the development of a tumor occupying the entire right hemiabdomen. The patient reports a voluntary weight loss during the last three months, and bowel transit disorders (constipation).

The family medical history and the pathological history of the patient are insignificant. The general examination of the systems is normal with the exception of a slowed bowel transit and abdominal distention. The local examination performed with the patient in a supine position reveals at palpation a well defined bulky tumor, about 22/20/23 cm in size, with a firm consistency and mobile on the superficial planes occupying the entire right hemiabdomen. A nonspecific infection is revealed by the bio-humoural tests.

The heart-lung X-ray reveals an enlarged volume of the heart and hydroaeric levels in the projection area of the colon.

The CT scan examination reveals a giant tumor developed in the right hemiabdomen, subhepatically, with maximum diameters of 22.5/20/24.5 cm, coming in contact with the abdominal wall, the lower limit reaching the

sacral promontory with a transmedian extension on the left side, at the level of the renal hilum, with a predominantly lipid component, heterogeneous (including also weakly moderate iodophil tissular areas), apparently well-defined, this imaging aspect suggesting a liposarcoma. The described formation fills the retroperitoneal space, with a significant compression effect on the duodenal frame and the pancreatic cephalic region, as well as on the intestinal loops and the colon, which appear completely displaced into the left iliac fossa. It comes into intimate contact with the inferior vena cava and the aorta at the level of the emergence of the renal vessels. No pulmonary or hepatic metastases are found.



Figure 1 - Frontal anterosuperior reconstruction



Figure 2 - Sagittal reconstruction on the right mid-clavicular line

Based on the medical history, the clinical and the paraclinical examination, the diagnosis of mesenteric cyst, possibly a liposarcoma, is made.

The treatment is strictly surgical, the complete excision of the tumor being proposed. It is decided to perform an exploratory laparotomy under general anesthesia with endotracheal intubation.

The approach used is the median transabdominal one through a xipho-suprapubic incision. The peritoneal cavity is reached, where the polylobed cyst formation with an approximate size of 20/20/25 cm is found occupying the right hemiabdomen, stretching from the visceral margin of the liver to the pelvis, sleeving on $\frac{3}{4}$ the IVC at the level of the renal adhesions, without locoregional adenopathies, without secondary hepatic or peritoneal disseminations. The complete excision of the tumor of the mesentery with gastric marginal resection is decided and performed. The surgical team performed the ablation of the tumor with oncological safety margins, histopathologically certified by the extemporaneous histopathological examination.

After ensuring an adequate control of the hemostasis, drain tubes are placed at the subhepatic, right parieto-colic and subcutaneous levels.

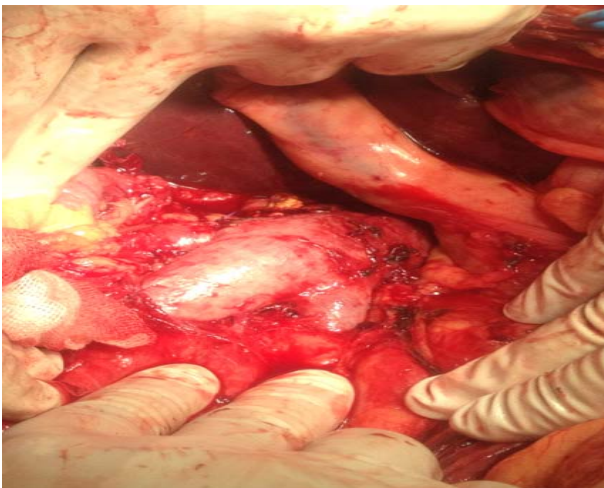


Figure 3 - IVC aspect and renal adhesions after the complete excision

The relatively well defined tumor with a weight of 5.5 kg, having a liposarcoma-like aspect, is sent for the anatomical pathology examination, resulting in the diagnosis of a well-differentiated liposarcoma with myxoid areas.



Figure 4 - Surgical piece (macroscopic aspect of liposarcoma)

Postoperatively, the patient had a slow, favorable evolution with a few febrile episodes (approx. 38° C) and diarrhea (4-6/day for 2 days) with the spontaneous remission of the diarrhea. An ultrasound was performed which revealed no pathological involvement. During the following period, the patient resumed normal bowel movement and in the absence of febrile episodes, 9 days after the surgical intervention, he was discharged surgically healed and was referred to the oncology department. The recommendations of the oncologist consisted in monochemotherapy with Doxorubicin as an adjuvant therapy for the surgical intervention and periodic reassessments. Over the course of 17 months the periodic MRI performed showed no signs of local or distant recurrence, and in terms of the clinical and biological aspects, the cardio-hematological toxicity that could have occurred after chemotherapy was absent.

In the case of our patient, without the surgical intervention, the tumoral mass would have continued to grow causing severe occlusive syndromes both in the gastrointestinal tract and the cardiovascular system, affecting the circulatory and respiratory components. The longer the waiting period, the higher the risk of pulmonary or hepatic metastases and the adjuvant therapy would have failed. Even if so far, the evolution has been favorable, the patient remains under close supervision, the studies mentioning the possibility of recurrences even after 10 years postoperatively.

Discussions

There are genetic studies regarding the influence of the vascular endothelial growth factor (VEGF) and of the platelet-derived growth factors (PDGF) on the development and the evolution of soft tissue tumors located in the extremities, but in the case of the retroperitoneal localization, these factors which influence angiogenesis and stromal proliferation are inconclusive. The same studies, which have just begun, also mention the possibility of a connection between the fibroblast growth factors (FGF) and the survival rate, respectively the patients diagnosed with liposarcoma who present an increased expression of the R-1 receptors for FGF have a higher survival rate [4]. But it is a well-known fact that the prognosis and the treatment are influenced by the histologic type and the observance of the resection margins within oncological limits. The basic treatment for these lesions is the surgical one, the excision with wide margins being preferred if it is possible. The tumors with low malignancy have a tendency to local recurrence and often become more biologically aggressive with each recurrence. The liposarcoma at this level seem to have a lower incidence in point of developing remote metastasis (7%) compared to other histological subtypes (15%-34% Shibata and collaborators noticed a longer survival rate in patients with partial resection versus those with only a biopsy. The resectability of the tumor depends on factors such as anatomical relations with adjacent vascular components or the invasion of the other organs, this aspect being determined preoperatively by means of imaging (CT, MRI, CT angiography) [5]. Although the combination with radiotherapy has proven beneficial in most cases of liposarcoma of the extremities or of the trunk, there are no data to support the same thing for the retroperitoneal ones. Another argument against the postoperative use of external radiotherapy for retroperitoneal liposarcoma is the increased frequency of radix enteritis. This has disastrous consequences for the patient especially since after the removal of the giant tumoral mass, the intestinal loops tend to fill the resection bed where they can be attached by the postoperative adhesions. On the other hand, at a dose of 50 Gray applied to the resection area, the

gastrointestinal toxicity is significant. The American College of Surgeons Oncology Group conducts studies regarding the optimum timing of administering radiotherapy in the case of soft tissue sarcomata situated retroperitoneally. The options taken into consideration are pre-operatively, in such an instance if there is a favorable answer to radiotherapy, obviously the resection will be more successful, intra-operatively or immediately after the removal of the tumoral mass, when the resection bed is well exposed and the viscera can be mechanically moved out of the way of the waves. The use of brachytherapy (targeted internal radiotherapy) is under discussion for the management of the patients with soft tissue sarcomata situated retroperitoneally. At this moment it seems to be quite toxic, especially if used in the upper abdominal region. Currently these practices are not recommended as a means of treatment other than for clinical research.

However, even chemotherapy is rather controversial. The superiority of various combinations of chemotherapy drugs versus monochemotherapy is reported, but the clinical trials do not provide sufficient evidence in their favor. Representative for this discussion is the combination of Ifosfamide with Doxorubicin which gives an increased toxicity, but no change regarding the survival rate has been detected.

Conclusions

- The factors involved in the development and recurrence of the retroperitoneal liposarcoma remain unidentified.
- The imaging techniques help the preoperative assessment of the tumor, providing significant information regarding the localization, origin, content, boundaries and relationships with the other organs, their compression or dislocation and regarding the major vascular elements from the retroperitoneal space.
- The large size of the tumor which compresses and displaces the adjacent structures makes the surgical treatment the basis of the therapeutic conduct.
- Retroperitoneal liposarcoma represent a special situation that may justify an aggressive surgical approach, sometimes, including

multiple resections for repeated recurrences and even sometimes incomplete resections, in the absence of metastases.

▪ Currently, the histological type, the tumor size and localization are the elements which guide the general prognosis and provide data regarding the possibility of the occurrence of metastases, the recurrence rate and the survival rate.

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