

CASE REPORTS

Giant retroperitoneal sarcomas

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Abstract

Retroperitoneal sarcomas are rare malignant tumors, which are developing from mesenchymal stem cells residing in muscle, fat, and connective tissues. Underlying the rarity of this kind of tumors in general population, the aim of this paper is to present three cases of retroperitoneal sarcomas operated in Surgical Department of Military Hospital of Craiova (a retroperitoneal liposarcoma, a dedifferentiated liposarcoma and a malignant fibrous histiocytoma). From clinical point of view, we note the poverty of symptoms and non-specificity of these and a great tolerability of retroperitoneal space that offers the possibility for a great development of the tumor. CT-scan and MRI are the best investigations for diagnosis but surgical exploration is the best way for a good evaluation of these tumors. From histological point of view, we try to present new features about these kinds of tumors in order to classify them. Results of surgery correlated with complementary therapies were good without per-operative mortality or postoperative morbidity but we noted a recidive of one tumor (with different histological pattern) 21 months after the surgical intervention. The rarity of retroperitoneal sarcomas, combined with the vast array of histologic subtypes, has complicated our understanding of these tumors and impeded the development of effective therapies.

Keywords: retroperitoneal sarcomas, histopathology.

Introduction

Retroperitoneal sarcomas are rare malignant tumors, which are developing from mesenchymal stem cells residing in muscle, fat, and connective tissues [1].

In a general surgical clinic, the incidence of these tumors is very low but, on the other hand, these tumors are very spectacular because of those volumes and also because of their vast array of histologic subtypes.

The aim of this paper is to present three cases of retroperitoneal sarcomas operated in Surgical Department of Military Hospital of Craiova during the last four years (about 0.4% from all newly malignancies diagnosed in our clinic in this period), with the accent on morphology of these tumors. Comparing our incidence with the literature, we discover much similitude: about 8 600 new cases annually in the United States that represent less than 1% of all newly malignancies [2].

Clinical and pathological reports

Case no. 1

A female patient (B.V., 49 years) without significant pathological antecedents is presenting in our Clinic in August 2004 for lumbar pain without remission to antispasmodic usual drugs.

Clinically, we note grade II obesity and a tumor not clearly delimited in the right abdominal quadrants.

Laboratory showed higher inflammatory tests and 1.22 mg% creatinine. At sonography we observed a big retroperitoneal tumor that modifies normal position of left kidney which is included in tumoral mass. Right kidney is over dimensioned (144/55 mm) with pelivascular dilatations.

Tumoral process involved, too, right kidney lodge with anterior translation of the right kidney. Intravenous urography confirmed altered positions of the kidneys and also showed a normal urinary function for all two.

CT-scan with intravenous contrast fluid was the most sensitive test showing a tumor located in right abdominal quadrant, and that involved all retroperitoneal space from the right lobe of liver to the pelvic level; tumor structure is mixed with a tissular part located to the median line on about 20 cm and laterally, a fatty part that involved right kidney without possibility to delimitate it from the tumor.

Surgical intervention was transperitoneal with retroperitoneal access after right parietal-colic detachment. The tumor was giant, involving all right abdominal part from the Bogros space, of right inferior abdominal quadrant to the right lobe of the liver, including right kidney. After resection we achieved a big tumor of 35/30 cm (7.2 kg) with multiple lobes apparently delimited but non-uniform colored

(white-yellow to red), with hemorrhagic and necrotic zones that includes right kidney. Histology presents a dedifferentiated retroperitoneal liposarcoma (Figures 1 and 2).

Evolution was simple in postoperative period but, after 21 months, patient was readmitted in our clinic without symptoms but with a local recidive discovered at sonography. Surgical intervention was the same like for primary tumor and we discover a tumor with same structure (13/10 cm), with a thin capsule that was excised totally (2.2 kg). No differences from primary tumor in histology of this recidive (dedifferentiated retroperitoneal liposarcoma).

Case no. 2

A male patient (A.S., 68 years) was admitted in our Clinic in February 2006 with edemas at lower limbs, a vague pain and a tumor located in the right abdominal quadrants. At local exam, we could describe a diffuse abdominal tumor, with moderate pain at palpation but without tenderness.

Laboratory results were in normal limits but sonography describes a giant retroperitoneal tumor located in right abdominal quadrants.

CT-scan offers a better description for this tumor that seems to involve right kidney (with normal secretion and excretion) and right suprarenalian gland. An intimate contact with inferior vena cava was described, too, for a length of 2 cm.

Surgical intervention was transperitoneal with retroperitoneal access after right parietal-colic detachment. Tumor involves retroperitoneal space from liver to the pelvis involving, too, as in CT-scan description, right kidney and right suprarenalian gland. Detachment from inferior vena cava was simple and without incidents.

Structure was lipomatous clinical with a thin capsule around the most part of the tumor, especially in the superior part of it. After dissection, right kidney and right suprarenalian gland were in good conditions so we could leave its in place. Tumor achieved had 30/27 cm (5.6 kg), with multiple lobes with different structures especially lipomatous (Figure 3).

Postoperative evolution was simple patient leaving the hospital in good conditions after seven days. Few sonographic exams and a CT-scan discovered no local recidive until now (17 months after the intervention).

Case no. 3

A male patient (T.I., 78 years) without significant pathological antecedents was admitted in our Clinic in May 2007 for a big abdominal tumor observed at simple inspection (Figure 4).

No pain or other symptoms were described but he accused a reduction in body mass with about 6–7 kg in the last two months. Local exam discover a giant abdominal tumor, homogenous, that seems to translate all abdominal organs to anterior. Main laboratory findings were in normal limits but sonography showed a giant retroperitoneal tumor that involved not only the right part of the abdomen but a

left part too and translate the right kidney to anterior. CT-scan showed a relative delimitation of the tumor from the retroperitoneal organs (kidney, suprarenalian glands, and great vessels) and a normal function of both kidneys, too.

Surgical abord was transperitoneal with retroperitoneal access after right parietal-colic detachment. Tumor was developed in the right part of the retroperitoneal space involving left part too, but was delimited from other organs with a capsule. Structure was relatively homogenous, but with few lobes that impose some problems at dissection in order to excise all tumor (Figure 5).

It was excised totally and postoperative evolution was simple, patient leaving hospital in good conditions in a 7-th postoperative day. Histology showed a malignant fibrous myxoid histiocytoma (Figures 6–8). A sonographic exploration showed no tumor at two months after intervention.

Results

Postoperative evolution was good in all three cases presented. There were no per operative mortality and no postoperative morbidity.

Follow-up of these patients was recorded for a median time of 28 months and we registered a local recidive (case no. 1) at 21 months after first intervention (she developed a local recidive – 2.2 kg – delimited from the rest of retroperitoneal tissues and that could be excised completely) with the same histopathological structure as primary tumor.

Discussions

Approximately 15% of soft tissue sarcomas arise in the retroperitoneum, one third of malignant tumors that arise in the retroperitoneum being sarcomas [3].

Sarcomas are developing from mesenchymal stem cells residing in muscle, fat, and connective tissues, but the origin of these stem cells is still unclear [1].

Liposarcoma represent between 40 and 50% from all retroperitoneal sarcomas and is classified in a heterogeneous group of neoplasia of soft tissue, becoming from mesenchymal multipotent stem cells with special proliferation of lipoblasts but of other types of stromal cells, too [4].

There are two main theories that try to explain the origin of mesenchymal stem cells. First, suggest that the origin could be found in local tissue pools and the second sustain the origin in bone marrow [5].

Many classifications of the retroperitoneal tumors aim to include all tumors primary located in the retroperitoneal space; it was excluded metastases or other tumoral extension from neighborhood. In present, there are three principal groups of tumors [4]:

- tumors of the specialized or common mesoderm or mesodermal tumors – that represent over 65% from all primary retroperitoneal tumors and that are more frequent in adults, has an important volume, multiple lobes, relatively delimited and has an important risk for local recidive;

Figure 1 – Fibroliposarcoma: zone with proliferant pattern with elliptic cells with unclear margins and small cytoplasmatic amount (HE staining, $\times 100$)

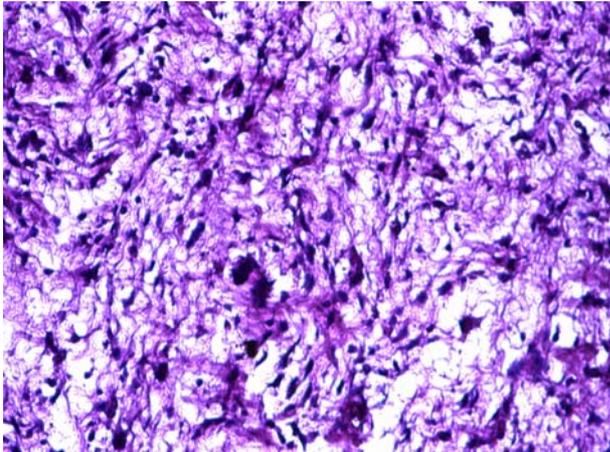
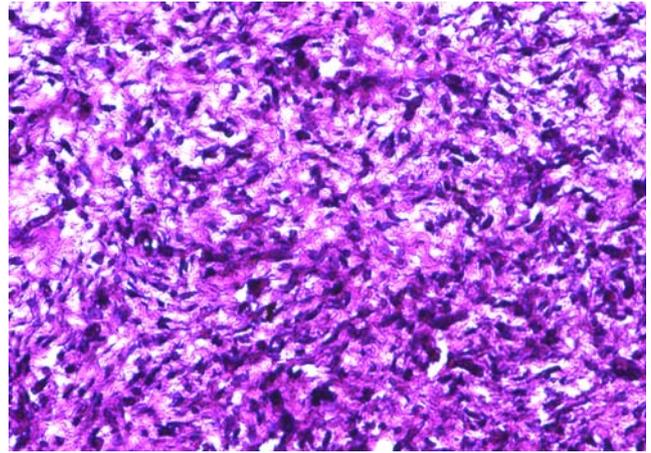


Figure 2 – Fibroliposarcoma: fibroblastic proliferation with plexyform capillary network (HE staining, $\times 100$)

Figure 3 – Necrobiosis (HE staining, $\times 100$)

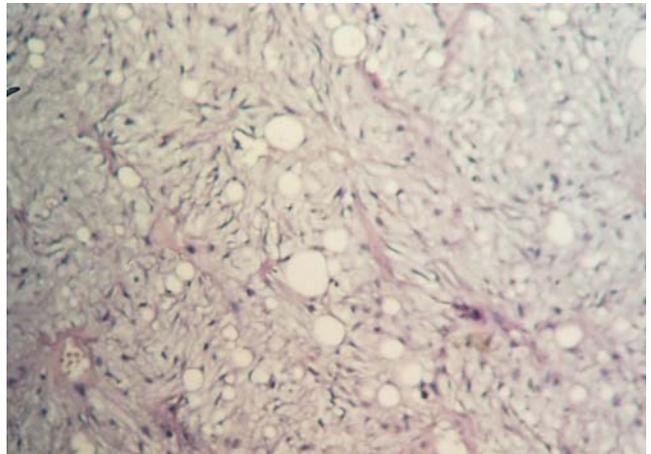
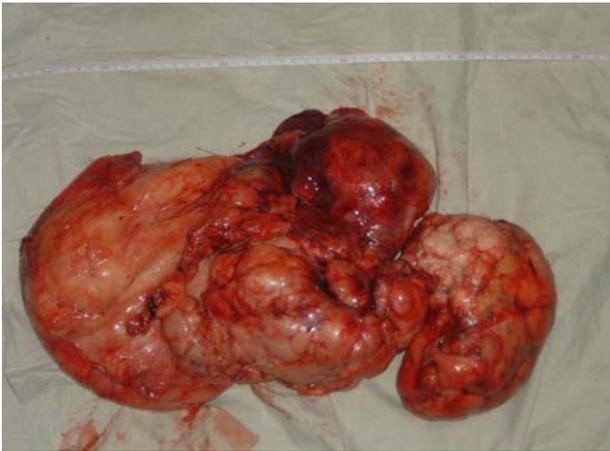
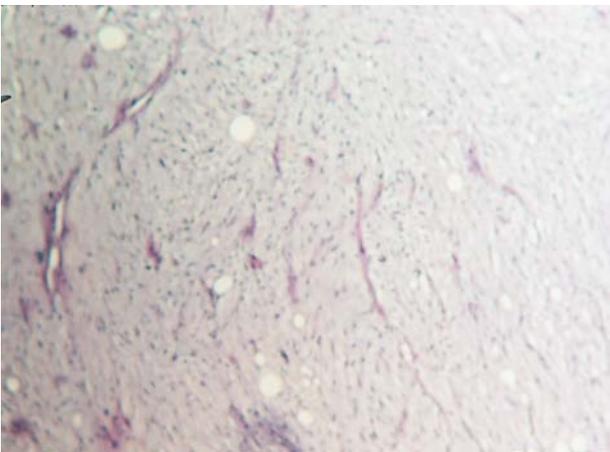
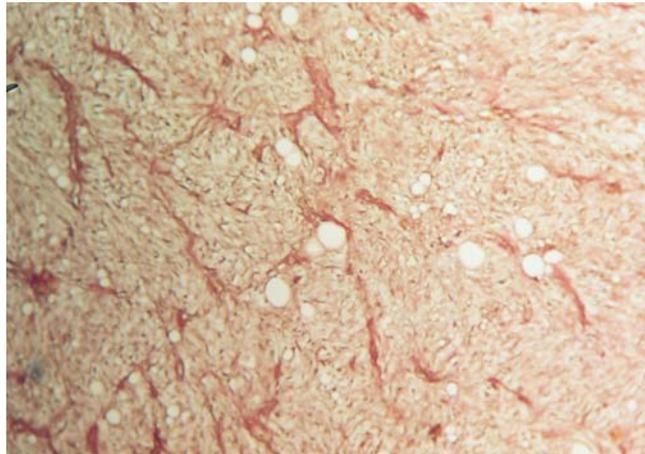


Figure 4 – Retroperitoneal tumor



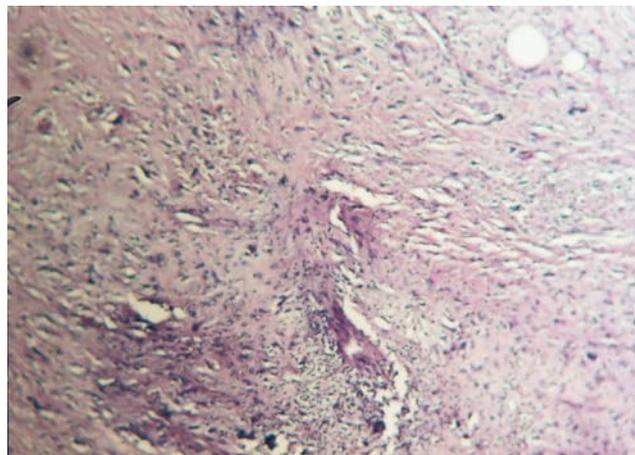
**Figure 5 – Malignant fibrous myxoid histiocytoma.
Macroscopic aspect**

**Figure 6 – Malignant fibrous myxoid histiocytoma:
zone with pleomorphic aspect with fibroblastic cells,
histiocytes and lymphocytic inflammatory infiltrate
(HE staining, $\times 10$)**



**Figure 7 – Malignant fibrous myxoid histiocytoma.
Myxoid type with arcuate vessels
(HE staining, $\times 10$)**

**Figure 8 – Malignant fibrous myxoid histiocytoma.
Myxoid zones with arcuate vessels
(Van Gieson staining, $\times 10$)**



- neuroectodermic tumors – more frequent in childhood;
- vestigial tumors – frequent in childhood, too.

Histological structure of mesodermal tumors could be homogenous to variable and includes tumors of adipose, muscular or vascular tissues and tumors becoming from non-differentiated or primary mesenchyma.

Liposarcoma is the most frequent sarcoma in adult patients. Histopathological features are variable, from the well-differentiated forms to others with excessive variability. Thus, dedifferentiated liposarcoma is characterized by coexistence of good and bad differentiated regions in the same tumor, or between the primary tumor and in metastatic or locally recidivated tumor. Bad differentiated regions could be similar with those from other types of retroperitoneal sarcomas like malignant fibrous histiocytoma or could be constituted from non-differentiated cells.

Malignant fibrous histiocytoma is a part of soft tissue fibrous-histiocytar tumors localized at the level of lower extremities, higher extremities or in retroperitoneal space. There are various histopathological aspects classified in five subtypes:

- storiform – pleomorphic malignant fibrous histiocytoma;
- myxoid malignant fibrous histiocytoma;
- giant-cell type;
- inflammatory malignant fibrous histiocytoma;
- angiomatoid type.

Association of myxoid and cellular regions characterizes our case (no. 3). An excess of acid mucopolysacharydes, storiform pattern becoming less clear, characterizes myxoid regions. On the other hand, vessels become more prominent, have thick-walled and often are curved with a wide arc and with tumor and inflammatory cells condensed along it. Seldom vessels could be very discrete with plexiforme distribution similar with myxoid liposarcoma. Cells from myxoid regions had all differentiation spectrums from well-differentiated fibroblasts to pleomorphic cells and polynuclear cells.

From clinical point of view, principal symptom was diffuse pain and presence of an abdominal mass. Retroperitoneal sarcomas have no sex or age prevalence, developing equally in men and women at any age.

Our cases suggest the presence of disease after the 50 years of age but the literature describes even younger patients. In almost all cases presented in literature (and in all our cases), retroperitoneal sarcomas was larger than 10 cm; tumors smaller than 5 cm are rarely diagnosed because of the patients that had no symptoms since they are larger in size [6–9].

Usually symptoms are related to the mass effect of the tumor or to local invasion of it being represented by the urinary symptoms, gastrointestinal obstruction, and lower extremity swelling or neurological signs.

Computed tomography (CT-scan) or magnetic resonance image (MRI) are the most useful tools for investigation and evaluation of retroperitoneal mass. These explorations demonstrated the retroperitoneal

origin of the tumor and also could assess the relationships with other organs (kidney, suprarenalian glands, liver, and vessels), but could even discover possible intra-abdominal metastasis or bone invasion. MRI offer a better evaluation of the retroperitoneum, few studies emphasized the importance of axial images in addition to sagittal and coronal views [1].

Once a retroperitoneal tumor has been identified, we must differentiate them from other retroperitoneal tumors like renal, adrenal or pancreatic tumors. A special attention must be paid for testicular examination (ultrasonography, and measurements of serum β -human chorionic gonadotropin) to exclude a testicular cancer with retroperitoneal metastasis. Surgical exploration is the most appropriate step for a retroperitoneal mass suspected of being a sarcoma [1, 10–12].

Because of heterogeneity of retroperitoneal sarcomas is difficult to establish a good prognostic score. To obtain it is necessary to study sufficient numbers of patients with sarcomas and to combine data from patients who were treated over several decades, who underwent markedly different treatment regimens, and who had histologic subtypes with dramatically different clinical behaviors [1].

If tumor size has not been identified as a predictor of survival since virtually all retroperitoneal sarcomas are larger than 5 cm at presentation, tumor grade has been reported as a significant factor in some studies [9, 13–17].

Conclusions

Retroperitoneal sarcomas are rare clinical entities that have varying clinical courses depending on their histologic subtype and grade.

The rarity of retroperitoneal sarcomas, combined with the vast array of histologic subtypes, has complicated our understanding of these tumors and impeded the development of effective therapies.

Because of great tolerability of retroperitoneal space these tumors has a long asymptomatic evolution that is correlate with its dimensions.

Even CT-scan and MRI offer a lot of information for diagnosis; surgical exploration is the most important step for evaluation of retroperitoneal tumors.

Surgery is the most important factor in retroperitoneal sarcomas treatment, followed by other complementary therapies.

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