CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 1050–1053

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Giant retroperitoneal leiomyosarcoma. Multiorgan block removal

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ARTICLE INFO

Article history:
Received 3 July 2014
Received in revised form
12 September 2014
Accepted 15 October 2014
Available online 31 October 2014

Keywords:
Retroperitoneal
Leiomyosarcoma
Multiorgan resection

ABSTRACT

INTRODUCTION: Retroperitoneal tumors are rare, mostly malignant. Locally aggressive, and more frequent in women in their 5th decade of life. Its symptoms are nonspecific, including abdominal pain and palpable mass. To diagnosis is helpful computed tomography and biopsy. It needs surgery for absolute healing.
PRESENTATION OF CASE: 67 years old man was admitted with back pain and fever. Abdominal imaging tests showed a 15 cm abdominal mass without clear organodependencia. Endoscopy with biopsies evidenced mesenchymal neoplasia of undetermined origin. In surgery we confirm its resecability and was necessary multiorgan resection. *Pathologic diagnosis*: well differentiated retroperitoneal leiomyosarcoma. Started adjuvant radiotherapy. In subsequent tests showed the presence of liver metastases.
DISCUSSION: Retroperitoneal tumors are developed from nerve, vascular, muscular, connective, supportive and fibroareolar tissue from this space. Its size does not modificate survival or resectability. We used TC and biopsy for its diagnose. Adjuvant therapy does not affect survival or quality of life, surgery remains the only curative option. Locoregional recurrence is the most influential figure in the prognosis. A large percentage of patients required a second surgery (between 45 and 82%).
CONCLUSION: The only curative option of retroperitoneal sarcomas is surgery, which usually requires multiple organ resection. Chemotherapy and radiotherapy are mostly a surgical supplement. Chemotherapy has not shown significant increase in survival.

1. Introduction

Retroperitoneal tumors represent a rare entity, most are malignant and sarcomas are most commonly. Sarcomas are malignant tumors derived from embrionary mesoderm that arise from skeletal and extraskeletal connective tissues, including the peripheral nervous system. The majority of soft tissue sarcomas present in the extremities; however, many other sites can be affected, including the retroperitoneum. Leiomyosarcomas are second in frequency at this location, and also one of the most aggressive. More frecuent in women between the fourth and sixth decades of life.[1] Most patients debut with abdominal pain, followed by palpable abdominal mass. Symtomps of local compresion are less frequent. The diagnosis is usually given by computerized tomography (CT) and biopsy.[2]

The only effective treatment known at the time is surgical resection R0, since chemotherapy is used only in metastasis, and radiotherapy is a surgical complement to decrease locoregional recurrence risk. To obtain surgical safe margin, is often necessary the resection of another adjacent infiltrated organs.[3] Recurrence, which is the most important prognostic factor in these patients, is very common.

We present a case of retroperitoneal leiomyosarcoma where was necessary multiorgan resection for its treatment.

2. Presentation of case

A 67 year old male was admitted for study of fever, back pain, constitutional symptoms and general illness with several months of evolution. During hospitalization the patient required repeated transfusions because of progressive anemia, needing more than twelve packed erythrocytes.

Physical examination revealed an abdominal mass in left upper quadrant, hard and fixed to deeper layers.

Ultrasound showed a enlarged spleen (14.5 cm) with difficult to access to splenic hilum due to a 15 cm diameter, heterogeneous and multi-lobed mass. Located above and medial to left kidney upper pole and lateral to aorta, in intimate contact with posterior spleen face. No involvement or liver metastasis was observed.

The admission CT report a tumor in left upper quadrant of 17 cm in diameter, heterogeneous hypo-and hyperdense areas and gas in superoanterior portion. Infiltrates spleen, left kidney and adrenal gland, colon and pancreas, with no clear dependence of any of

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http://dx.doi.org/10.1016/j.ijscr.2014.10.061

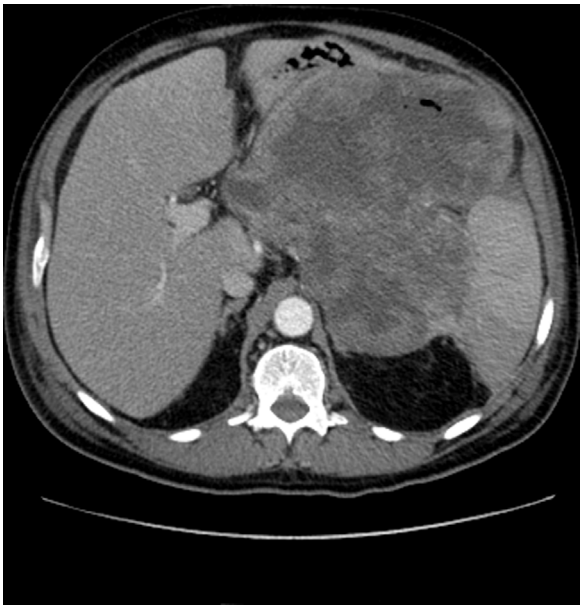

Fig. 1. Abdominal CT showing tumor located between spleen and stomach with retroperitoneal infiltration.

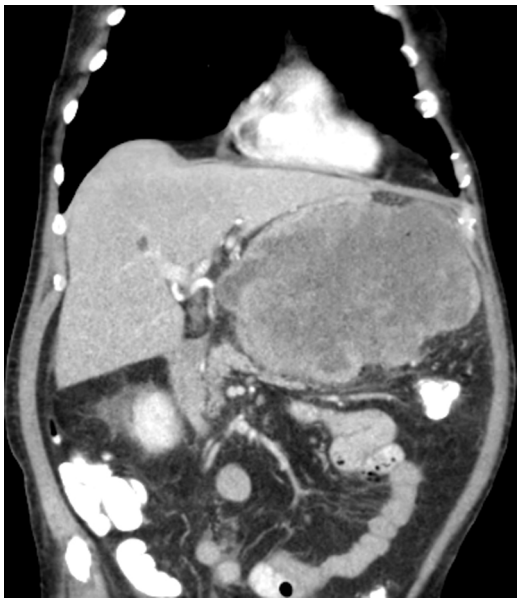

Fig. 2. Coronal CT image demonstrating the involvement of the whole area corresponding to left upper quadrant of the abdomen.

these structures. The patient had gastrointestinal bleeding, thus we did oral endoscopy with biopsy, which evidenced ulceration of the gastric mucosa by extrinsic neoplasia. Biopsies were informed as fusiform cell proliferation and mesenchymal neoplastic nature, unable to determine its origin.

Debating exposure the case at the digestive tumors interdisciplinary committee, it was decided practise an open mass biopsy, after which we had patologycal diagnosis: well differentiated leiomyosarcoma. We decided not to perform percutaneous biopsy because of the risk of spread and the low diagnostic sensitivity of this technique in our hospital.

The patient was programed to an exploratory laparotomy to decided tumor resectability.

Planned surgery is based on the CT image, showing a dissection plane starting laterally to the left side of the spleen, behind the pancreas neck, and including part of the colon with the transverse mesocolon and stomach. During surgery we confirmed this was possible and practiced block tumor removal, including full gastric resection, spleen, pancreas and 20 cm distal transverse colon. Intraoperative retroperitoneal tissue biopsy showed no infiltration in renal or adrenal area.

We obtained a diagnosis of well-differentiated retroperitoneal leiomyosarcoma that ulcerated stomach wall and infiltrated spleen, pancreas and transverse colon serosa. Mitotic index of 15 mitoses per 50 high power fields and surgical margins were free of disease. Metastasis was not noted on any of the lymph nodes removed.

Postoperatively, the patient presented a left retroperitoneal abscess fistulized to preexisting renal cyst, which require percutaneous drainage for its resolution.

As adjuvant treatment, the patient started radiotherapy. In subsequent image test, nine months later, there were evident liver lesions, and proceeded to fine needle puncture-aspiration ultrasound guided, confirming that those corresponded to metastasis. Thus began chemotherapy treatment with gemcitabine and docetaxel, but lesions have not size reduction after six sessions, so the patient currently continues with palliative chemotherapy and is followed by Oncology Service. The patient died 23 months after surgery (Figs. 1–5).

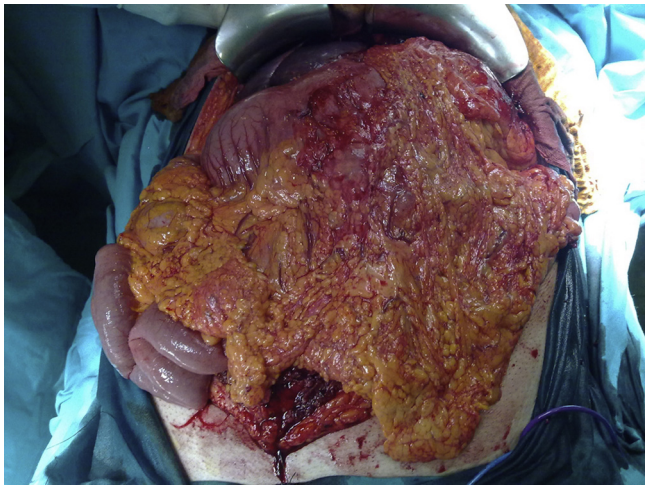

Fig. 3. Surgical field showing the affected area and the absence of local spread or multiple peritoneal involvement.

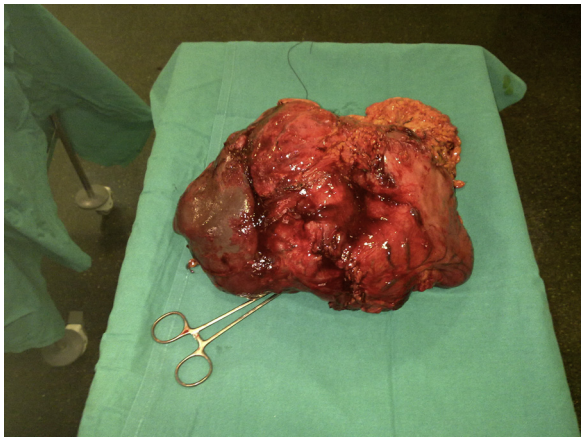

Fig. 4. Piece of surgical resection including stomach, spleen, left transverse colon and pancreatic tail.

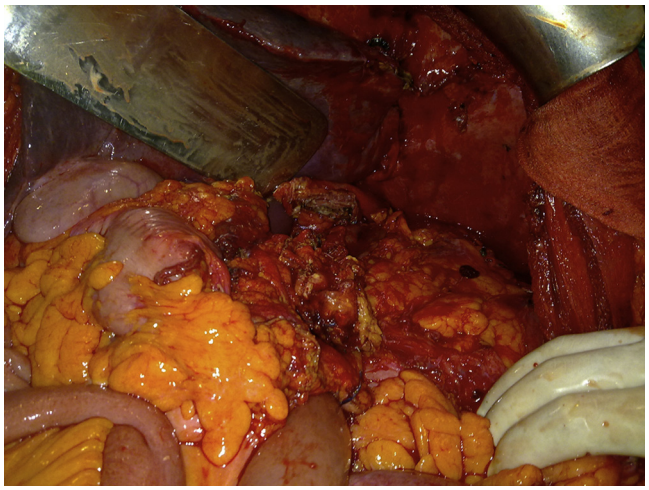

Fig. 5. Surgical field when resection was completed.

3. Discussion

The retroperitoneum is defined anteriorly by the peritoneal extensions anchoring the transverse colon, the small bowel, and the ascending and descending colon. The posterior aspect of this space is a muscular wall comprising the psoas major and minor, the quadratus lumborum, obturator internus, pyriformis muscle and the tendinous portion of the transversus abdominis muscle. The retroperitoneum is bordered superiorly by the diaphragm, inferiorly by the levator ani muscles, and laterally by the ascending and descending colon in combination with the peripheral margin of the quadratus lumborum muscles.

Retroperitoneal tumors are defined as those that are formed from nerve, vascular, muscular, connective, supportive and fibroareolar tissue, excluding organs and great vessels that are in this space. At this level 85% of tumors are malignant, and of these, 50% are sarcomas.[4] The most common subtypes of sarcomas at this level are liposarcoma, with the best prognosis, and leiomyosarcomas, one of the most aggressive.

The average size of these tumors at diagnosis is 16 cm, since they do not start giving symptoms until they reach 10 cm in diameter. The retroperitoneum has adaptability, allowing tumor growth without the rest of the body perceiving changes. Its growth is slow and insidious. However, size is not an impediment to total resection neither data for worse survival.[5] Tumor growth is centrifugal, being the most immature cells in the outer layer of the tumor, compressing surrounding structures.

The symptoms presented by these tumors are nonspecific and varied, depending on the affected organ and its location.[6] The most common symptoms are abdominal or back pain, and palpable abdominal mass. Usually also present gastrointestinal disorders, urinary discomfort, lower limb edema, sweating or constitutional symptoms.

For the diagnosis, the most sensitive imaging test and higher performance is CT. It can help determinate histological tumor type because of the indirect data (necrotic areas, fat or muscle tissue),[7] helps to delineate the tumor and its relationship to other structures, distinguishing between invasion or contact; staged the tumor, because it shows node and distant metastases, and sometimes provides information of the origin of the tumor. puncture/biopsy is commonly used for diagnosis, but is more reliable if it is guided by image tests, CT, as otherwise can be inconclusive.[3]

The use of adjuvant therapy has not demonstrated to increase survival neither provide a better quality of life. Surgery is the only procedure that has proven to be curative.

It needs a complete microscopic tumor resection R0 for an acceptable survival as locoregional recurrence is the most influential data in the prognosis. Recurrences can occur more frequently in incomplete resections (macro or microscopic). For complete tumor resection is necessary removing neighbors organs by more than 50% of the time, the most frequently resected organs are kidney and adrenal gland, but pancreas, spleen, liver, colon, duodenum and cava are also affected on numerous occasions. These resections imply increased morbidity though to mortality, and longer and more complex surgeries. Pancreatic manipulation (total sections and especially partial) increases the risk of complications especially postoperatively. Sometimes different specialists are required for resection the different infiltrated structures by the tumor. Resectability tumor can only be confirmed during surgery, and when it is not possible, is usually to do extensive infiltration of the mesenteric root, neurovascular extensive infiltration, distant metastasis or peritoneal sarcomatosis.[8]

Complete resection was achieved in only 39% of cases in some series.

Nodal metastases and tumor size, have not prognostic significance, provided they can be removed in opposite to distant metastases, which are usually caused by hematogenous dissemination. The presence of distant metastasis should not suggest a palliative treatment, if it can be completely removed both the original tumor and metastasis. Resection of metastases after surgery is necessary if you want to increase patient survival.[9]

A large percentage of patients with R0 resection require rescue surgery (between 45 and 82%).[10] Posterior median survival is under 10 months. In nonresecable tumors patients, the median survival is lower than 6 months. Patients with complete resection, which do not require subsequent surgery have an average survival of 5 years.[11]

4. Conclusions

Currently, surgery is the only “curative” option for retroperitoneal sarcomas. Before surgery, image tests are needed to delineate the tumor and the affected organs to prepare a good surgical strategy that sometimes require more than one specialist. Because the tumor at diagnosis is usually voluminous, wide approaches are preferred, especially if it requires multiple organ resection.

Aggressive surgical treatment is justified only in patients in which complete tumor resection is feasible. In our patient, the resection was carried out with curative intent to attempt to improve his quality of life severely limited by persistent anemia. The patient survived 23 months after surgery with an acceptable quality of life.

Chemotherapy and radiotherapy are in most cases a surgical complement although chemotherapy has not demonstrated a significant increase in survival.

Conflict of interest

Any authors have not conflicts of interest.

Funding

We have not any source of funding for our research.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contributions

Dr. Mateo Vallejo, Dr. Medina Achirica and Dr. Esteban Ramos participated in the surgical intervention and patient monitoring. Dr. Dominguez Reinado wrote this case report. Dr. Melero Brenes and Dr. Diaz Oteros search bibliography of this case.

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