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Giant retroperitoneal leiomyosarcoma. Multiorgan block removal

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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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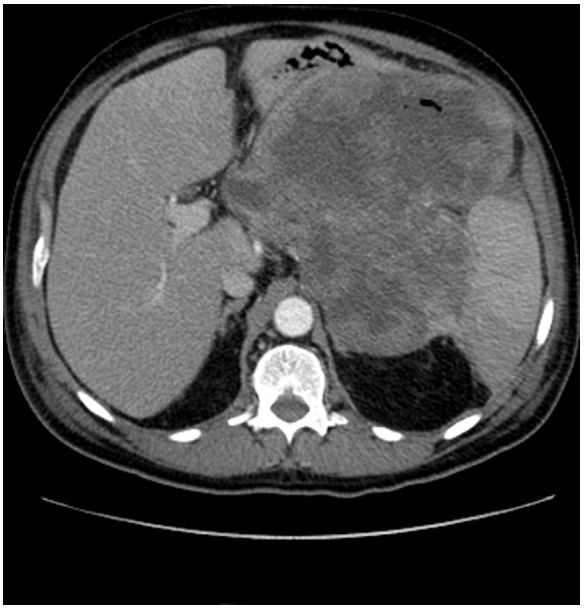

Fig. 1. Abdominal CT showing tumor located between spleen and stomach with retroperitoneal infiltration.

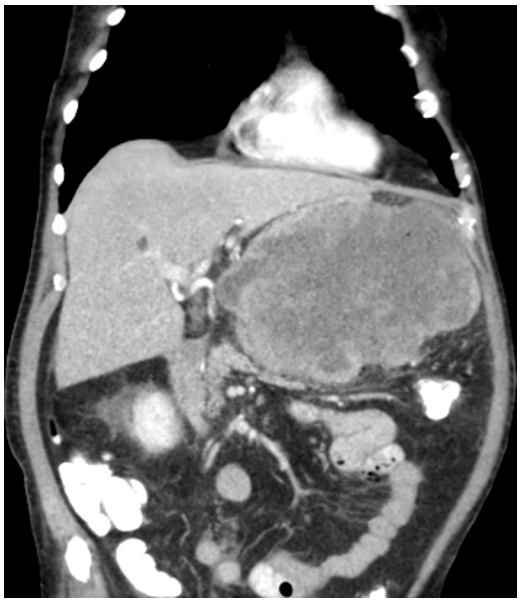

Fig. 2. Coronal CT image demonstrating the involvement of the whole area corresponding to left upper quadrant of the abdomen.

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).

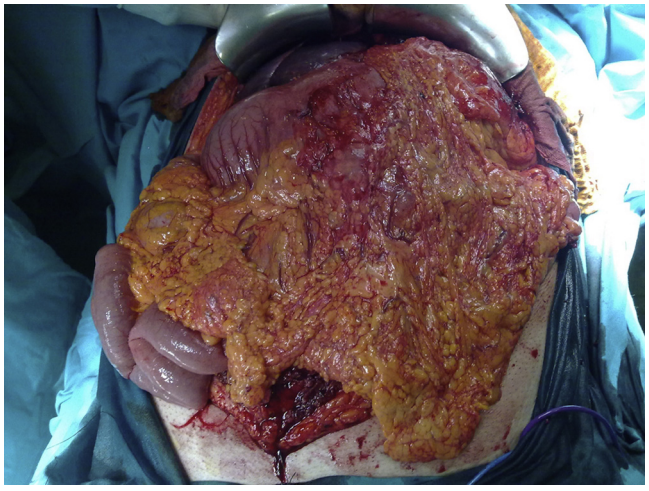

Fig. 3. Surgical field showing the affected area and the absence of local spread or multiple peritoneal involvement.

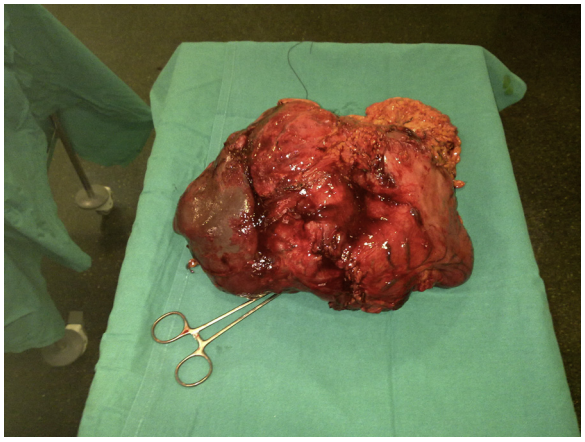

Fig. 4. Piece of surgical resection including stomach, spleen, left transverse colon and pancreatic tail.