ABSTRACT

Primary retroperitoneal tumors are a rare heterogeneous group of neoplasms. Treatment involves complete macroscopic resection of the tumor with free margins. Currently, laparoscopy has become an interesting approach for treating either benign or malignant solid tumors. Major advantages are less postoperative pain, fast recovery, low morbidity and good cosmetic outcome. The authors present the case of a 47-year-old woman with a small retroperitoneal leiomyosarcoma (5 cm) who underwent successful laparoscopic tumor resection. She followed a good postoperative course without complications. She was discharged from the hospital on the first postoperative day (24 hours after the procedure). The patient is alive and showed no signs of tumor recurrence during a twelve-month-follow-up period. Therefore, laparoscopy seems to be a good option for treating small retroperitoneal sarcomas. This procedure avoids a large incision, inflicts less postoperative pain, and promotes rapid recovery and good cosmetic results.

Key words. Laparoscopic surgery; retroperitoneal sarcoma; leiomyosarcoma; soft tissue neoplasms; immunohistochemistry.

RESUMO

Ressecção laparoscópica de leiomiossarcoma retroperitoneal: relato de caso

Os tumores retroperitoneais representam um grupo heterogêneo raro de neoplasias. O princípio do tratamento é a ressecção completa macroscópica do tumor com as margens livres. A via laparoscópica tem se tornado abordagem interessante no tratamento de tumores sólidos benignos e malignos. As maiores vantagens são baixa morbidade, menos dor pós-operatória, rápida recuperação e bom resultado cosmético. Os autores apresentam mulher de 47 anos com pequeno leiomiossarcoma retroperitoneal (5 cm), submetida a ressecção laparoscópica do tumor com sucesso. A paciente apresentou evolução pós-operatória sem complicações e recebeu alta hospitalar 24 horas após o procedimento. Atualmente, está viva sem sinais de recidiva do tumor aos doze meses de seguimento. O acesso laparoscópico parece uma boa opção para o tratamento de pequenos sarcomas retroperitoneais.
INTRODUCTION

Primary retroperitoneal tumors are a rare heterogeneous group of neoplasms that are most often malignant (85%). Sarcoma is the most common lesion (around 50%) among malignant tumors. Liposarcomas are the most common histologic subtype, followed by leiomyosarcomas.1 Retroperitoneal sarcomas are usually an insidious disease. They have no specific clinical symptoms or signs and are mainly asymptomatic. Diagnosis is usually incidental and is reached by means of imaging studies. When at an advanced stage, sarcomas can be large, thus becoming symptomatic. The most common manifestations are abdominal or back pain, palpable abdominal mass, and constitutional symptoms like anemia and weight loss. Patients may also show signs of bowel or ureteral obstruction if the tumor spreads to surrounding organs.1,2 Abdominal computed tomography or magnetic resonance imaging are appropriate to determine tumor size and invasion of adjacent structures.3 Biopsy should not be done if retroperitoneal sarcoma is suspected, given the risk of transperitoneal spread and implantation.4

Surgery still remains the first-line treatment of retroperitoneal sarcomas, representing the only potentially curative therapy. The primary aim of surgery is to completely excise the tumor with negative margins (R0-R1). A major prognostic factor is complete resection of the tumor.5 The benefit of surgical resection is related to American Joint Commission on Cancer (AJCC) stages I to III (65 to 76% risk reduction for mortality).6 Many centers have reported complete resection rates for primary retroperitoneal tumors (over 90%), leading to greater long-term survival.3 Confirming diagnosis is usually made by pathological and immunohistochemical analysis.

Laparoscopic resection of retroperitoneal tumors is emerging as a potential surgical option in select cases.7 The enthusiasm for laparoscopy is based on several advantages, such as smaller incisions with better aesthetic results, improved visualization of abdominal and pelvic cavities, minimal blood loss, lower rate of postoperative complications, decreased surgical site infection, shorter hospital stay, reduced pain and quicker return to normal activities.

Unfortunately, most retroperitoneal sarcomas will recur. The 5-year survival rate is reported to be around 40 to 74% for patients who underwent complete surgical resection.7 Recidive occurs in approximately 47% of patients, often within a 2-year follow-up period.7 Local recurrence can still be treated by subsequent surgical resection that can lead to second complete resection and allow survival.7 The most important prognostic factor associated with survival is the radicality of the surgical resection. Tumor size and recurrence of sarcoma do not seem to be independent predictors of death.8–11 Postoperative radiotherapy may control local recurrence and has been recommended for patients with microscopically comprised margins.12 A few reports have been published about laparoscopic resection of retroperitoneal sarcomas.13,14

To date and to our knowledge, there is no report of laparoscopic surgical treatment of retroperitoneal sarcoma in Brazil. The authors report the case of a patient with retroperitoneal leiomyosarcoma which was successfully treated with complete laparoscopic resection done by a single surgical team.

CASE REPORT

A 47-year-old asymptomatic woman was referred to the surgical oncology service for management of a retroperitoneal tumor. A solid heterogeneous retroperitoneal mass had been previously and incidentally detected on abdominal computed tomography on routine screening. The mass was estimated to be...
around 4 cm in diameter and was located between the left infrarenal area and psoas muscle (Figure 1). Colonoscopy was reportedly normal and serum tumor markers were negative. Distant metastases were not observed on thorax computed tomography. Her medical history included three previous surgeries. These surgical procedures were a cesarean delivery (twenty years ago), an aesthetic dermolipectomy (five years ago), and laparoscopic cholecystectomy due to symptomatic cholelithiasis (one year ago). On physical examination, a mobile and hard mass was palpable 5 cm deep into the left flank.

Therefore, the preoperative diagnostic hypothesis was retroperitoneal solid tumor of unknown origin (probably a retroperitoneal schwannoma), and total resection of the lesion by laparoscopic approach was indicated.

The surgical procedure was performed with four trocars: a 10-mm trocar in the epigastrium (a 30-degree laparoscope was used), a 10-mm trocar in the umbilicus, an 11-mm trocar in the hypogastrium, and a 5-mm trocar in the left flank. During abdominal cavity inspection, a retroperitoneal mass measuring 5 cm was observed. It was a hard and mobile tumor, situated below the left infrarenal area and along the left psoas muscle, without invasion of the sigmoid colon. Moreover, no lymphadenopathy was noticed.

Left-sigmoid mesocolon was thus incised with Ultracision® device. Inferior mesenteric vessels and left ureter were isolated and preserved. Complete resection of the mass was performed with 1 cm tri-dimensional margin excision (Figure 2). The mass was entirely removed into an endobag through a previous small infra-umbilical transversal incision (Pfannenstiel), according to oncologic surgical principles. There was no resection of adjacent organs.

Postoperative course was carried out without complications. Oral feeding was started 12 hours after surgery and the patient was discharged from the hospital on the first postoperative day (24 hours after procedure).

Histological analysis revealed a high-grade malignant mesenchymal tumor with nuclear atypias and an elevated mitotic rate (25 mitosis per 10 high power fields).

Figure 1. Computed tomography showing retroperitoneal sarcoma

Figure 2. Aspects of laparoscopic dissection of retroperitonal sarcoma (white – sarcoma; blue – left colon, sigmoid colon; green – left ureter).
The immunohistochemical study was positive for desmin and actin, confirming the final pathological diagnosis of leiomyosarcoma (Figures 3 and 4). All resected margins were macroscopically negative ($R_1$). Adjuvant radiotherapy was performed. The patient is alive without signs of tumor recurrence (twelve-month follow-up period).

**DISCUSSION**

Retroperitoneal sarcomas are rare mesenchymal neoplasms accounting for 0.1–0.2% of all cancers and about 10–20% of all soft-tissue sarcomas.\(^2,4\) The most prevalent histological type is liposarcoma (41–55%), followed by leiomyosarcoma (8–18%), and malignant fibrous histiocytoma.\(^2,3,6\) Soft-tissue sarcomas are usually considered to be sporadically acquired lesions, although some familial disorders have been described, such as retinoblastoma, neurofibromatosis, and Li-Fraumeni syndrome. Soft-tissue sarcomas may occur at any age, with most diagnosis being made when patients are between 54 to 65 years old. No gender or race predilection has been reported.\(^1\)

Differential diagnosis often includes gastrointestinal stromal sarcoma and benign disorders, such as schwannoma, paraganglioma, lipoma, lymphangiofibroma, ganglioneuroma, and desmoid tumor.\(^2,4,7\)

In the case reported, the patient was asymptomatic with an abdominal computed tomography finding of a retroperitoneal mass of unknown origin, characterizing a typical incidental discovery, as most cases described in the literature.\(^2,4,6,7\) Complete surgical resection of retroperitoneal tumors represents a major treatment option and the most important prognostic factor for survival. However, it can be a challenge, even for experienced surgeons, because tumors are close and can involve retroperitoneal vital structures.\(^13\) Multi-organ and vascular resections may be required (in approximately 50% and up to 18% of cases, respectively), often determined intraoperatively, requiring en bloc resection.\(^1,7\) Organs frequently resected due to infiltration include kidney (32–46%), colon (25–30%), adrenal gland (18%), pancreas (15%), and spleen (10%).\(^1,3,5\)

According to what has been reported in the literature, 1–3% perioperative mortality and 8–44% complication rates have been associated to open surgery. Postoperative complications may include infection, anastomotic leak, intra-abdominal abscess, hemorrhage, small bowel obstruction, myocardial infarction, atrial fibrillation, and deep venous thrombosis. The median length of hospital stay has been described as seven days. Tumor size ranging from 2 to 48 cm has been reported.\(^3\)
In this case, given that preoperative imaging was not suggestive of adjacent organ involvement, the patient was thus referred for laparoscopic resection. Complete resection was feasible with macroscopically negative margins (R1), with an excellent oncological, functional, and aesthetic outcome. There were no intraoperative injuries. Moreover, there was minimal blood loss, with no postoperative complications. The patient was discharged from the hospital 24 hours after surgery. The advantages of laparoscopic resection of retroperitoneal mass were detailed in a few reports in the literature. By means of laparoscopic approach, a retroperitoneal tumor can be completely removed through a minimally invasive procedure, even in a deep location. Another advantage associated with laparoscopic procedure includes the possibility of a second look in the case of disease recurrence.

One of the major limiting factors of laparoscopy is infiltration to surrounding vital structures. It seems that tumor size is not a contraindication to laparoscopic surgery. Current literature has described retroperitoneal tumors up to 12 cm in diameter that were successfully resected by laparoscopy. However, large and heavy tumors are expected to be more difficult to manage via laparoscopic technique.

Although laparoscopy has been described as a safe and effective approach for the management of retroperitoneal malignant mass, it is not yet a consensus. The risk of tumor dissemination cannot be ruled out. Further studies involving a greater number of cases may be necessary for its definitive indication.

In the present case, leiomyosarcoma was the histopathologic type. It represents the second most common subtype of retroperitoneal sarcomas. Despite the absence of scientific evidence to support the use of adjuvant radiotherapy in the management of retroperitoneal sarcomas, we have decided to recommend it due to microscopically comprised margins that have been associated with high local recurrence.

In conclusion, complete surgical resection is the mainstay of treatment of sarcomas of the retroperitoneum. Laparoscopic approach seems a good option for the treatment of these tumors in selected cases (small tumors), such as the one described in the present report. This approach is a safe, feasible, and effective alternative. Therefore, it does not violate oncological principles when it is performed by a skilled laparoscopic team.

**REFERENCES**