


Retroperitoneal Neoplasia

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Abstract

The retroperitoneal space is the anatomical region located in the lower abdomen, between the posterior parietal peritoneum and the fascia of the lumbar musculature, from the underside of the diaphragm to the pelvic floor. At the front, it continues with the posterior surface of the liver, the duodenum-pancreas area and part of the ascending and descending colon.

Sarcomas comprise one-third of retroperitoneal tumours, with two predominant histological subtypes, namely liposarcoma (70%) and leiomyosarcoma (15%).

Imaging is used to confirm the diagnosis, assess the presence of metastasis and determine the possibility of resection, with Computed Tomography (CT) being the most commonly used tool.

This case describes a 65-year-old female patient who presented with a giant, palpable abdominal mass that had been developing for several months. She underwent surgery and the mass was removed. Histopathological examination of the tumour mass revealed a well-differentiated liposarcoma with a mixed sclerosing and infiltrative variant.

Keywords: Lower Abdomen; Computed Tomography; Tumour

Introduction

Retroperitoneal tumours are defined as a group of neoplasms that originate in the retroperitoneum. Most are derived from mesenchymal, neurogenic or embryonic cells and are usually detected when they have reached a large size. Most are malignant in nature; three types are defined in their classification: liposarcoma, leiomyosarcoma and

malignant fibrous histiocytoma. Retroperitoneal Soft Tissue Sarcoma (STS) usually occurs in patients around 50 years of age, but it can occur at any age. Retroperitoneal STS is diagnosed with equal frequency in men and women. Liposarcoma is the most common sarcoma, with a higher incidence in women aged 50 to 70 years. It is subdivided into well-differentiated, myxoid and pleomorphic types. Well-differentiated liposarcomas that arise in the body wall/trunk or limb are called atypical lipomatous tumours, as they tend not to metastasise to distant organs and the prognosis is favourable.

Most dedifferentiated liposarcomas are high-grade tumours (grade 2 or 3). Compared to well-differentiated low-grade liposarcomas, they have higher local and distant recurrence rates (distant recurrence rate of 20 to 30% versus 0%) and a six-fold increased risk of death. Leiomyosarcoma is the most common malignant tumour and predominates in middle-aged women, characterised as large soft tissue masses with extensive areas of necrosis. Malignant fibrous histiocytoma is the most common soft tissue sarcoma, predominantly in older men, characterised by large, infiltrating solid masses, areas of necrosis and the presence of dystrophic calcifications [1].

Retroperitoneal soft tissue sarcoma generally does not cause symptoms until the tumour reaches a size sufficient to compress or invade surrounding anatomical structures. Most tumours are already large and locally advanced when initially detected, with an average size at diagnosis of approximately 15 cm.

Some patients may present with an incidental abdominal mass that is asymptomatic or minimally symptomatic. Others may present with abdominal pain or symptoms related to the mass effect of the tumour on surrounding structures, such as lower limb oedema due to compression of retroperitoneal structures, referred pain due to compression of neighbouring structures, weight loss due to local invasion of the gastrointestinal tract, serous ascites due to portal venous compression; It may also be accompanied by fever, night sweats, and, in rare cases, paraneoplastic hypoglycaemia, which is usually secondary to tumour production of large insulin-like growth factor 2.

On physical examination, abdominal exploration may reveal an abdominal mass. Other possible findings may include ascites, hepatic masses or lower limb oedema. Male patients with a retroperitoneal mass should also undergo a testicular examination to assess for the presence of a mass, as testicular cancer can metastasise to the retroperitoneal lymph nodes and retroperitoneal tumours can spread through the inguinal canal to the scrotum. The first signs are often an increase in abdominal circumference and a palpable mass. Patients often report a feeling of gastric fullness and difficulty eating due to gastric and digestive compression. There is often a discrepancy between the volume of the tumour and the lack of symptoms when the mass is detected. The average size at diagnosis is approximately 20 cm and smaller masses are often detected incidentally during diagnostic tests for unrelated symptoms [2].

Diagnostic imaging is one of the most important tools for defining the diagnostic and therapeutic approach, with plain and contrast-enhanced tomography being of great importance. In patients with localised primary retroperitoneal sarcoma, resectable disease and no evidence of distant metastasis on staging imaging studies, surgery with complete macroscopic resection offers the only possibility of cure; there is no clear role for radiotherapy in the vast majority of patients with resectable retroperitoneal sarcoma. For selected patients with respectable disease and specific histologies at high risk of local recurrence (well-differentiated liposarcoma and low-grade dedifferentiated liposarcoma), we suggest preoperative (i.e., neoadjuvant) radiotherapy plus surgery rather than surgery alone, as data suggest a lower risk of locoregional recurrence with this approach. However, surgical resection alone is an appropriate alternative, as randomised trials evaluating preoperative radiotherapy have not demonstrated an overall survival advantage over surgery alone.

In the case of tumours of mesenchymal origin, the first-line treatment is surgical resection. For adrenal tumours, treatment depends not only on whether the tumour is benign or malignant, but also on whether it is functional or non-functional and on its size. Benign non-functional tumours smaller than 2.5 cm are monitored. The rest, non-functioning benign tumours larger than 2.5 cm, functioning benign tumours or malignant tumours, are treated by surgical excision. In malignant retroperitoneal tumours, it is possible to optimise survival and reduce locoregional recurrence through surgery aimed at achieving disease-free margins, which often requires extensive and combined resections of neighbouring organs. The status of the tumour after treatment is described by the Residual tumour (R) classification, which defines how complete the surgical resection of a malignant neoplasm was.

R0 resection indicates a resection with negative microscopic margins in which no macroscopic or microscopic tumour remains in the primary bed of Retroperitoneal Sarcoma (RPS). R1 resection indicates removal of all macroscopic disease, but the microscopic margins are positive for RPS. R2 resection indicates macroscopic residual disease with a macroscopic residual tumour that was not resected with macroscopic involvement [3]. Currently, there is no treatment available that is as effective as surgery for these patients. Since the first publications on this disease, the therapeutic approach has been primarily surgical [1].

In recent years, in addition to surgical treatment of retroperitoneal sarcoma, there has been increased interest in the usefulness of perioperative radiotherapy. In a matched case-control analysis using data from the national clinical oncology database jointly administered by the American Cancer Society and the American College of Surgeons, the addition of perioperative radiotherapy to surgical treatment was shown to be associated with a better prognosis compared to surgery alone. Both patients who received radiotherapy before surgery and those treated with radiotherapy after surgery had better clinical outcomes than those treated with surgery alone. These findings suggest that perioperative radiotherapy may provide an additional benefit in the management of these patients. However, retroperitoneal sarcomas are characterised by a poor prognosis, with limited survival and a high recurrence rate. Overall five-year survival varies widely, ranging from 22% to 66% [4,5].

Case Presentation

This is a 65-year-old female patient with a history of right oophorectomy for ovarian cyst and haemolytic anaemia 25 years ago. She presents with an 8-month history of progressive weight loss, permanent abdominal discomfort with increased volume causing early postprandial discomfort and occasional nausea leading to vomiting. On physical examination, the patient was calm, with blood pressure of 128/78 mmHg, heart rate of 70 beats per minute, respiratory rate of 20 per minute, temperature of 36.2 °C, oxygen saturation of 85% in ambient air and generalised pallor. Abdomen with Pfannenstiel-type scar, distended, hard, not painful, with generalised dullness and no pathological peritoneal signs (Fig. 1).

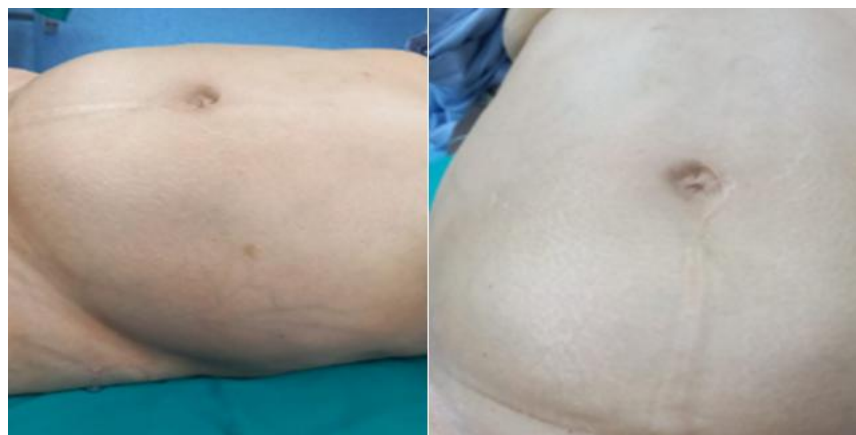


Figure 1: Abdomen before surgical treatment.

Laboratory tests show: Red blood cells 4,220,000, haemoglobin 12.7 g/dL, haematocrit 39.2%, platelets $75 \times 10^3/\mu\text{L}$, white blood cells 3,030, neutrophils 61%, C-reactive protein 249 mg/L, Glucose 89 mg, creatinine 09 mg, CA 72.4: 0.81, CA 19.9: 6.15, CA 125: 6.06, CEA: 1.66, AFP: 1.09, liver function tests, LDH, GGT and alkaline phosphatase within normal parameters.

Simple and contrast-enhanced Computed Tomography (CT) scan of the abdomen reports: large mixed mass with areas of solid fatty tissue density and an area of dense liquid density, measuring $28 \times 22 \times 13$ cm. It extends from the pelvic cavity in the right hemiabdomen to the ipsilateral hypochondrium, producing a significant mass effect on the adjacent structures, displacing them upwards and to the left; it does not enhance with intravenous contrast and appears to depend on the right ovary, suggesting a mature giant teratoma, no adenopathies and the presence of several diverticula in the left colon. She is admitted to hospital, her platelet deficiency is corrected and she is prepared for surgery with a diagnosis of intra-abdominal tumour.

Surgical Treatment

The patient was taken to the operating theatre, where she underwent a laparotomy with a xipho-pubic incision + tumour resection + incidental appendectomy, revealing a tumour measuring approximately $40 \text{ cm} \times 30 \text{ cm}$, multilobulated, with regular edges, hypervascularised, lipomatous and gelatinous consistency with hard areas, originating in the retroperitoneum from the pelvic cavity and the entire right hemiabdomen, completely crossing the midline with easily released adhesions, completely displacing the intra-abdominal structures towards the anterior abdominal wall; right kidney, ipsilateral ureter and renal vessels completely displaced towards the left hemiabdomen, very tense, with thrombosis of some peripheral venous vessels, anterior surface of the stomach, pancreas, spleen, cecal appendix, cecum and entire colon (right and left) completely adhered to the tumour weighing 18 kg; another isolated tumour dependent on the retroperitoneum, with the same characteristics, measuring approximately $10 \text{ cm} \times 7 \text{ cm}$ and weighing 2 kg, embracing the aorta and inferior vena cava. A mixed drain was placed in the abdominal cavity and blood loss during the procedure was estimated at approximately 2000 cm.

In the immediate postoperative period, she was treated with hydration, blood transfusion, plasma, broad-spectrum antibiotic therapy and analgesia. She progressed satisfactorily with adequate respiratory, renal and digestive function, with little serohematic production due to drainage and no abdominal signs suggesting complications. Follow-up laboratory tests show normalisation of haematocrit, adequate renal function and no evidence of infection. She is discharged 48 hours' post-surgery with analgesia and outpatient antibiotic therapy. The abdominal drainage is removed after 8 days with minimal serohemorrhagic

production (Fig. 2).



Figure 2: Post-surgical abdomen.

The histopathological study reports: retroperitoneal mass divided into two parts, the larger mass measuring 39x22x12 cm and weighing 18 kg and the smaller mass measuring 19x14x9 cm and weighing 2 kg, with a smooth external surface, well defined, encapsulated by a thin, poorly vascularised capsule. Histopathological diagnosis (provisional): mesenchymal neoplasm of adipose origin, consistent with well-differentiated liposarcoma (mixed sclerosing and inflammatory variant). The definitive diagnosis will be made after complementing with immunohistochemistry studies and, if necessary, s100 and ki-67 (Fig. 3).

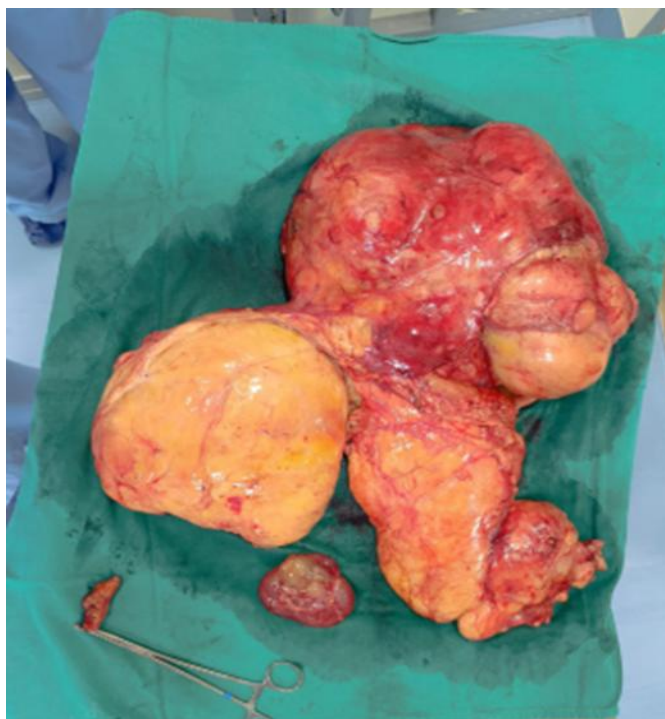


Figure 3: (Macroscopic View): Tumour masses: Cecal appendix (with forceps); Small tumour 2 kg; Large tumour 18 kg.

Discussion

Retroperitoneal tumours are the most common type of neoplasm, accounting for around 70% of retroperitoneal sarcomas. Symptoms may be delayed, mainly due to the mass causing discomfort when it reaches a large size, as was the case with the patient described here. The patient did not present a pathognomonic clinical picture for any specific pathology, so paraclinical tests were important in guiding us towards an existing abdominal pathology, bearing in mind that computed tomography is the study of choice for an initial evaluation, as it allows us to take into account the size, composition and relationship with the

surrounding structures.

Conclusion

Surgical resolution is the treatment of choice and the timely intervention performed on the patient mentioned was of great importance for a satisfactory recovery, as her post-surgical stay has significantly improved her quality of life.

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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Data Availability Statement

Not applicable.

Ethical Statement

The project did not meet the definition of human subject research under the purview of the IRB according to federal regulations and therefore, was exempt.

Informed Consent Statement

Written informed consent was obtained from the patient for the publication of this case report; no form of identification has been used in the publication of this article.

Authors' Contributions

All authors contributed equally to this paper.

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