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Original Research Paper

Surgery

# RETROPERITONEAL HIGH-GRADE SARCOMA IN A 76-YEARS-OLD MAN: Case Report.

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**ABSTRACT** Retroperitoneal soft tissue sarcomas (RPS) are rare tumours which account for approximately 12-15% of all soft tissue sarcomas with a mean incidence of 2.7 per million. RPS are frequently incidental findings in the work-up for non-related symptoms or diseases and can grow to an extremely large size in the retroperitoneum before symptoms or signs of abdominal pain, back pain, bowel obstruction or a palpable abdominal mass develop. Surgical resection is the only hope for cure and is therefore the treatment of choice for localized disease. After tumour grade, the long-term survival following RPS resection is most dependent on the completeness of surgical resection. Other important factors are patient age, tumour subtype, tumour size, multifocality and centralized multidisciplinary management in a specialist sarcoma centre.

The authors present a 76-year-old man presented with abdominal pain and a palpable mass in the right hemiabdomen.

The abdomen CT scan revealed a voluminous retroperitoneal neoformation at the level of the right side, with axial diameters of 17x12 cm and cranio-caudal extension of 15 cm. He underwent an incisional abdominal biopsy of the neoformation with an eco-guided retroperitoneal surgical access. The histological diagnosis revealed the presence of a poorly differentiated malignant neoplasm, epithelioid-like, apparently mesenchymal, so the medical team decides to subject the patient to surgery to remove the neoformation in the right hemi-abdomen.

The removed mass is then sent to the pathological anatomy and the histological diagnosis subsequently confirms the diagnosis of high-grade sarcoma with a well-differentiated liposarcoma component.

**KEYWORDS :**

## INTRODUCTION

Sarcomas are heterogeneous and clinically challenging soft tissue (muscles, connective tissues, blood and lymphatic vessels, nerves, ligaments and adipose tissue) and bone cancers.

Sarcomas represent the second most common type of solid tumours in children and adolescents and comprise an important group of secondary malignancies. In adults, soft tissue sarcomas affect about 5 people in 100,000 and represent 1 percent of all human malignancies.

Survival 5 years after diagnosis depends on the aggressiveness of the disease and on how timely the diagnosis is; considering all the stages of severity, it is around 55 percent.

Soft tissue sarcomas can be localized practically in all parts of the body, in about two out of three cases these tumours form at the level of the limbs or the superficial trunk, in the other cases they affect the head, neck, internal organs (including uterus) or internally the trunk (mainly the retroperitoneum, i.e. the posterior part of the abdominal cavity, rich in adipose and connective tissue). When develop in the abdomen, the symptoms can be much vaguer and more non-specific, such as weight loss, lack of appetite and fatigue. In some cases, abdominal swelling is noted. Rarely they can obstruct the passage of stool causing intestinal blockages, induce bleeding from the gastrointestinal tract or cause oedema of the lower limbs. Since symptoms are so general, the diagnosis of retroperitoneal and visceral sarcomas is often late.

In comparison to extremity soft tissue sarcomas, the prognosis of RPS is significantly worse. This difference is largely due to the difficulties that surgeons face in achieving wide resection margins, which relates to the location of tumour within the retroperitoneum, that frequently results in a large tumour size and complex anatomical relationships with critical vasculature and viscera at presentation.

In view of the complexity of the retroperitoneal space and the multitude of organs involved, achieving optimal resection margins can be challenging.

The most frequent sarcoma subtypes in the retroperitoneum in adults over 55 are well-differentiated liposarcoma (WDL) and dedifferentiated liposarcoma (DDL) (40%) and leiomy osarcoma (LMS) (27%).

Even though retroperitoneal sarcomas are rare tumours, they can be encountered by a wide variety of clinicians as they can be incidental findings on imaging or present with non-specific symptoms and signs. Surgical resection can offer hope of cure and patient outcomes are improved when patients are managed in high-volume specialist centres. Failure to recognize retroperitoneal sarcomas on imaging can lead to inappropriate management in inexperienced centres. Therefore, it is critical that a diagnosis of retroperitoneal sarcoma should be considered in the differential diagnosis of a retroperitoneal mass with prompt referral to a soft tissue sarcoma unit.

## CASE PRESENTATION

A 76-years-old man was sent to the hospital by his family doctor after a kidney ultrasound, performed for pain in the right hemiabdomen and showing a voluminous NDD neoformation.

Patient's past medical history includes non-insulin-dependent

diabetes mellitus in therapy with oral hypoglycaemic, right kidney lithotripsy and appendicectomy.

At the presentation in our ER department the patient was alert, oriented, collaborative, quiet and not particularly suffering. He reported a feeling of weight associated with pain to the right hemiabdomen, referred at the homolateral side. He also was apyretic, with normal bowel function and he did not present nausea and vomiting.

Abdominal physical examination revealed a palpable mass in the right hemiabdomen, minimally painful on palpation, in the absence of signs of peritonism.

In the suspicion of dealing with a sarcoma, hospitalisation was indicated for further diagnostic investigations and to set the therapeutic procedure in the most appropriate way.

So, an emergency contrast enhanced and nonenhanced abdomen CT scan was performed, which pointed out a voluminous retroperitoneal neoformation at the level of the right side, with axial diameters of 17x12 cm and cranio-caudal extension of 15 cm and with ipsilateral hydroureter onephrosis. It appeared in close relationship with the hepatis tip, with the lower pole of the right kidney which appeared displaced superiorly, with the duodenal C, which was also displaced super-medially, lapping the inferior vena cava and appearing indissociable from the right psoas muscle.

There was also calico-pyelic ectasia of the right kidney and absent opacification of the excretory path, incorporated into the proximal ureteral tract by the mass. The mid-distal ureter was recognizable and of regular calibre.

Spleen, pancreas, adrenal glands and left kidney appeared regular.

No free fluid flaps in the abdomen.

In order to characterise the mass and establish its nature, an incisional abdominal biopsy of the retroperitoneal neoformation was planned. The histological diagnosis revealed the following: "Poorly differentiated malignant neoplasm, epithelioid-like, apparently mesenchymal. Phenotypic characterization: CK7 - / CAM5.2 - / SOX10 - / CD34 - / ACT muscle-specific - / DES - / Myogenin - / VIM + ".

With MR imaging of the abdomen and pelvis performed in the basic conditions and after the administration of paramagnetic contrast enhanced intravenously (Prohance) were confirmed size of the mass (Figure 1) and the relationship of this lesion with the lower pole of the right kidney, compressed and displaced anteriorly, with the duodenal C and the inferior vena cava; the mass also had a marked infiltrative component towards the right psoas muscle, presumably its site of origin.

A lobulation of the neoplasm, on the cranial and medial side (about 2 cm), infiltrated the pillar of the diaphragm (Figure 2). Some lymph nodes with a short axis of less than one centimetre in the paraortic area, the largest of about 15 x 7 mm (Figure 3).

Small flap of free pouring into the pelvic cavity.

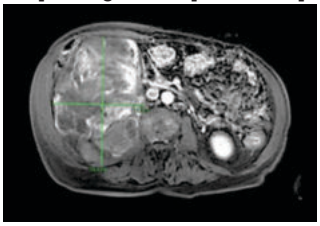

**Figure 1**

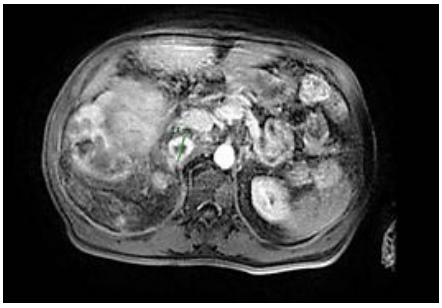

**Figure 2**

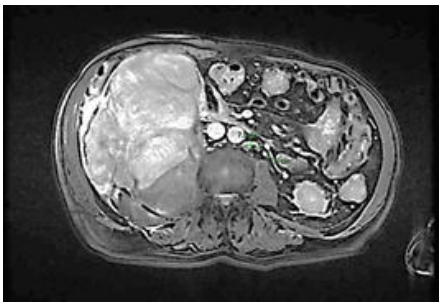

**Figure 3**

After the acquisition of all the images necessary for the study of the mass, the surgical procedure was carried out.

**The surgery was divided into two stages:**

1. An Urological time in which the placement of a right ureteral stent 6x24 took place. The ascending pyelography on the right showed hydronephrosis with ureteral dilatation.
2. A Surgical period in which a right lombotomy was performed and upon exploration of the peritoneum, a neoformation of hard consistency was found, not very mobile onto the surrounding planes and closely adhered to the deep plane. The surgical team proceed with the detachment and exeresis of the mass (Figure 4); during this manoeuvre, due to the intense inflammatory process around the mass and the consequent presence of numerous adhesions between the mass and the surrounding tissues, the right kidney was torn and it was therefore necessary to perform a right nephrectomy.

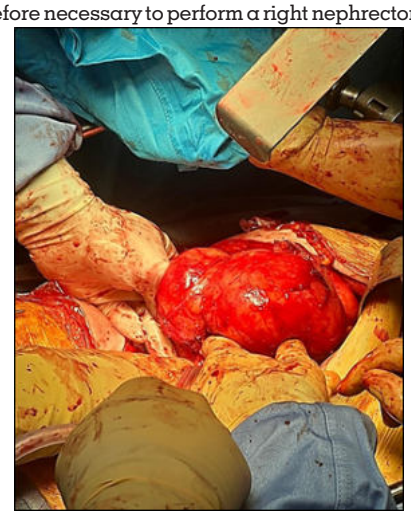

**Figure 4**

The removed mass was sent to pathological anatomy and the definitive histological examination revealed the presence of a neoformation with a plurinodular profile, fibrous design, increased in consistency and measuring 21x18x13 cm (Figure 5), proving to be a high-grade sarcoma (according to FNCLCC System), referable to dedifferentiated liposarcoma.

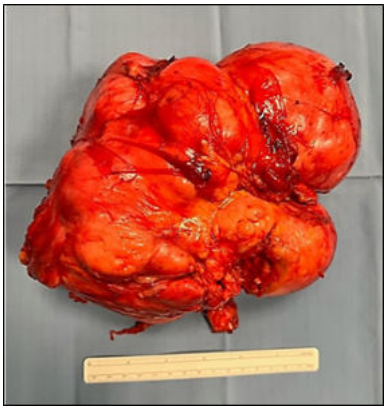

**Figure 5**

## DISCUSSION

Sarcomas differentiate based on the morphological appearance of cancer cells and their similarity to their normal tissues.

**Tumours with adipose differentiation are called:**

- Lipomas, if they are benign forms without the possibility of malignant evolution;
- Liposarcomas, on the other hand, if they are malignant neoplasms with a different spectrum of aggression and can originate wherever there is adipose tissue in the body, although the most common form generally occurs in the retroperitoneum. They are more common in people between the ages of 50 and 65.

Sarcoma is diagnosed with the help of computed tomography (CT) and magnetic resonance imaging (MRI). These imaging techniques help identify an abnormal mass of tissue.
The finding of a soft tissue swelling with a diameter of at least 3 cm in the deep tissues should lead to suspect the presence of a soft tissue sarcoma.

Surgery is the cornerstone of the treatment of forms in the localised phase and remains the mainstay of treatment for retroperitoneal sarcomas.

The tumour size, location and relationship (i.e. adjacent, encasement or invasion) to adjacent viscera, parietal wall, bone and neurovascular structures must be defined to plan for possible adjacent visceral resection. Resection of the ipsilateral kidney and adjacent hemi-colon is often required and any abnormality of the contralateral kidney or involvement of the contralateral renal vein should be reported. Common causes for non-resectability or contraindications to resectability are metastases, encasement of the celiac axis, porta hepatis and superior mesenteric vessels or extensive involvement of bone or spinal cord. In addition, lumbar vessels and collateral veins in the retroperitoneum can be a source of significant intra-operative blood loss.

The aim of surgical resection should be to achieve a macroscopic complete R0/R1 resection.

In the majority of cases, one or more organs need to be resected together with the tumour in order to achieve complete resection.

## REFERENCES


1. Grünewald, T. G. et al. (2020). Sarcoma treatment in the era of molecular medicine. EMBO Mol Med. 12(11).
2. Messiou C, et al. (2017). Primary retroperitoneal soft tissue sarcoma: Imaging appearances, pitfalls and diagnostic algorithm. Eur J Surg Oncol. 43(7):1191-1198.
3. Bonvalot S, et al. (2009). Primary retroperitoneal sarcomas: a multivariate analysis of surgical factors associated with local control. J Clin Oncol. 27:31-7.
4. Morosi C, et al. (2014). Correlation between radiological assessment and histopathological diagnosis in retroperitoneal tumors: analysis of 291 consecutive patients at a tertiary reference sarcoma center. Eur J Surg Oncol. 40:1662-70.
5. Mullinax JE, et al. (2011). Current diagnosis and management of retroperitoneal sarcoma. Cancer Control. 18:177-87.
6. Bonvalot S, et al. (2010). Aggressive surgery in retroperitoneal soft tissue sarcoma carried out at high-volume centers is safe and is associated with improved local control. Ann Surg Oncol. 17:1507-14.
7. Gronchi A, et al. (2009). Aggressive surgical policies in a retrospectively reviewed single-institution case series of retroperitoneal soft tissue sarcoma patients. J Clin Oncol. 27:24-30.
8. Gronchi A, et al. (2012). Frontline extended surgery is associated with improved survival in retroperitoneal low- to intermediate-grade soft tissue sarcomas. Ann Oncol. 23:1067-73.
9. Strauss DC, et al. (2010). Surgical management of primary retroperitoneal sarcoma. Br J Surg. 97:698-706
10. Brennan MF, et al. (2014). Lessons learned from the study of 10,000 patients with soft tissue sarcoma. Ann Surg. 260:416-21; discussion 421-2.
11. Gronchi A, et al. (2004). Retroperitoneal soft tissue sarcomas: patterns of recurrence in 167 patients treated at a single institution. Cancer. 100:2448-55.
12. Lahat G, et al. (2008). New perspectives for staging and prognosis in soft tissue sarcoma. Ann Surg Oncol. 15:2739-48.
13. Trans-Atlantic RPSWG. (2015). Management of primary retroperitoneal sarcoma (RPS) in the adult: a consensus approach from the Trans-Atlantic RPS Working Group. Ann Surg Oncol. 22:256-63.
14. Miah AB, et al. (2014). Optimal management of primary retroperitoneal sarcoma: an update. Expert Rev Anticancer Ther. 14(5):565-79.
15. Bonvalot S, et al. (2012). Technical considerations in surgery for retroperitoneal sarcomas: position paper from E-surg, a master class in sarcoma surgery, and EORTC-STBSG. Ann Surg Oncol. 19(9):2981-91.