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Case Report

Another Big Mystery Emerges. A Case of Huge Retroperitoneal Liposarcoma and a Literature Review

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Abstract

Soft tissue tumours arising in the large retroperitoneal cavity are relatively scarce. Among them, liposarcoma (RPL) constitutes the most common type encountered in clinical practice. Typically defined by a diameter of 20-30 cm or more, or by a weight of 15-20 kg or more, these tumours remain asymptomatic for a long time and only become apparent when they reach a significant size. Treatment is arduous and may require extensive surgical procedures that can excise several adjacent structures, possibly supplemented by adjuvant radiotherapy. Notwithstanding successful treatment, the recurrence rate remains very high. Herein, we report a rare case of giant RPLS in our institution and a literature review.

Keywords: Giant Liposarcoma; Retroperitoneal Liposarcoma; Rare Soft Tissue Tumour; Giant Tumour.

Introduction

Retroperitoneal tumours (RPTs) are an unusual heterogeneous group of neoplasms arising in the retroperitoneal compartment from cells in soft tissues (fat, muscle, bone, etc.), the lymphatic system, or blood vessels [1]. They are divided into benign and malignant types, and around 70-80% of them are considered malignant even if they only constitute 0.1-0.2% of all malignant tumours [2]. Nowadays, soft tissue sarcoma (STS) is recognized as the most common RPT type, and retroperitoneal liposarcoma (RPL) as the most common soft tissue sarcoma [3]. Retroperitoneal liposarcoma is a rare, typically asymptomatic tumor that develops in the retroperitoneal space and usually occurs in patients aged between 40 and 60 years; men and women are affected equally[4]. It represents 40% of all retroperitoneal STS. According to the World Health Organisation (WHO), 4 types of liposarcoma are classified as follows: well-differentiated and dedifferentiated types, which are more frequent in the retroperitoneal cavity, and pleomorphic and myxoid types, which often appear in the extremities [5]. Owing to the large space afforded by the retroperitoneal cavity, RPL can reach an extremely large size (average: 20-30cm) and weight (average: 15-20kg) [6,7]. Consequently, RPL is generally asymptomatic during the early phase of the disease and mainly causes symptoms when it reaches a significant size through pressure on neighbouring structures (ureters, kidneys, bowels, pancreas, vessels, or intraperitoneal retroperitoneal organs) and, on a lesser scale, through organ invasion, making the patient's prognosis very unfavourable [7]. This feature hinders early diagnosis and subsequent prompt and effective treatment. Herein, we present a rare case of giant RPL and a comprehensive literature review.

Case Presentation

We report the case of a 46-year-old man, without medical or surgical history, admitted to the emergency department with epigastric pain, fatigue, asthenia, weight loss (12 kg in 6 months), and increased abdominal volume. On history, the patient described feeling exhausted for several days, associated with significant loss of appetite over the past few months, and postprandial discomfort. He reports no other associated complaints. On arrival,

hemodynamic parameters were normal. Clinical examination revealed normal cardiopulmonary function, a distended abdomen, and palpation of a mass extending from the right hypochondrium to the right iliac fossa (Figure 1). The mass was hard and painless, with peristalsis present in the left lower abdomen. The rest of the clinical examination was normal. Bearing in mind the patient's condition, further investigations were carried out. Blood biology showed mainly a significant inflammatory syndrome associated with elevated LDH levels. Abdominal CT imaging revealed a first retroperitoneal mass measuring 20 cm x 16 cm with hypo dense content in the centre suggesting necrosis, and a second mass measuring 20 cm x 25 cm with cystic lesions. These masses pushed the right kidney to the left anterior and the liver to the left, inducing compression of the inferior vena cava and infiltration of the right psoas muscle. An abdominal MRI confirmed the presence of a voluminous tumor with 2 cysts and fleshy components, pushing back the abdominal structures (kidneys, intestines, pancreas, and liver) (Figure 2). There was no vascular infiltration or thrombosis. The vascular pedicle of the tumour mass was located inferiorly alongside the sacral promontory. The diagnosis of retroperitoneal liposarcoma was retained and surgery was rapidly performed. Surgical resection yielded a total of 4 tumour lesions of different sizes and consistency with a total weight of 16 kg (Figure 3). Anatomopathological examination confirmed the RPL's diagnosis and revealed 3 different associated histological subtypes: well-differentiated type for 2 small lesions, myxoid (1 lesion), and undifferentiated type (1 lesion). According to the grading system of the National Federation of Centres de Lutte Contre le Cancer (FNCLCC), the tumour was classified as grade 2 (score 5: dedifferentiation 3, necrosis 1 mitosis 1). Chromosomal abnormalities in the region of chromosome 12 q13-15 were positive. The patient did not receive adjuvant radiotherapy or chemotherapy. After five months, an abdominal CT scan revealed a recurrence of the tumour mass. The patient underwent re-operation with resection of the tumour mass, including the proximal insertion of the psoas muscle, the posterior insertion of the right diaphragmatic dome, and a right hemicolectomy. Six months after the re-operation, a new abdominal CT scan was carried out, and no signs of recurrence were observed. At 12 months, the patient was completely disease-free.

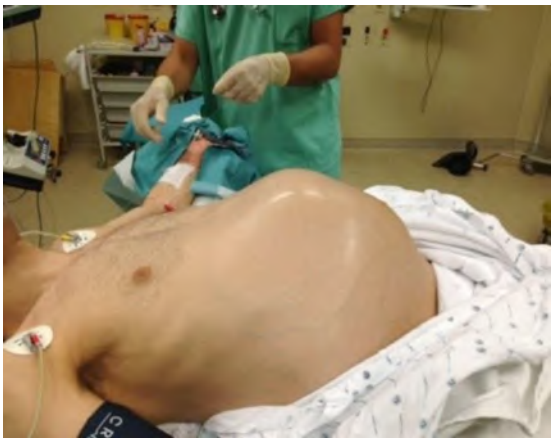

Figure 1: Clinical examination of the patient reveals a distended abdomen with palpation of a mass extending from the right hypochondrium to the right iliac fossa.

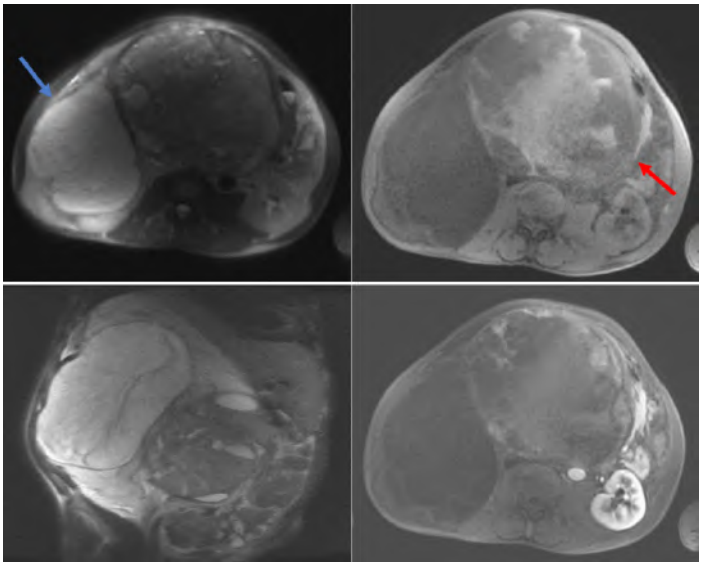

Figure 2: Abdominal MRI demonstrating the presence of a voluminous tumour with fleshy (red arrow), cystic components (2, blue arrow) pushing back the abdominal structures (kidneys, intestines, pancreas and liver).

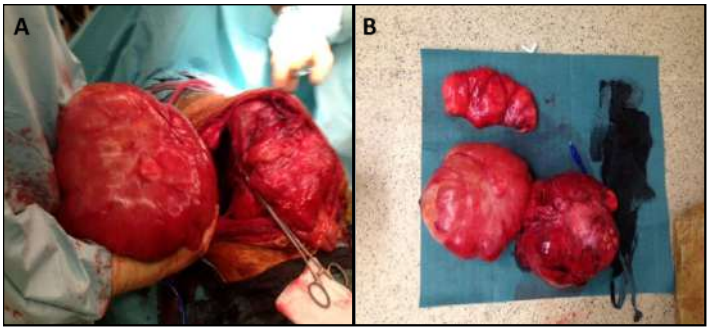

Figure 3: Intraoperative images showing the large mixed-component mass and the removal of 3 other associated masses (joined together).

Discussion

Retroperitoneal liposarcoma (RPL) is a scarce mesenchymal neoplasm, accounting for approximately 40-45% of adult retroperitoneal soft tissue sarcomas (the most prevalent), and 0.1-0.2% of all malignant tumours [8]. It typically affects individuals aged between 40 and 60 years, with a relatively uniform distribution between genders [9]. While known, the underlying risk factors for this condition are not fully understood, and several studies have suggested a probable link with certain chromosomal abnormalities as in our patient [10]. According to the literature, the American Cancer Society (ASC) outlines several risk factors that may increase the developing risk of RPL as follows; previous radiation (especially for other malignancies), a family history of cancer, lymphatic system trauma, and exposure to hazardous chemicals [11].

Other than being a rare form of cancer, the manifestations of RPL are usually unremarkable and non-specific when present, and are often identified at an advanced stage, characterized by a large abdominal mass [12]. Consequently, many patients exhibit large tumours and, based on histopathological features, often have a poor prognosis. Typical complaints include a palpable tumour, pain or fullness in the abdomen or flank and early satiety as in our patient, pain or swelling in the lower extremities, or symptoms of urinary or bowel obstruction [13].

In clinical routine, computed tomography (CT) is largely employed for the diagnosis and preoperative evaluation of retroperitoneal liposarcoma [14]. Nevertheless, magnetic resonance imaging (MRI) is now considered the gold standard as it affords higher soft tissue resolution, enabling a more accurate diagnosis of retroperitoneal tumours [15]. MRI also provides a clear view of the tumour's blood vessels, enabling tumour characteristics to be identified and tumour invasion to be assessed.

The World Health Organisation (WHO) described 4 types of liposarcoma classified as follows: well-differentiated and dedifferentiated types, which are more frequent in the retroperitoneal cavity as in our patient, and pleomorphic and myxoid types, which often appear in the extremities [2,5]. Our case is quite unique in that it presents with 3 different subtypes within the same mass, highlighting its heterogeneity and complicating its management. Prognostic factors affecting patients with retroperitoneal liposarcoma include mitosis rate, tumour size, negative resection margins, and histopathological type [16]. The larger the tumour, the higher the rate of distant metastases. According to the latest data in the literature, tumours larger than 20 cm can lead to distant metastasis rates of 50-60% at five years [16].

Currently, surgical resection remains the cornerstone modality for retroperitoneal tumours [17]. Nevertheless, most patients still require supplementary treatment modalities following successful resection of the tumour, due to the higher recurrence rate of RPL compared with LPS in other locations [17]. The efficacy of radiotherapy and chemotherapy in RPL remains controversial. Debate has been sparked over the role of radiotherapy, particularly by the STRASS and STREXIT trials, which demonstrated its efficacy in specific scenarios (certain types of liposarcoma) [18,19]. Some retrospective studies suggest that preoperative radiotherapy improves the probability of obtaining R0-R1 resections [20]. Other studies go further, showing an increased survival rate in patients with liposarcomas larger than 10cm, but prudence is required for tumours below this size, and long-term benefits have not been thoroughly evaluated [21]. Anthracycline-based chemotherapy regimens (doxorubicin) and Ifosfamide are currently considered the first-line treatment for advanced or metastatic LPS [22]. Nevertheless, an extensive phase III randomized controlled trial carried out by the European Organisation for Research and Treatment of Cancer (EORTC) concluded that this combined treatment did not improve overall survival (OS) or recurrence rates [23]. Targeted therapies have been investigated for RPL but their effectiveness and benefits have yet to be established [24,25].

Conclusion

Retroperitoneal liposarcoma is a multifaceted and rare tumour with a high potential for loco regional invasion and relapse. It may be very large and remain asymptomatic for a long time. Its unique characteristics, particularly its diagnosis and the large size of the tumour, afford valuable indications for the overall management. Computed tomography (CT) and magnetic resonance imaging (MRI) are useful complementary examination methods. Surgery is the preferred therapeutic approach at present. Although the efficacy of radiotherapy and chemotherapy in the treatment of RPLPS remains to be ascertained, targeted therapy holds promise

as a treatment strategy and is a new area to investigate in the future. This case highlights the complex management of a rare retroperitoneal tumour, showcasing the challenges in achieving radical resection. Our understanding of liposarcoma is fundamental and involves raising awareness among healthcare professionals to ensure early detection and intervention.

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