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Retroperitoneal Leiomyosarcoma Presenting as a Large Abdominal Mass with Synchronous Pulmonary Metastases: A Case Report

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Abstract **Case Report**

Leiomyosarcoma (LMS) is a rare malignant mesenchymal neoplasm arising from smooth muscle cells and accounts for approximately 10–20% of all soft tissue sarcomas (WHO Classification of Tumours Editorial Board, 2020). Retroperitoneal leiomyosarcoma is an uncommon subtype that frequently presents at an advanced stage because of its deep anatomical location and nonspecific clinical manifestations. We report the case of a 57-year-old man who presented with progressive unintentional weight loss, excessive sweating, and a palpable left-sided abdominal mass. Contrast-enhanced computed tomography demonstrated a 12.0 × 15.0 × 16.0 cm well-defined, lobulated, heterogeneously enhancing retroperitoneal soft tissue mass with areas of central necrosis causing mild left hydronephrosis due to mass effect. Staging investigations identified multiple bilateral pulmonary nodules suspicious for metastatic disease. The patient underwent exploratory laparotomy with complete surgical excision of the tumour. Histopathological examination confirmed FNCLCC Grade 2 retroperitoneal leiomyosarcoma (pT3) with microscopically negative surgical margins (R0). Immunohistochemistry demonstrated positivity for caldesmon, desmin, smooth muscle actin (SMA), and CD117, while DOG1, CD34, S100, STAT6, MDM2, CAM5.2, and ERG were negative, supporting the diagnosis of leiomyosarcoma. Following multidisciplinary sarcoma team discussion, the patient commenced systemic chemotherapy for metastatic disease. This case highlights the diagnostic challenges associated with retroperitoneal leiomyosarcoma and underscores the importance of early diagnosis, complete surgical resection, accurate histopathological evaluation, and multidisciplinary management in optimising patient outcomes.

Keywords: Leiomyosarcoma; Retroperitoneal leiomyosarcoma; Retroperitoneal sarcoma; Soft tissue sarcoma; Pulmonary metastases.

INTRODUCTION

Leiomyosarcoma (LMS) is a malignant mesenchymal neoplasm arising from smooth muscle cells and accounts for approximately 10–20% of all soft tissue sarcomas (WHO Classification of Tumours Editorial Board, 2020). It most commonly arises in the uterus, retroperitoneum, and deep soft tissues of the extremities. Retroperitoneal leiomyosarcoma represents one of the principal histological subtypes of primary retroperitoneal sarcoma and is characterised by aggressive biological behaviour, with a high propensity for local recurrence and haematogenous metastasis, particularly to the lungs and liver (Casali *et al.,* 2021; Gronchi *et al.,* 2021).

Because of the large potential space within the retroperitoneum, these tumours frequently remain asymptomatic until they reach a considerable size. As a result, patients often present with vague constitutional symptoms such as abdominal discomfort, early satiety, weight loss, or a palpable abdominal mass rather than symptoms directly attributable to the tumour itself. Compression of adjacent organs may result in secondary manifestations including hydronephrosis, bowel displacement, or vascular compression, often reflecting advanced disease at presentation (Messiou *et al.,* 2017).

Contrast-enhanced computed tomography is the imaging modality of choice for the initial evaluation of retroperitoneal masses and provides valuable information regarding tumour size, anatomical relationships, and metastatic disease. Magnetic resonance imaging offers complementary assessment of local tumour extent, particularly when involvement of adjacent structures is suspected. Nevertheless, imaging findings alone are insufficient for definitive diagnosis because several retroperitoneal neoplasms, including

gastrointestinal stromal tumours, liposarcomas, solitary fibrous tumours, and malignant peripheral nerve sheath tumours, may demonstrate overlapping radiological characteristics. Histopathological examination supported by immunohistochemistry therefore remains the gold standard for diagnosis (WHO Classification of Tumours Editorial Board, 2020).

Complete surgical excision with microscopically negative margins remains the cornerstone of treatment and is the most important determinant of long-term survival. Despite advances in surgical management, retroperitoneal leiomyosarcoma continues to carry a significant risk of both local recurrence and distant metastasis, highlighting the importance of multidisciplinary management involving specialist sarcoma surgeons, radiologists, pathologists, and medical oncologists (Gronchi *et al.,* 2021; Casali *et al.,* 2021). We present a case of retroperitoneal leiomyosarcoma presenting as a large abdominal mass with synchronous pulmonary metastases to illustrate the diagnostic challenges, clinicopathological features, and multidisciplinary management of this uncommon malignancy.

Case Presentation

A 57-year-old man presented with a two-month history of progressive unintentional weight loss of approximately 20 kg, accompanied by excessive sweating and a gradually enlarging left-sided abdominal swelling. His medical history was significant for type 2 diabetes mellitus, hypertension, dyslipidaemia, gastro-oesophageal reflux disease, and gout. His previous surgical history included mesh repair of a right inguinal hernia ten years earlier.

On physical examination, the patient appeared clinically stable. Abdominal examination revealed a firm, non-tender, palpable mass measuring approximately 10 cm over the left side of the abdomen. The overlying skin was normal, with no evidence of erythema or ulceration. No palpable lymphadenopathy was identified.

In view of the patient's constitutional symptoms and palpable abdominal mass, urgent abdominal imaging was arranged. Contrast-enhanced computed tomography (CT) of the abdomen and pelvis demonstrated a large, well-defined, lobulated, heterogeneously enhancing soft tissue mass measuring 12.0 × 15.0 × 16.0 cm, containing areas of central necrosis with surrounding fat stranding. The lesion was located within the left retroperitoneum, abutting the left psoas muscle without a distinct intervening fat plane and producing mass effect on adjacent structures, resulting in mild left-sided hydronephrosis. The radiological differential diagnosis included gastrointestinal stromal tumour (GIST), a tumour arising in relation to the gonadal vasculature, and other primary retroperitoneal neoplasms. Upper gastrointestinal endoscopy and colonoscopy demonstrated colonic polyps and sigmoid diverticulosis but no evidence of a primary gastrointestinal malignancy.

Staging CT of the thorax demonstrated multiple bilateral pulmonary nodules suspicious for metastatic disease, the largest measuring 7.2 mm within the anterior segment of the right upper lobe. Additional nodules were identified in the right middle lobe, right lower lobe, left upper lobe, and left lower lobe. Whole-body nuclear medicine imaging also suggested multiple bilateral pulmonary metastases without evidence of additional distant metastatic disease. Magnetic resonance imaging confirmed a left retroperitoneal neoplasm without direct invasion of adjacent retroperitoneal structures or evidence of intra-abdominal metastatic spread.

Following discussion at the specialist sarcoma multidisciplinary team meeting, the patient underwent exploratory laparotomy with complete wide excision of the retroperitoneal tumour.

Gross pathological examination demonstrated an unoriented retroperitoneal soft tissue specimen measuring 19 × 13 × 11 cm, containing a well-circumscribed tan-coloured tumour measuring 15 × 12 × 10 cm with focal fibrosis and less than 10% tumour necrosis. Histopathological examination demonstrated a highly cellular spindle cell neoplasm composed of intersecting fascicles of elongated cells with eosinophilic cytoplasm and characteristic cigar-shaped nuclei showing moderate pleomorphism and hyperchromasia. Mitotic activity was readily identified, with approximately 13 mitoses per 10 high-power fields. Scattered multinucleated giant cells and focal lymphocytic inflammatory infiltrates were also present.

Immunohistochemical analysis demonstrated diffuse positivity for caldesmon, desmin, smooth muscle actin (SMA), and CD117, while DOG1, CD34, S100, STAT6, MDM2, CAM5.2, and ERG were negative. The overall morphological and immunophenotypic features were consistent with leiomyosarcoma.

The final histopathological diagnosis was FNCLCC Grade 2 retroperitoneal leiomyosarcoma (pT3) with complete (R0) surgical excision. The tumour measured 15 cm in greatest dimension, demonstrated less than 10% tumour necrosis, and showed no evidence of lymphovascular invasion. All surgical margins were free of tumour, with the closest margin measuring 0.2 cm.

Following postoperative multidisciplinary team review, the patient commenced systemic chemotherapy because of synchronous pulmonary metastatic disease. Baseline fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) performed following surgery demonstrated complete resection of the primary tumour with resolution of the previously observed left hydronephrosis. Mild uptake

within the anterior abdominal wall was consistent with postoperative change. Although no evidence of local recurrence or additional metastatic lesions was identified, interval enlargement and increased metabolic activity of the bilateral pulmonary nodules were observed, consistent with metastatic progression.

DISCUSSION

Retroperitoneal leiomyosarcoma is an uncommon malignant mesenchymal neoplasm arising from smooth muscle cells and represents one of the principal histological subtypes of primary retroperitoneal sarcoma (WHO Classification of Tumours Editorial Board, 2020; Gronchi *et al.,* 2021). Owing to the large potential space of the retroperitoneum, these tumours frequently attain considerable size before becoming clinically apparent, resulting in delayed diagnosis and advanced disease at presentation. Consequently, constitutional symptoms such as weight loss, abdominal discomfort, early satiety, or a palpable abdominal mass are often the initial manifestations rather than symptoms related to direct organ invasion (Casali *et al.,* 2021). This pattern was clearly demonstrated in our patient, who presented with significant unintentional weight loss and a large palpable abdominal mass that had already reached 15 cm at the time of surgical excision.

Cross-sectional imaging plays a pivotal role in the evaluation of retroperitoneal masses by defining tumour size, anatomical relationships, and the presence of metastatic disease. In the present case, contrast-enhanced computed tomography demonstrated a large heterogeneous retroperitoneal mass with central necrosis and mass effect on adjacent structures, while magnetic resonance imaging confirmed its retroperitoneal origin without direct invasion of surrounding organs. Nevertheless, radiological appearances remain non-specific, and several retroperitoneal neoplasms, including gastrointestinal stromal tumour (GIST), liposarcoma, solitary fibrous tumour, and malignant peripheral nerve sheath tumour, may demonstrate overlapping imaging characteristics (Messiou *et al.,* 2017). Consequently, definitive diagnosis relies on histopathological examination supported by immunohistochemistry.

Histologically, leiomyosarcoma is characterised by intersecting fascicles of spindle cells with eosinophilic cytoplasm and elongated blunt-ended ("cigar-shaped") nuclei, accompanied by variable degrees of nuclear pleomorphism, mitotic activity, and tumour necrosis. These features contribute to tumour grading using the French Federation of Cancer Centers Sarcoma Group (FNCLCC) grading system, which remains an important prognostic indicator (WHO Classification of Tumours Editorial Board, 2020). In the present case, the tumour was classified as FNCLCC Grade 2 with a mitotic rate of 13 mitoses per 10 high-power fields and less than 10% tumour necrosis, consistent with an intermediate-grade sarcoma.

Immunohistochemistry plays an essential role in distinguishing leiomyosarcoma from other spindle cell tumours of the retroperitoneum. Our tumour demonstrated diffuse positivity for caldesmon, desmin, smooth muscle actin (SMA), and CD117, while DOG1, CD34, S100, STAT6, MDM2, CAM5.2, and ERG were negative. Although CD117 positivity initially raised the possibility of a gastrointestinal stromal tumour, the absence of DOG1 and CD34 expression together with the characteristic histomorphological features supported the diagnosis of leiomyosarcoma. This case therefore highlights the importance of interpreting immunohistochemical markers within the context of the overall pathological findings rather than relying on a single marker.

Comparison with previously published cases further emphasises the characteristic clinical behaviour of retroperitoneal leiomyosarcoma. Hermi *et al.,* (2023) described a giant retroperitoneal leiomyosarcoma presenting as a progressively enlarging abdominal mass, illustrating the tendency of these tumours to remain clinically silent until they reach considerable size. Similarly, Vallejo *et al.,* (2014) reported a 15 cm retroperitoneal leiomyosarcoma in a 67-year-old man, closely resembling the tumour size observed in our patient. More recently, Boujguenna *et al.,* (2025) reported a retroperitoneal leiomyosarcoma in which imaging findings alone were insufficient to establish the diagnosis, with histopathological examination and immunohistochemistry ultimately confirming the tumour subtype. These reports are consistent with our findings and reinforce the diagnostic challenges associated with retroperitoneal leiomyosarcoma. However, unlike several previously published cases that presented with localised disease, our patient had synchronous bilateral pulmonary nodules suspicious for metastatic disease at initial presentation, highlighting the aggressive biological behaviour and early haematogenous dissemination associated with this malignancy.

Complete surgical excision with microscopically negative margins remains the cornerstone of treatment and represents the single most important prognostic factor for patients with retroperitoneal leiomyosarcoma (Gronchi *et al.,* 2021; Casali *et al.,* 2021). Achieving an R0 resection has consistently been associated with improved local disease control and overall survival. Despite the considerable size of the tumour and its close relationship to adjacent retroperitoneal structures, complete excision with negative margins was successfully achieved in the present case, demonstrating the importance of meticulous surgical planning within a specialist multidisciplinary sarcoma service.

Pulmonary metastases are the most common site of distant spread because leiomyosarcoma

predominantly disseminates through the haematogenous route. Multiple bilateral pulmonary nodules were identified during initial staging and subsequently demonstrated interval enlargement and increased metabolic activity on postoperative FDG PET/CT, consistent with metastatic disease. Nevertheless, no evidence of local recurrence or additional metastatic lesions was identified following surgical resection. Current international guidelines recommend that patients with metastatic soft tissue sarcoma should be managed through a multidisciplinary team, with systemic chemotherapy considered according to tumour biology, disease burden, patient performance status, and individual treatment goals (Casali *et al.,* 2021). Accordingly, our patient commenced systemic chemotherapy following multidisciplinary discussion.

The prognosis of retroperitoneal leiomyosarcoma is influenced by several clinicopathological factors, including tumour size, histological grade, mitotic activity, surgical margin status, and the presence of metastatic disease at diagnosis. Although complete surgical resection offers the greatest opportunity for prolonged survival, both local recurrence and distant metastasis remain common, necessitating long-term surveillance with cross-sectional imaging. Early diagnosis remains challenging because of the non-specific clinical presentation; however, prompt recognition, accurate histopathological diagnosis, radical surgical resection, and coordinated multidisciplinary management remain fundamental to optimising patient outcomes.

The present case contributes to the limited literature describing retroperitoneal leiomyosarcoma presenting as a large abdominal mass with synchronous pulmonary metastases. It reinforces the importance of considering retroperitoneal sarcoma in the differential diagnosis of large intra-abdominal masses and highlights the indispensable role of histopathological examination and immunohistochemistry in establishing the definitive diagnosis. Furthermore, it illustrates that complete surgical excision, even in patients with advanced disease, remains a key component of management within a specialist sarcoma multidisciplinary team.

Despite advances in surgical techniques and systemic therapy, five-year overall survival remains limited, particularly in patients presenting with metastatic disease, highlighting the importance of long-term surveillance and multidisciplinary follow-up.

Conclusion

Retroperitoneal leiomyosarcoma is a rare and aggressive soft tissue sarcoma that frequently presents late because of its deep anatomical location and non-specific clinical manifestations. This case demonstrates the diagnostic challenges associated with large retroperitoneal masses and highlights the importance of maintaining a broad differential diagnosis in patients presenting with constitutional symptoms and an abdominal mass. Although imaging is essential for tumour localisation and staging, definitive diagnosis relies on histopathological examination with immunohistochemical analysis.

Complete surgical excision with negative margins remains the cornerstone of treatment and offers the best opportunity for local disease control. Nevertheless, the presence of synchronous pulmonary metastases in our patient illustrates the aggressive biological behaviour of retroperitoneal leiomyosarcoma and the importance of multidisciplinary management involving specialist sarcoma surgeons, radiologists, pathologists, and medical oncologists. Long-term surveillance remains essential because of the substantial risk of local recurrence and distant metastatic progression.

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Conflict of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

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