

# Laparoscopic resection of retroperitoneal metastasis originating from sternal leiomyosarcoma – a case report

## Laparoskopická resekce retroperitoneální metastázy pocházející ze sternálního leiomyosarkomu – kazuistika

Ioannidis O.<sup>1</sup>, Kotidis E.<sup>1</sup>, Anestiadou E.<sup>1</sup>, Matzarakis I.<sup>2</sup>, Skliris J. P.<sup>2</sup>, Symeonidis S.<sup>1</sup>, Bitsianis S.<sup>1</sup>, Kerasidou O.<sup>1</sup>, Kaprianou M.<sup>1</sup>, Chatzianestiadou C.<sup>1</sup>, Salta-Poupnara E.<sup>1</sup>, Tserkezidis F.<sup>1</sup>, Angelopoulos K.<sup>1</sup>, Angelopoulos S.<sup>1</sup>

<sup>1</sup> Fourth Department of Surgery, Medical School, Faculty of Health Sciences, Aristotle University of Thessaloniki, General Hospital "George Papanikolaou", Thessaloniki, Greece

<sup>2</sup> Department of Pathology, General Hospital "George Papanikolaou", Thessaloniki, Greece

### Summary

**Background:** Leiomyosarcomas are malignancies of smooth muscle differentiation that can metastasize early through hematogenous spread. Leiomyosarcoma of the sternum is extremely rare, with an incidence of < 0.7% among malignant bone tumors. **Case:** We report the case of a 66-year-old male with a retroperitoneal metastasis located in the right pararenal space, originating from a sternal leiomyosarcoma. Three years earlier, the patient had undergone partial excision of the sternum and costal cartilages, followed by thoracic wall osteosynthesis, for the primary tumor. Follow-up imaging with CT and PET-CT revealed a mass in the right perirenal space, situated between the upper pole of the right kidney and the right adrenal gland. In the absence of other metastatic lesions and following multidisciplinary oncology team discussion, the patient underwent laparoscopic en bloc metastasectomy and right adrenalectomy. **Results:** Histopathological examination confirmed the metastatic nature of the tumor with negative resection margins. Postoperatively, the patient experienced fever due to atelectasis, which resolved with intravenous antibiotics. He was discharged on the 6<sup>th</sup> postoperative day and remains free of recurrence at a two-year follow-up. **Conclusion:** Minimally invasive surgical techniques allow for oncological resection of retroperitoneal metastatic lesions with low perioperative morbidity and mortality. Resection of isolated lymph node metastases has been shown to improve overall survival and disease control.

### Key words

retroperitoneal metastasis – lymph node metastasis – leiomyosarcoma – sternum – minimally invasive metastasectomy – adrenalectomy

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Elissavet Anestiadou, MD, MSc, MSc, PhD

Fourth Department of Surgery  
Medical School, Faculty of Health Sciences

Aristotle University of Thessaloniki  
General Hospital "George Papanikolaou"

57010 Thessaloniki, Greece  
e-mail: elissavetxatz@gmail.com

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## Souhrn

**Východiska:** Leiomyosarkomy jsou malignity hladkého svalstva, které mohou časně metastazovat hematogenním šířením. Leiomyosarkom sternu je extrémně vzácný, mezi maligními nádory kostí má incidenci  $< 0,7\%$ . **Případ:** Prezентujeme případ 66letého muže s retroperitoneální metastázou lokalizovanou v pravém pararenálním prostoru, pocházející z leiomyosarkomu sternu. Pacient před 3 lety podstoupil parciální resekci sternu a žebních chrupavek následnou osteosyntézou hrudní stěny kvůli primárnímu nádoru. V rámci dalšího sledování CT a PET-CT odhalily masu v pravém perirenálním prostoru, mezi horním pólem pravé ledviny a pravou nadledvinou. Při absenci dalších metastatických lézí a po rozhodnutí multidisciplinárního onkologického týmu byla provedena laparoskopická en bloc metastazektomie a pravostranná adrenalectomie. **Výsledky:** Histopatologické vyšetření potvrdilo metastatickou povahu nádoru s negativními resekcními okraji. Pooperačně se u pacienta objevila horečka způsobená atelektázou, která byla zvládnuta intravenózními antibiotiky. Z nemocnice byl propuštěn šestý pooperační den a po dvouletém sledování je bez recidivy. **Závěr:** Minimálně invazivní chirurgické techniky umožňují onkologickou resekci retroperitoneálních metastatických lézí s nízkou perioperační morbiditou a mortalitou. Bylo prokázáno, že resekce izolovaných metastáz do lymfatických uzlin zlepšuje celkové přežití a kontrolu onemocnění.

## Klíčová slova

retroperitoneální metastáza – metastáza do lymfatických uzlin – leiomyosarkom – sternum – minimálně invazivní metastazektomie – adrenalectomie

## Introduction

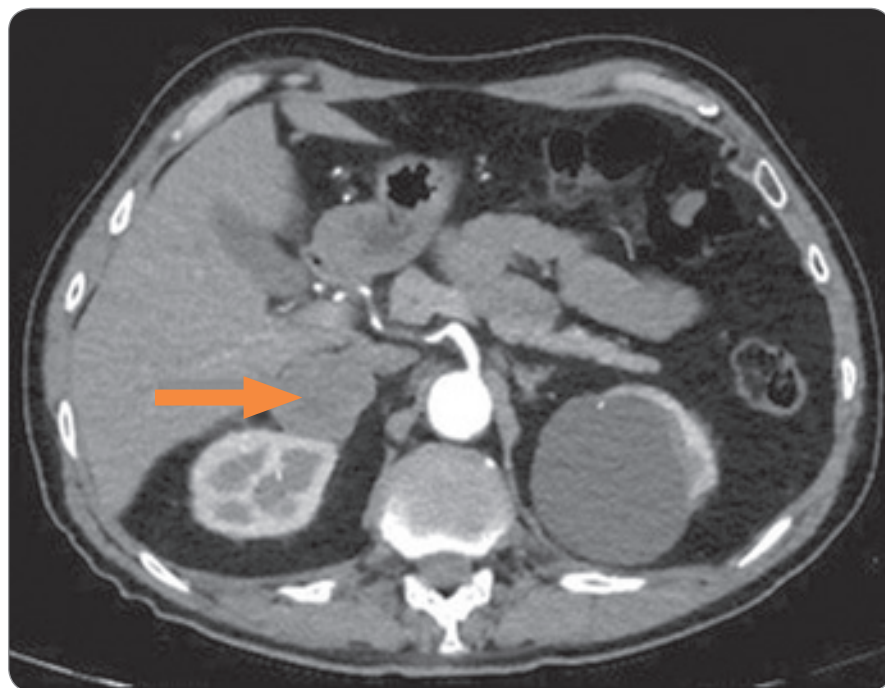
Primary leiomyosarcoma (LMS) of the bone was first described in 1965 by Evans and Sanerkin and is a rare and aggressive primary osseous sarcoma, representing less than  $< 0.7\%$  of all primary malignant bone tumors [1,2]. It is a high-grade destructive osseous tumor with poor prognosis, depending on the stage at diagnosis, and may arise *de novo* or after previous radiation [3,4]. Primary leiomyosarcoma of the sternum is an extremely uncommon clinical entity,

with few references reported in the literature worldwide [5,6]. Surgical resection is highly due to a poor prognosis of 35% overall survival (OS) rate [6]. The following case report is about a 66-year old patient, presented with retroperitoneal metastasis secondary to primary leiomyosarcoma of the sternum excised two years before, and is described in accordance to the Case Report (CARE) guidelines [7]. Both the patient and his relatives were thoroughly informed and a written informed consent was ob-

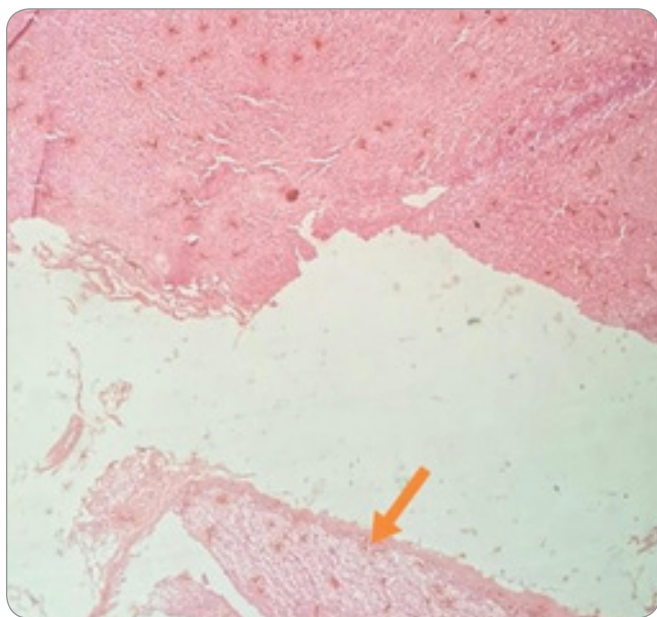
tained for publication of the present case report and the accompanying images. Copies of the written consent are available for review by the Editor-in-Chief of this journal. The present study highlights the importance of awareness of primary bone leiomyosarcoma metastatic potential, as well as the role of minimally invasive surgery in achieving low morbidity and mortality rates after resection of isolated metastatic lesions.

## Case report

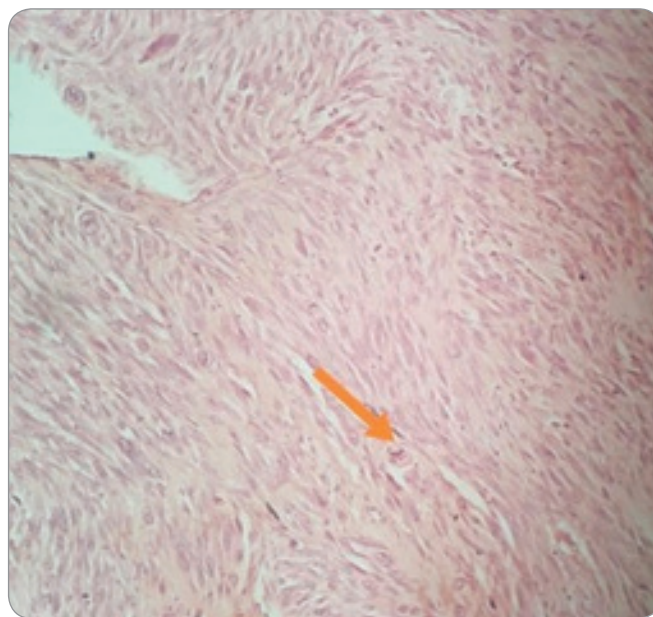
We report a case of a 66-year-old male, diagnosed with primary leiomyosarcoma of the sternum three years before, without history of irradiation. Past medical history was free of comorbidities. Partial sternal resection along with resection of adjacent chest wall structures with negative fatty margins and reconstruction of thoracic wall with methylmethacrylate prosthesis was performed. No adjuvant therapy followed, after making shared decisions with the patient due to confirmation of negative margins in the final pathology report. The postoperative course was uneventful and without any symptoms, until a nodal metastatic lesion in the retroperitoneum was encountered at a three-year follow-up. Particularly, follow-up computed tomography revealed a solitary metastatic lesion in the right pararenal space, between the upper pole of the right kidney and the right adrenal gland, measuring  $4.1 \times 3.3 \times 3.5$  cm (Fig. 1). The



**Fig. 1. Enhanced computed tomography revealing a metastatic lesion (arrow) in close proximity to the right adrenal gland and kidney.**



**Fig. 2.** Tumor spindle cells arranged in fascicles on the upper part of the image. Adjacent normal adrenal gland (arrow). Hematoxylin and eosin staining, 4x.



**Fig. 3.** Higher power of neoplastic cells, exhibiting marked polymorphism and mitotic activity (arrow). Hematoxylin and eosin staining, 20x.

lesion was in close proximity to the right renal vein and seemed to be infiltrating the right adrenal gland. PET-CT also confirmed the aforementioned finding, revealing a hypermetabolic lesion in the right pararenal space, with no signs of recurrence in the chest wall. The rest of laboratory data, including serum levels of alkaline phosphatase, calcium, phosphate, and tumor markers, were normal. Core needle biopsy was also performed and the histopathological examination was indicative of a metastatic mesenchymal malignant neoplasm compatible with leiomyosarcoma. Based on long disease-free interval, absence of other metastatic lesions and good physical and biological status of the patient, the oncology group approved the surgical management. The patient underwent en block laparoscopic surgical excision of the retroperitoneal metastatic lesion, combined with right adrenalectomy due to the assumed infiltration of part of the adrenal gland. Under general anesthesia, the patient was placed in a full lateral left decubitus position. Transperitoneal approach for was chosen and four ports were used. Severe adhesions were noted around the right adrenal central vein. This area was dissected by sharp dissection and the metastatic nodal mass was

completely resected macroscopically en block with the right adrenal gland. The operative time was 135 minutes, and blood loss was minimal. Immediately after surgery, the patient returned to the ward and remained stable. On the 2<sup>nd</sup> postoperative day, he developed fever up to 38.3 °C, which was attributed to atelectasis, based on chest X-rays and after pulmonary consultation, intravenous antibiotic coverage with piperacillin/tazobactam and intensive respiratory physiotherapy began. The patient was discharged on 7<sup>th</sup> postoperative day.

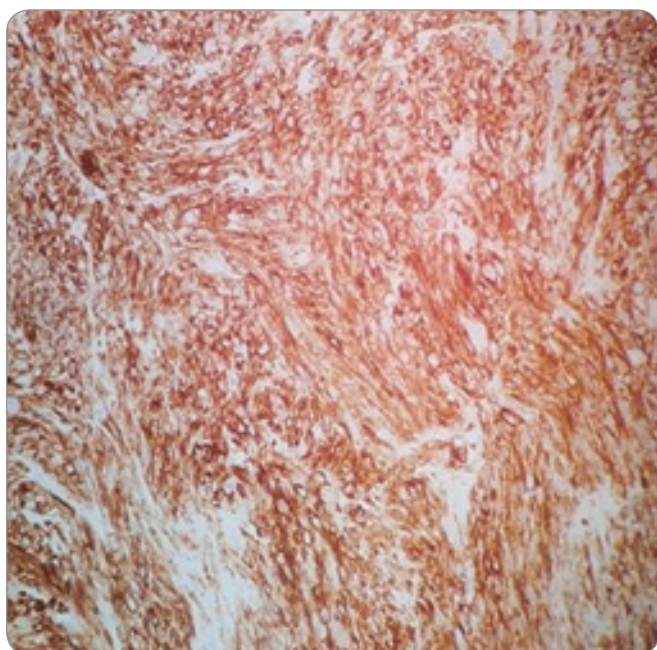
The mass was sent for histological examination. The mass was solid and whitish and measured 5.7 × 4.5 × 4 cm. The histology report confirmed the preoperative diagnosis of retroperitoneal metastasis originating from the primary sternal leiomyosarcoma, as well as the negative resection margins (Fig. 2), describing a mesenchymal malignant tumor with neoplastic cells exhibiting marked pleomorphism and increased mitotic activity (Fig. 3). During immunohistochemical evaluation, tumor cells stained positive for caldesmon and desmin (Fig. 4, 5). No adjuvant therapy was administered. At a two-year follow-up, the patient remains free of symptoms and recurrence, according to the findings of

physical examination, whole body CT, and PET-CT.

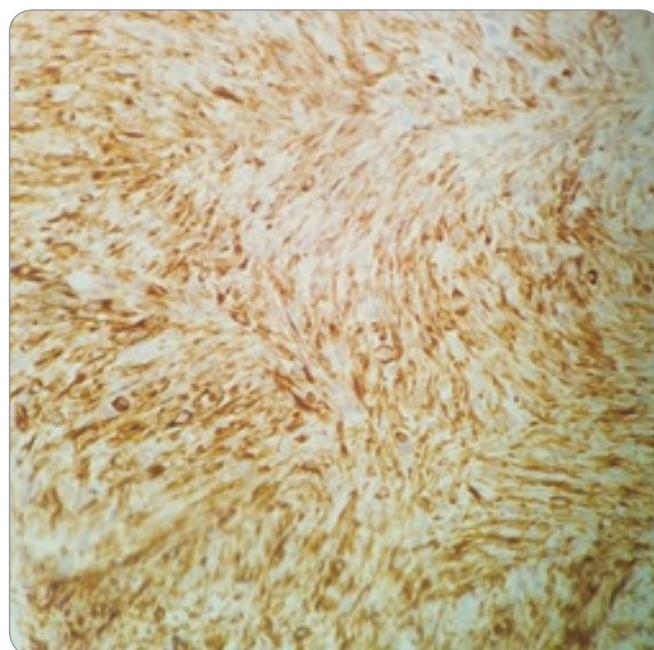
### Discussion

Primary LMS of the bones represents an uncommon malignant intraosseous neoplasm, characterized by spindle cells with smooth muscle differentiation, without matrix production [8]. It represents less than 0.7% of all malignant bone tumors and, thus, knowledge of biology and clinical behavior is limited [9]. Main locations affected include the long bones, particularly distal femur and proximal tibia, in 70% of cases, followed by the craniofacial bones in about 20% of the cases [4]. Sternum primary leiomyosarcoma is extremely rarely reported in the literature. Aoki et al. described a case of primary postirradiation LMS of the sternum in a patient who had received irradiation due to mediastinal seminoma 11 years ago. The management plan included preoperative chemotherapy with cisplatin, doxorubicin and IFOVADEC (ifomide, vindesine, doxorubicin, and dacarbazine), as well as resection of the upper one-third of the sternum, the medial one-third of the bilateral clavicles, and the anterior part of the first, second, and third ribs. The authors mention that the patient re-





**Fig. 4.** During immunohistochemical evaluation, tumor cells stained positive for smooth muscle marker caldesmon (20×).



**Fig. 5.** Positivity for desmin is also evident among sarcomatous cells (20×).

mained recurrence-free during a three-year follow-up [5]. A case of chest wall leiomyosarcoma affecting the left 7<sup>th</sup>, 8<sup>th</sup>, and 9<sup>th</sup> ribs was also described by Nagai and colleagues, which was managed with chest wall resection and reconstruction with polypropylene mesh. The patient underwent a total of three liver metastasectomies, three lung metastasectomies, and proton-beam radiotherapy for sacral metastasis, and was still of recurrence 14 years after chest surgery [10].

Leiomyosarcomas, however, are more common in uterus, gastrointestinal tract, and retroperitoneum, presenting as abdominal masses [11]. Particularly, the retroperitoneum is a common site of origin of leiomyosarcomas, constituting 12–69% of cases [12]. Therefore, clinical examination and a thorough investigation are important to distinguish a primary tumor from a metastatic one [5]. In our case, this was achieved by core needle biopsy.

Primary bone leiomyosarcoma should be managed by multidisciplinary teams in specialized reference centers [2]. Surgical excision with negative margins remains the cornerstone of management [2]. Adjuvant therapy with chemoradiation is recommended in case of

positive specimen margins [6]. However, leiomyosarcoma of the bone is accompanied by metastasis in 50% of patients within 33 months, especially in those patients treated by surgical resection only, as in the case presented [5].

Few data are available regarding optimal management of metastatic leiomyosarcoma of the bone [5]. A case series by Rekhi et al. showed that metastatic potential in these tumors is high, independent of tumor stage [13]. In addition, metastatic disease is more likely to present within the first year after the appearance of the primary tumor, with the lungs being the most common site of metastasis, while metastases to the liver, adrenal glands, kidneys, and lymph nodes have also been reported, but with a lower rate [2]. Chemotherapy is often used in combination with personalized management and is also the main option for palliative treatment [5]. Recine et al. highlight the poor outcome of surgical resection of metastatic leiomyosarcoma of the bone, while some other authors propose that complete surgical resection of the tumor offers longer overall survival rates [2]. According to Katsuhiko et al., metastasectomy for patients with advanced soft tissue sarcomas should be performed only if potential advantages

outweigh disadvantages [14]. Palliative cryoablation has also been proposed for advanced unresectable malignancies causing compression effects [15]. So, no commonly accepted guidelines have been established about the management of metastatic leiomyosarcoma of the bone due to poor prognosis and limited cases reported.

## Conclusion

Primary leiomyosarcoma of the bone is a scarce but aggressive clinical entity, with few reports of cases originating from the sternum. More clinical trials are necessary in order to improve the knowledge of the disease's biology and metastatic potential, as well as to enrich our armamentarium with targeted therapeutic agents. There is no consensus and established guidelines regarding the role of surgery and adjuvant therapy in metastatic disease. However, minimally invasive surgery seems to offer the opportunity for excision of retroperitoneal metastatic lesion with minimal morbidity and mortality rates.

## Statements

All authors undersigned certify that they do not have any financial or personal relationships that might bias the content of this work.

All authors undersigned certify that they have the written consent of the identifiable person in order to present the case in this scientific paper.

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