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Case Report of a 10 cm Subdued Abdominal Mass: Diagnosis and Management of a Rare Primitive Mesenteric Leiomyosarcoma

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Introduction: Specific management schemes of the exceedingly rare primitive mesenchymal leiomyosarcomas do not exist. Reporting their behavior and management is necessary to be able to construct evidence-based directives.
Case Report: We report a case of a 42-year-old Caucasian woman that presented with a large indolent abdominal mass evolving over 3 months. Two differential diagnosis were looked upon, either GIST or leiomyosarcoma. The abdomino-pelvic ct-scan proved that this tumor could be completely resected, so the patient was successfully operated within 2 weeks of the preliminary diagnosis. A monobloc negative margin resection was performed and the patient recovered on the surgical floor. The anatomopathological studies and immunohistochemistry tests confirmed the presence of a primitive mesenchymal leiomyosarcoma. The patient was recommended a follow up every 4 months with a thoraco-abdomino-pelvic ct-scan due to her economic status.

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Conclusion: The management of rare tumors is always challenging due to the lack of knowledge concerning its behavior and response to the various treatment modalities. Reporting this rare case of mesenchymal leiomyosarcoma and similar cases will enable a better understanding of this disease and its treatment.

Keywords: Leiomyosarcoma; mesenteric leiomyosarcoma; abdominal mass; surgical management.

1. INTRODUCTION

Leiomyosarcomas are soft tissue malignant tumors that represent 10% of all sarcomas [1]. These mesenchymal tumors stem from the smooth muscle lineage and can be found wherever there is a vein, in the retroperitoneum, and the uterus. Out of this rare entity descends an uncommon subtype reported merely 15 times worldwide according to our research on PubMed: The primitive mesenteric leiomyosarcoma. A case series and literature review by kato and Al published in 2016 stated that since 1998, only 13 cases were reported and identified properly by immunohistochemistry [2]. So far, vague diagnostic and treatment guidelines are bound to this malignancy. To add value to our case report, while presenting the 16th case of a confirmed primitive mesenteric leiomyosarcoma, we're going to elaborate the evidence-based process that guided the management at our University hospital in Lebanon.

2. THE CASE

A 42-year-old pre-menopausal Caucasian woman, with no previous medical history, presented for an indolent palpable mass that emerged and expanded over the past 3 months. The patient had no pain but complained of a recent lack of comfort and a pressure like feeling on her left flank while lying on her back. Moreover, she mentioned during her interrogatory a loss of 5 kg during this period without changes in her dietary habits. The patient had no other gastrointestinal, gynecological, or urinary symptoms and did not smoke. Her surgical history included two c-sections and a rhinoplasty. She had a clean family history free of any form of familial cancer. Her physical exam showed a 10 cm indolent mass extending from her left flank down to the left lower quadrant.

She was prescribed blood tests including CEA, CA19.9 that came back within the normal ranges. The abdominal ultrasonography revealed a 10 cm solid mass within the left lower quadrant displaying features that favor a mesenchymal tumor (Fig. 1). Upon these results, an abdominal-pelvic CT scan was performed to identify adequately the relationship of the mass with its surroundings. It disclosed a large partially necrotic solid mass lesion within the left lower quadrant, displaying features that suggest a mesenteric GIST (Fig. 2). No regional, or distal lymph nodes were identified on the CT-scan, but two non-significant hepatic lesions and multiple small lytic vertebral lesions were recognized. Keeping in mind GIST and sarcomas as the two probable diagnosis a Pet scan was ordered to determine the nature of this lesion and to rule out the presence of distant metastasis. The patient's Pet-scan revealed a large highly suspicious hypermetabolic soft tissue mass with necrotic areas in the left abdomen with hypermetabolic foci on both side of the pelvis. These foci represent either tumoral deposits, lymph node involvement or, inflammatory bowel uptake. She had also an increased uptake in the left side suspicious to be originating from the ovary.

Figure 1

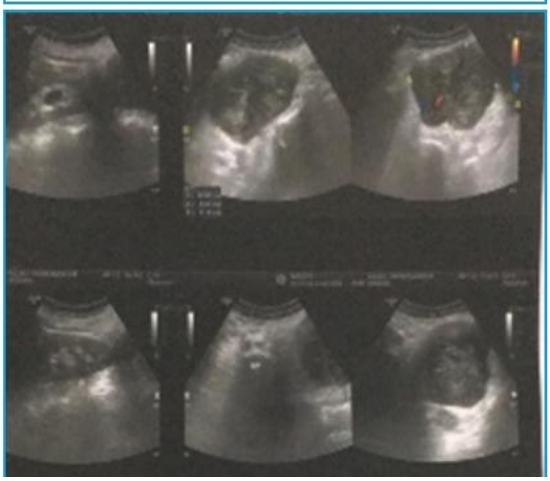

Fig. 1. Ultrasound that shows a focal heterogenous hypoechogenic solid mass lesion within the left

This round mass with homogeneous density and a uniform enhancement could be either a GIST, a lymphoma, a sarcoma, or a metastasis. The hepatic lesions were no longer a threat since the

Pet-scan didn't highlight them. Therefor we were dealing with a localised mass that could benefit from neoadjuvant chemotherapy to reduce its size. Keeping in mind that a preoperative biopsy is not recommended when a GIST is suspected [3], and since the mass can be resected completely with few risks on the surrounding organs the patient scheduled for an operation within two weeks following the extensive paraclinical investigations.

Figure 2

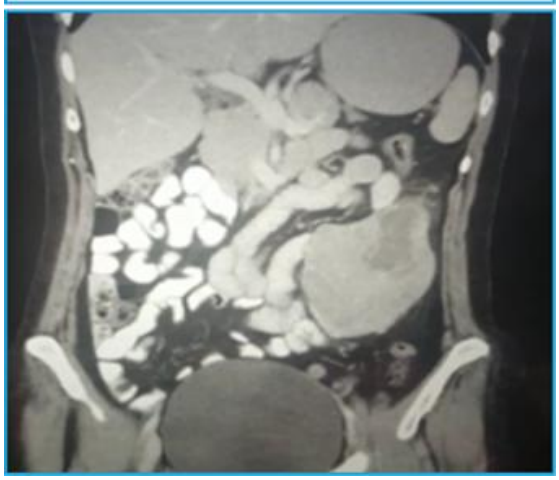

Fig. 2. Ct-scan that shows a large partially necrotic solid mass lesion with smooth edges recognized within the left lower quadrant

The patient was operated and a complete monobloc resection of the growth was performed (Fig. 3).

The tumour was proximal to the left urethra and since the paraclinical imagining couldn't affirm its involvement a ureteral catheter was also scheduled before the resection to avoid its injury. The patient had a supine position hands and legs apart, and after the insertion of the ureteral catheter, through a large median incision the 10 cm growth accolated to the left mesocolon was detached completely in one piece with negative margins. A part of the left colon was sacrificed since its vascular dissection was intricate. A direct end to end colic anastomosis was performed with a left oophorectomy. The left ovary displayed no signs of malignancy, but it was excised because of the increased uptake seen on the Pet-scan. The abdominal exploration showed no signs of carcinosis or hepatic lesions.

Postoperatively, the patient was stable and recovered on the surgical floor. She complained of some discomfort managed by pain killers. The urinary catheter was removed after two days and the patient was discharged after 5 days on pain killers.

The anatomopathological studies revealed a growth of fusiform cells, a sarcoma, that could be either a GIST or a leiomyosarcoma. The immune-histopathological studies confirmed it to be a primitive mesenteric leiomyosarcoma. Due to the rarity of this disease, the pieces were sent to be reexamined abroad. The results were concurrent with ours. Unfortunately, histopathological photos are not available because of a technical problem.

There are no guidelines set out for this specific subtype of sarcomas. The patient was followed up according to the leiomyosarcoma follow up guidelines. Due to financial issues the patient was advised to revisit us every 4 months for the next three years with a thoraco-abdominal-pelvic Ct-scan. The thoracic scan was added to the recommended abdominal-pelvic scan instead of a chest X-ray since leiomyosarcomas metastasis frequently in the lungs. Her first visit to our clinic after 4 months showed a completely clean scan with no signs of metastasis or new growths.

3. DISCUSSION

According to the problem solving in oncology, there are several investigational tools available to complete the assessment of an abdominal mass. An ultrasound can identify the growth and provide information on its origin and nature. It is highly sensitive and specific for diagnosing a cystic abdominal lesion [4]. However, its efficacy in localizing the origin of the lesion is not valuable, especially in cases of a large mass. Computed tomography (CT) allows accurate detection and characterization of abdominal tumours, which helps the surgeon prepare for the surgery. Moreover, Investigators concluded that both ultrasounds and CT-scans are excellent diagnostic tools of a clinically suspected abdominal mass, with sensitivity and specificity values exceeding 95% [4]. However, CT-scans are slightly better in detecting the origin of the mass and predicting the pathologic diagnosis. A study in 1981 proved that a CT-scan can accelerate the diagnosis and reduce the overall cost of hospitalization [4]. So, when the patient presents with a large abdominal mass, it is preferable to get a Ct-scan pre-operatively.

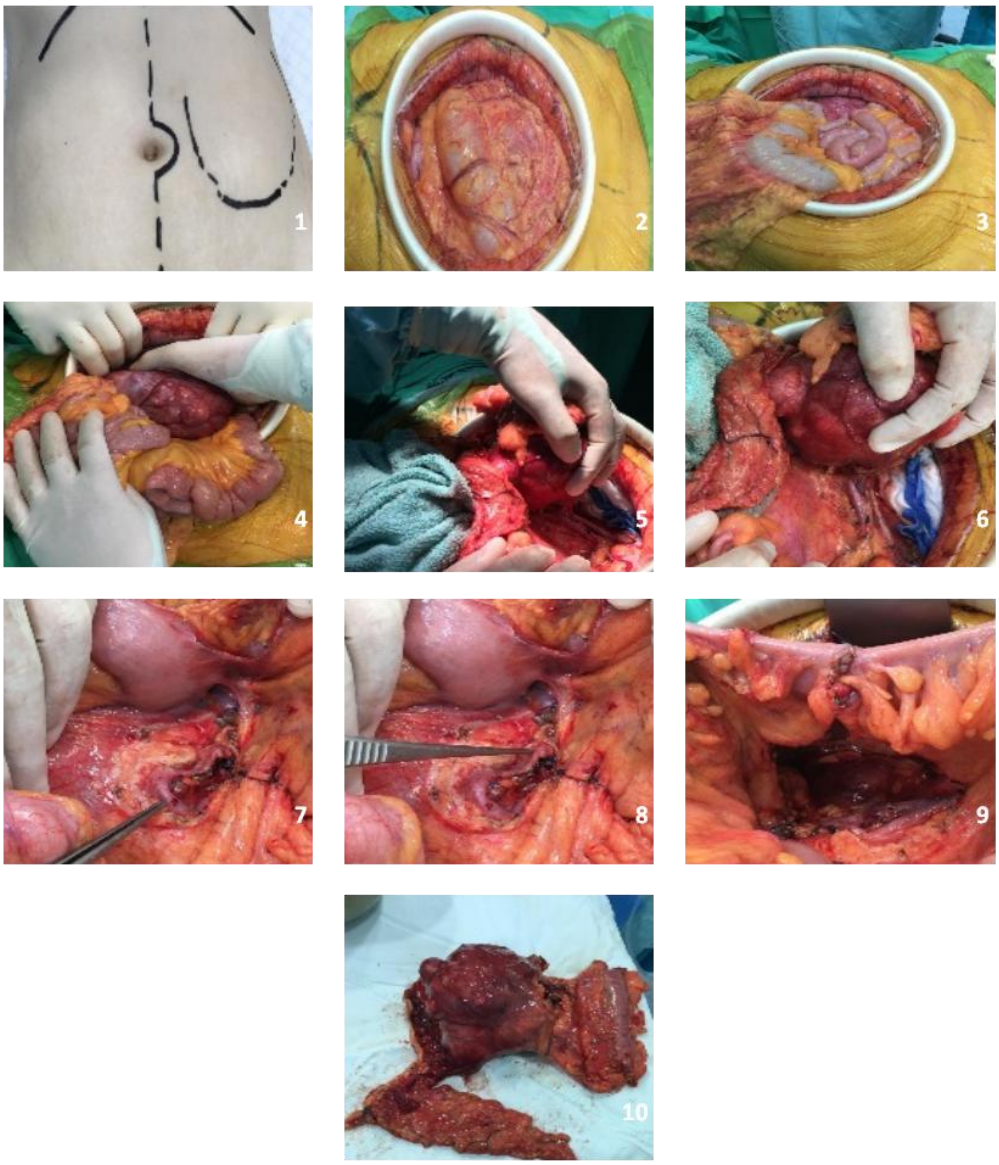

Fig. 3. The operation. 1. Delimitation of the mass and the incision line. 2,3,4. Exposure of the mass. 5,6. Attachment of the mass to the mesentery. 7. The inferior mesenteric artery. 8. The left colic artery. 9. Terminal colic manual anastomosis. 10. The mass with its colic and mesenteric attachment

A meta-analysis done in 2004 on the value of pet-scan in the detection and grading of sarcomas, showed that a PET can discriminate between sarcomas, benign tumours and low- and high-grade sarcomas based on the mean SUV [5]. It is not indicated as part of the standard treatment of a sarcoma however it should be used to detect and grade sarcomas, and to evaluate the treatment of a locally advanced case. Another study published in 2018 in clinical sarcoma research concluded that PET–CT in high-grade bone and soft tissue sarcoma can add significant benefit to routine CT/MRI staging [6]. They estimated a 21% overall potential benefit for PET–CT over CT/MRI, especially by 'upstaging' the grade of the disease. 12% of cases were upstaged to a M1 following the PET–CT [6].

Primitive leiomyosarcomas of the mesocolon are extremely rare. Leiomyosarcomas are soft tissue malignant mesenchymal tumours that derive from the smooth muscle lineage that commonly metastasize to the liver as well as the lung [7]. Their incidence ranges between 10% and 20% and it increases with age and peaks at the age of

70 [8]. Leiomyosarcomas are second to liposarcomas as soft tissue and abdomino-pelvic sarcomas [9]. Usually their incidence and location are influenced by gender: women have a higher incidence of retroperitoneal and inferior vena cava leiomyosarcomas where as men present more with non-cutaneous and cutaneous soft tissue sarcomas [1]. Uterine leiomyosarcomas increase in the perimenopausal years and they are the most common site specific leiomyosarcomas [1]. Few clear causal or predisposing factors have emerged, however, Epstein-Barr virus (EBV) infections in the setting of severe immunosuppression could be linked to their development [10]. No solid evidence was found in literature discussing a possible malignant transformation of leiomyomas.

Typically, leiomyosarcomas do not produce symptoms until they grow large enough to compress or invade contiguous structures [1]. Exceptionally pain, early satiety, and obstructive gastrointestinal symptoms may occur early in the disease course. Because of the elasticity of the mesentery, mesenteric leiomyosarcomas grow to large sizes by the time of diagnosis. Their relationship with highly mobile and flexible tissue enables them to occupy the intraperitoneal space without causing early obstructive symptoms.

Anatomopathologically, leiomyosarcomas have cells displaying features of the smooth muscle lineage. Commonly they are reactive for actin, desmin, and h-caldesmon. Unlike GIST tumours, leiomyosarcomas are negative for CD117 on immunohistochemistry [8].

Generally, in case of an unknown abdominal mass diagnosed on a physical exam, physicians should always look for its primary site of origin. After a complete medical history and physical examination basic laboratory tests could be ordered with tumour markers to help guide the diagnosis. Imaging will determine the resectability of a growth or therefor the need of a FNA biopsy. As mentioned earlier, the ultrasonography can determine the nature of the tumour with a vague definition of the relationship of the mass with its surroundings. This will be solved with a Ct-scan that could be initially ordered without an ultrasound since studies proved it to be superior keeping in mind money and time factors. The Pet-scan will determine the distant and local metastasis and its specifically helpful for a factual staging of a leiomyosarcoma.

Even though these tumors are generally large at presentation and often invade vital structures, most of these tumors are resectable with negative margins [1]. Nodal dissection is unnecessary since nodal involvement is very rare. The kidney, colon, pancreas, and spleen are the most commonly resected organs. Pathologic evaluation after complete resection is necessary for a definite diagnosis. In case of an unresectable mass, biopsy is required preoperatively. Unresectable tumours usually have either an extensive vascular involvement, peritoneal implants, or distant metastasis. Spinal cord involvement and mesenteric root attachments will also prevent complete resection of the growth.

Because of the low tolerance to radiation of the abdominal and retroperitoneal organs, delivery of adequate radiotherapy is often difficult in case of unresectable tumors. There is encouraging evidence for intraoperative radiation therapy to the tumor bed, but this technique is still considered investigational and can be performed only in select centers. Due to its rarity and low incidence, the standard chemotherapy regimen for recurrence and metastasis is not yet established. The role of postoperative chemotherapy remains unproven, uterine leiomyosarcoma seems to be more responsive to many chemotherapy agents, in contrast with visceral leiomyosarcomas [9].

In general, the prognosis depends of the grade size and location. Mesenteric leiomyosarcomas have a poor prognosis with an overall 5-year survival limited to 20% [2,7]. The prognosis improves with a negative margins surgical resection [11,12]. The follow up procedure is based on the type of resection. Complete resection enables a physical exam and a thoraco-abdomino-pelvic scan follow up every three to 6 months for the first 3 years then annually. Whereas, sarcomas that have been partially resected are recommended to be followed up with a CT-scan and physical examination every 3 to 6 months for 3 years then every 6 months for the next 3 years and finally once a year [2,13].

4. CONCLUSION

Mesenteric leiomyosarcomas are extremely rare therefor reporting them is a necessity to be able to establish better evidence-based management guideline.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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