UTERINE LEIOMYOSARCOMA – A CASE REPORT

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ABSTRACT

Uterine leiomyosarcoma is a rare malignant neoplasm with a very poor prognosis and higher prevalence in pre- and peri-menopause. It accounts for only 1-2% of uterine malignancies. We report the case of a 75-year-old postmenopausal woman who presented a large abdominal mass compatible with uterine leiomyoma. She underwent a total abdominal hysterectomy with bilateral salpingo-oophorectomy obtaining a surgical piece weighing 9820g. The diagnosis of Uterine leiomyosarcoma is made by histopathological examination after surgery. This clinical case demonstrates that it is essential to consider a possible malignancy diagnosis to reduce the prevalence of occult Uterine leiomyosarcoma.

KEYWORDS

Leiomyosarcoma, Diagnosis, Uterus, Hysterectomy

Introduction

Uterine sarcomas account for 3-8% of all uterine malignancies, with leiomyosarcoma (LMS) being the most frequent histological subtype [1]. Uterine LMS is a rare and extremely aggressive malignant mesenchymal tumour of the smooth muscle lining. It represents 1% of all malignant diseases of the female genital tract, accounting for approximately 60% of all uterine sarcomas. It usually presents as a voluminous tumour in women over 40 years of age. It is associated with nonspecific symptoms, most often vaginal bleeding (56%), palpable pelvic mass (54%), or pelvic pain (22%) [2]. However, most women are asymptomatic at the time of diagnosis. In rarer cases, the LMS can prolapse through the cervix [3]. Uterine LMS have complex genetic aberrations without any specific molecular or genetic marker and are often diagnosed in the advanced stages of the disease [4]. Magnetic resonance imaging is the ideal imaging technique to characterize uterine masses. However, although certain features are suspicious of LMS, the differential diagnosis with leiomyoma is difficult. In the same way, tumour biopsy is associated with a non-negligible rate of false negative results [5]. In general, in many patients, the diagnosis is made postoperatively when a benign leiomyoma is expected. LMS presents as a malignant tumour with poor prognosis even when confined to the uterus at the time of diagnosis. The 5-year survival rate varies between 18.8 and 68%. Even for stages I – II, the 5-year survival rate varies between 40 and 85%, with a relapse rate of 38–50% [1,2].

Case report

A 75-year-old Caucasian woman with spontaneous menopause at age 50 was admitted to the orthopaedics service following a transtrochanteric fracture of the right femur. She was referred to the Gynecology service due to the presence of a large palpable abdominal mass identified during this period. In addition, the patient reported increased abdominal volume for about 6 months. Physical examination revealed the presence of an abdominopelvic mass, lateralized to the right, non-movable, which extended to the xiphoid appendix. Abdominal ultrasound revealed the existence of a large mass, which occupied the entire hemi-abdomen on the right, exerting compression on the liver and right kidney. It was a mass with a solid echostructure, well-defined borders, doppler colour score 2-3 with minimal cystic areas that could correspond to hemorrhagic or hyaline degeneration in the context of a possible giant leiomyoma. No free fluid was identified in Douglas’ pouch. Contrast-enhanced computed tomography (CT) scan showed a tumour mass measuring approximately 30 x 15 cm, markedly hypervascular, with some areas of central necrosis and calcifications, which extended from the median region of the lower abdomen to the lower liver border, conditioning moulding, and compressing the vesicular and pyelocaliceal excretion systems bilaterally (compatible...
with very large uterine leiomyoma). No other alterations were found, namely adenomegaly. (Figure 1) Analytically, tumour markers CA 125, CA19-9 and carcinoembryonic antigen (CEA) were within normal limits, and serum lactate dehydrogenase (LDH) increased slightly. However, although the main suspicion was based on a uterine leiomyoma, the possibility of malignancy was not excluded, considering the rapid mass increase in postmenopausal women. A total abdominal hysterectomy with bilateral salpingo-oophorectomy was performed, obtaining a surgical piece of total hysterectomy and bilateral adnexectomy weighing 9820g. (Figure 2) The anatomopathological report of the uterus describes the presence of a nodule, on the right lateral portion, with 26 cm of the longest axis, heterogeneous, with hemorrhagic areas and areas of necrosis, compatible with probable LMS. The immunohistochemical exam confirmed the diagnosis. The patient underwent a new CT scan for tumour staging that did not reveal the existence of metastases and was referred for an oncology gynaecology consultation. (Figure 3)

**Discussion**

Uterine leiomyoma is the most common pelvic neoplasm in women, affecting 70% of white women and 80% of black women throughout life. Uterine sarcomas are significantly rarer and have a poor prognosis [1,5,6].

Leiomyomas, responsive to estrogen and progesterone, show greater growth in women of childbearing age. The first symptoms can develop as early as the third decade of life in black women and in the fourth decade of life in white women. Normally, after menopause, leiomyoma stabilizes or shrinks in size. In contrast, increased age is considered a risk factor for the development of uterine sarcomas, which are more frequent after menopause [7].

Leiomyomas do not appear to progress to sarcomas. However, these two entities often coexist [8]. Currently, available imaging methods for evaluation, diagnosis and differentiation between benign and malignant leiomyoma are not very specific, making diagnosing LMS an enormous challenge. Magnetic resonance imaging (MRI) might offer some information but is not entirely accurate. On ultrasound, both sarcoma and leiomyoma present as focal uterine tumours, and both may have central necrosis [9]. Also, the imaging diagnosis proved to be quite challenging in this clinical case. Both ultrasound and CT showed the existence of a giant leiomyoma as the most likely diagnosis, which in some cases can lead to delayed diagnosis. Furthermore, it is known that in more than 70% of women, uterine leiomyoma has a similar symptomatic presentation to LMS, which can lead to a later intervention, as happened in the present case, where the only symptom reported by the patient was an increase of abdominal volume after 6 months of evolution.

Analytically, there may be an increase in serum LDH, often produced by LMS, which may help differentiate from leiomyoma, as in the present case. However, the diagnosis is often unexpected, only confirmed by histopathological examination after a myomectomy or hysterectomy performed due to presumable leiomyoma [9]. The prevalence of LMS presumed to be leiomyomas at the time of surgery ranges from less than 1 to 13 per 10,000 surgeries [6]. Up to 33% of women with a recent diagnosis of uterine LMS have distant metastases, mainly in the liver and lungs [10,11].

In the clinical case presented, the imaging exams, carried out before and after the surgery, did not reveal the presence of distant metastases. This further supported the hypothesis that it was a giant leiomyoma, with the patient undergoing only abdominal surgery hysterectomy with bilateral salpingo-oophorectomy. Uterine LMS, even when confined to the uterus
at the diagnosis, is an aggressive tumour with a high risk of recurrence [12]. Currently, LMS staging is based on the 2017 International Federation of Gynecology and Obstetrics (FIGO) classification, which does not predict the survival of these patients. Thus, some studies suggest the prognostic usefulness of nomograms that incorporate variables such as patient age, tumor size, grade, local extension, distant metastases, and mitotic rate [13].

Conclusion
This clinical case demonstrates that it is essential to bear in mind a possible diagnosis of malignancy to reduce the prevalence of occult LMS. To this end, risk groups must be meticulously determined to prevent late diagnosis of this clinical entity as well as additional surgical procedures.

Abbreviations
- LMS – leiomyosarcoma
- CT - computed tomography
- CEA - carcinoembryonic antigen
- LDH - lactate dehydrogenase
- MRI - Magnetic resonance imaging
- FIGO - International Federation of Gynecology and Obstetrics

Author’s contributions:
Ana Rita Reis Mateus e Ana Carmo Rosa Rodrigues Casquilho devised the project, the main conceptual ideas and the proof outline. Rita Torres Martins worked out almost all of the technical details. Diana Patrícia de Castro Almeida was involved in planning and supervising the work. Ana Rita Reis Mateus wrote the manuscript. All authors discussed the results and commented on the manuscript.

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Conflict of interest
There are no conflicts of interest to declare by any of the authors of this study.

References