






CASE REPORT

Uterine leiomyosarcoma in a teenager: case report and literature review

Leiomiomasarcoma uterino en una adolescente: reporte de caso y revisión de la literatura

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Resumen

El leiomiomasarcoma uterino es un tumor raro, agresivo y de difícil diagnóstico, originado en el músculo liso uterino. Representa solo el 1,0 % de las neoplasias uterinas, pero es el sarcoma uterino más común y con mayor mortalidad. Este afecta principalmente a mujeres perimenopáusicas o posmenopáusicas, con síntomas inespecíficos que simulan leiomiomas benignos. El diagnóstico definitivo requiere de evaluación histopatológica posquirúrgica, ya que no hay biomarcadores o imágenes confiables preoperatorias. Se presenta un caso clínico inusual, en una paciente adolescente, a quien se le indicó manejo quirúrgico radical por vía abdominal mediante histerectomía abdominal total con salpingectomía bilateral, considerada de alto riesgo según lo descrito en el reporte histopatológico e inmunohistoquímico, con receptores hormonales positivos, por lo que en manejo multidisciplinario conjunto con oncología médica y pediatría se encontró apropiado el tratamiento sistémico adyuvante con quimioterapia para evitar y disminuir su tasa de recaída a cinco años y aumentar su pronóstico.

Conflicts of interest

The authors declare no conflicts of interest.

Citation

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Palabras clave: leiomiomasarcoma; leiomioma; neoplasias uterinas; adolescente; informes de casos.

Abstract

Uterine leiomyosarcoma is a rare, aggressive, and difficult-to-diagnose tumor that originates from the smooth muscle of the uterus. It accounts for only 1.0% of uterine neoplasms, yet it is the most common uterine sarcoma and has the highest mortality rate. It predominantly affects perimenopausal

or postmenopausal women and presents with nonspecific symptoms that mimic benign leiomyomas. A definitive diagnosis requires postoperative histopathological evaluation, as there are no reliable preoperative biomarkers or imaging techniques. We present an unusual clinical case involving an adolescent patient for whom radical surgical management was indicated via an abdominal approach, by means of total abdominal hysterectomy with bilateral salpingectomy. The patient was considered high risk based on the histopathological and immunohistochemical reports, including positive hormone receptors. Therefore, in multidisciplinary management involving medical oncology and pediatrics, adjuvant systemic treatment with chemotherapy was deemed appropriate to avoid and reduce the five-year recurrence rate and improve her prognosis.

Keywords: leiomyosarcoma; leiomyoma; uterine neoplasms; adolescent; case reports.

Introduction

Uterine leiomyosarcoma (uLMS) is a rare, aggressive malignant tumor that originates from the smooth muscle of the uterus. It is difficult to diagnose and has a poor prognosis. It accounts for about 1.0% of all uterine cancers, but it is the most common type of uterine sarcoma and contributes disproportionately to uterine cancer mortality due to its aggressive clinical behavior (1-2).

It typically occurs in perimenopausal or postmenopausal women, with a median age at diagnosis of 54 years (2-3). Symptoms may be nonspecific and resemble those of benign leiomyomas, and distinguishing between them usually requires histopathological evaluation after surgical resection, as there are no sufficiently specific clinical or imaging markers to reliably differentiate the two entities before surgery (3).

The primary treatment is hysterectomy with complete resection of the macroscopic tumor. Although it has a poor prognosis, with high relapse rates and low survival, systematic lymphadenectomy and oophorectomy have not shown a clear benefit in improving outcomes (4-5).

This paper reports a rare clinical case: a uLMS-NOS (Not Otherwise Specified) in an adolescent patient with no known risk factors, representing a notable exception to the typical epidemiological profile of this neoplasm. The relevance of this work lies in emphasizing the need to consider this entity in the differential diagnosis of rapidly growing uterine masses, even in young patients,

and it contributes to the current, albeit limited, literature on uterine sarcomas in the pediatric population, with a comprehensive diagnostic and therapeutic approach that justifies the radical treatment performed.

Clinical case description

A 17-year-old nulliparous teenager with no relevant medical history, who had menarche at age 14, began sexual activity at age 16, and started family planning at age 15, has had a subdermal implant for one year and is also using combined oral contraceptives. She denied any sexually transmitted infections. Her mother and father had a history of hypertension and obesity, with adequate blood pressure control. The patient presented to the clinic after 12 months of evolution due to a progressive increase in abdominal volume, associated with irregular menstrual cycles, persistent spotting, and no weight loss. Transvaginal pelvic ultrasound and abdominal computed tomography (CT) scan revealed a large abdominopelvic mass, dependent on the uterus, with heterogeneous features and multiple areas of necrosis, without evidence of adnexal lesions or in other areas of the abdomen or pelvis, suspicious for leiomyoma versus leiomyosarcoma. Additionally, an CT scan of the chest showed a 23 mm x 39 mm lesion in the anterior mediastinum, along with mediastinal lymphadenopathy up to 12 mm at the level of the L2 lumbar vertebra, and signs of a prevascular mediastinal lymph node conglomerate, initially suggestive of an inflammatory/reactive etiology.

Abdominal and pelvic magnetic resonance imaging (MRI) revealed a large solid lesion of uterine origin extending into the upper abdomen, showing heterogeneous signal intensity, obliteration of the endometrial cavity, and patchy, heterogeneous enhancement. After administration of paramagnetic contrast material, the lesion was highly suggestive of uterine sarcoma, measuring 108 mm anteroposterior × 185 mm longitudinal × 164 mm transverse ([Figure 1](#)). Tumor markers were negative (quantitative β -HCG <1.2, alpha-fetoprotein = 0.98; CA 125 = 18.1; CA 19-9 = 2; and lactate dehydrogenase = 165).

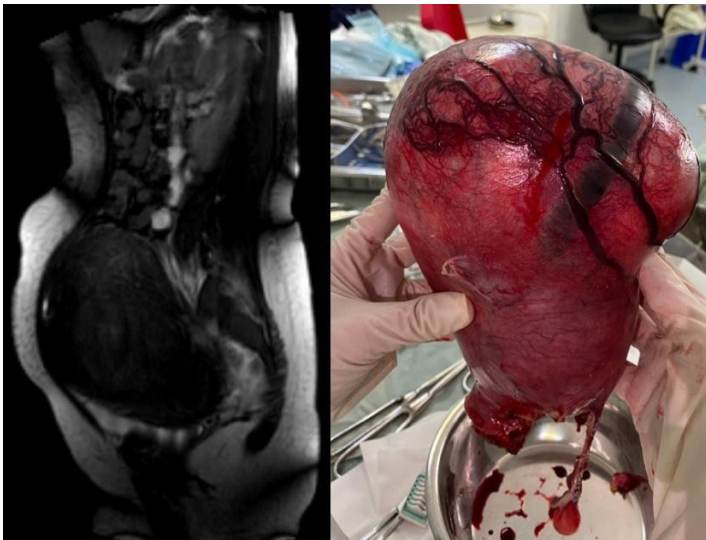


Figure 1. Imaging correlation of nuclear magnetic resonance and anatomical specimen of the uterine-dependent mass.

Open surgical management was indicated. Intraoperatively, a mass approximately 20 cm in size was found originating from the uterus, with necrotic degeneration, serosal, friable, and highly vascularized uterine tissue, without a single, clearly defined mass ([Figure 2A](#)), and compression of the right ureter requiring the placement of a double-J stent. The ovaries and fallopian tubes appeared macroscopically normal, so an extended total abdominal hysterectomy with bilateral salpingectomy was performed via laparotomy to preserve the ovaries ([Figure 2B](#)). The procedure was completed without complications, with intraoperative blood loss of approximately 200 ml, and no blood products were needed before, during, or after the intervention. The immediate postoperative course was uneventful, with a 5-day hospital stay after surgery, for a total of 10 days from admission to the institution.



Figure 2. Intraoperative macroscopic findings of the uterine-dependent lesion. **A.** Intraoperative anatomical specimen of a highly vascularized uterine-dependent mass. **B.** Postoperative anatomical specimen from a total abdominal hysterectomy with bilateral salpingectomy via laparotomy.

The histopathological study reported a 15 cm uLMS-NOS tumor with 80.0% myometrial invasion (8/10 mm thickness), 30.0% tumor necrosis, and 12 mitoses per 10 high-power fields ([Figure 3](#)). The surgical margins, cervix, parametria, and Fallopian tubes were described as free of tumor involvement. Immunohistochemical analysis showed

positivity for vimentin, desmin, smooth muscle actin, myogenin, and h-caldesmon (focal), with a Ki-67 index of 25.0%; negativity for inhibin, S100, TFE3, PAX8, and CK AE1/AE3; and positive hormone receptors (estrogen and progesterone) ([Figure 4](#)). These findings confirmed the diagnosis of uLMS, meeting the criteria for high-grade malignancy.

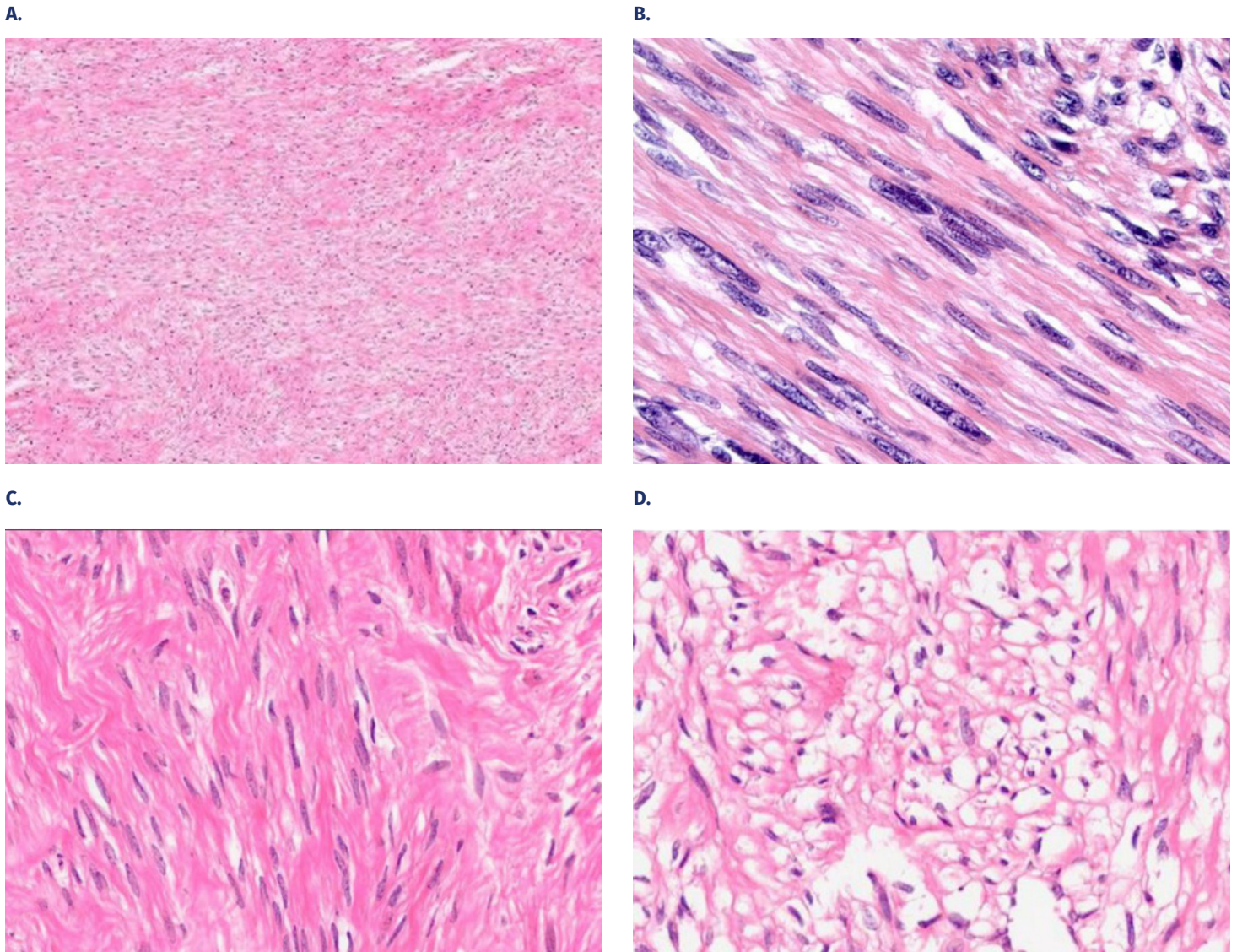


Figure 3. Histological slide. **A.** Tumor in myometrium arranged in cell fascicles and bundles of predominantly eosinophilic spindle-shaped cells, arranged perpendicularly to each other, both longitudinally and *en face*. **B.** Elongated (cigar-shaped) tumor cell nuclei, typical of leiomyosarcoma. **C and D.** Tumor cells with elongated nuclei with blunt ends.

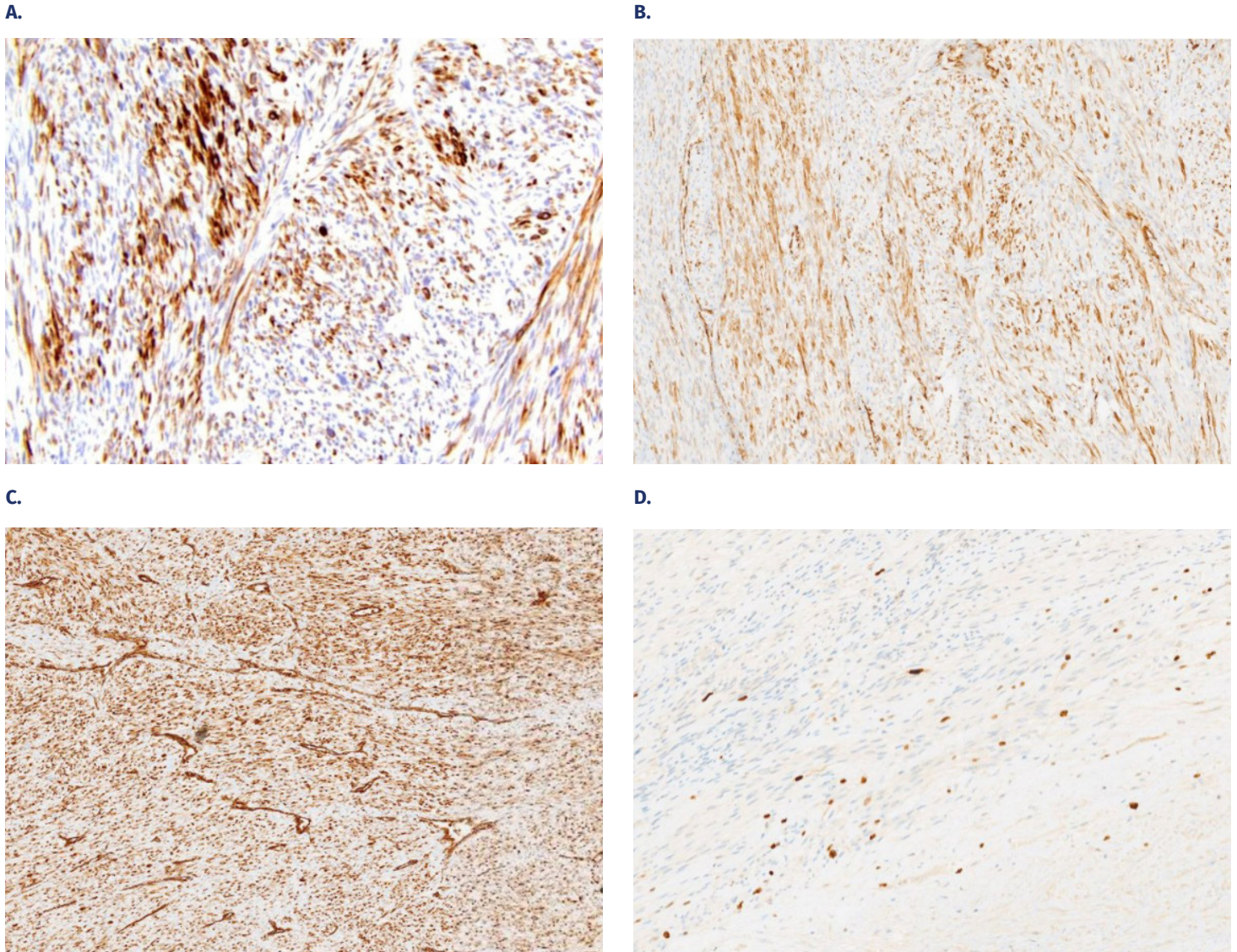


Figure 4. Immunohistochemistry. **A.** Desmin staining. **B.** H-caldesmon staining. **C.** Vimentin staining. **D.** KI-67 staining.

The case was discussed at a medical meeting involving pediatric medical oncology, radiology, gynecologic oncology, thoracic surgery, and surgical oncology. It was concluded that the tumor was high-risk due to the patient's age and tumor size, despite being stage I (T1BNXM0). According to the histopathological results, no sarcomatous overgrowth was evident, but it was considered to have a poor prognosis and a high five-year relapse rate due to the tumor's characteristics, with an estimated disease-free survival of less than 50%. Although positron emission tomography (PET) showed low-uptake mediastinal lymphadenopathies and a thymic mass suggestive of inflammatory/reactive hyperplasia, the decision was made to prioritize initiating adjuvant systemic chemotherapy

with ifosfamide combined with doxorubicin, including mesna for urothelial protection, along with antiemetics and hematological support with pegfilgrastim, and observe the mediastinal lesions, given the possibility that they represent normal thymic activity in adolescence.

Approximately 10 months after surgical management, the patient has completed four cycles of chemotherapy prescribed by the initial medical board and has been closely monitored for 8 months by specialists in medical oncology, gynecologic oncology, and thoracic surgery. During this period, she has not needed any additional interventions or hospitalizations related to relapses of the neoplastic pathology and remains asymptomatic.

A follow-up PET scan shows no evidence of hypermetabolic lesions typical of tumor glycolytic activity or morphological lesions suggestive of locoregional or distant involvement of a known primary tumor, and no signs of masses or lymphadenopathy in the mediastinal area. Over the same 10 months, the previously described mediastinal lesions have remained stable, with no evidence of growth, further supporting the hypothesis of a reactive origin as determined in the initial evaluation.

Discussion

uLMS-NOS is a malignant mesenchymal neoplasm of the uterine smooth muscle and is difficult to diagnose due to its rarity and aggressive nature, with a poor prognosis marked by five-year survival rates of less than 30.0% (1, 3).

Regarding diagnosis, uLMS-NOS lacks distinctive clinical features compared to uterine leiomyomas, making preoperative diagnosis challenging (2). Most cases are diagnosed in postmenopausal women, with a median age of presentation of 54 years, and should be strongly suspected when there is tumor growth similar to a uterine fibroid (6); however, this case involved a patient of reproductive age.

Historically, a woman with a rapidly enlarging uterine mass (defined as an increase of more than 6 cm within 6-12 months) was thought to have a higher risk of sarcoma (7-8), as was the clinical case with this patient. Although later research has shown that both leiomyomas and leiomyosarcomas tend to grow quickly, tumor size or the growth of a pre-existing uterine mass does not necessarily indicate a risk of malignancy (9-10).

In recent years, efforts have been made to develop preoperative diagnostic tests for evaluating suspicious uterine tumors, with each test showing varying degrees and ranges of accuracy (11). Magnetic resonance imaging (MRI), serum lactate dehydrogenase (LDH) markers, PET, and pelvic ultrasound have been proposed for differential diagnosis. MRI has proven to be more sensitive than ultrasound or PET, but the studies examining these findings are small and have low reproducibility (12-13). Diagnostic accuracy improves (up to a specificity of 0.96) when contrast-enhanced MRI is used compared to diffusion-weighted MRI to distinguish a leiomyosarcoma from

a uterine leiomyoma (14). This is mainly because MRI employs multiparametric algorithms that integrate morphological and functional criteria, such as the Oman-Canada Scoring System of Myometrial Masses (OCSSMM), which assigns scores to features such as non-cystic T2 hyperintensity, diffusion restriction, and other findings, thereby classifying lesions into five categories. A score of category III or higher indicates a high risk of sarcoma or smooth muscle tumors of uncertain malignant potential (STUMP), with a sensitivity of 92.3% and a negative predictive value of 98.6% (15).

In the patient described in this report, contrast-enhanced MRI was used as a diagnostic aid, along with PET to identify secondary findings. Although there are few studies on the role of PET in the diagnosis of uLMS-NOS, a small study comparing PET with MRI and ultrasound in five women reported a sensitivity of 100.0% for PET, versus 80.0% and 40.0% for MRI and ultrasound, respectively (16); however, more data are needed, and MRI remains the best current modality for preoperative evaluation of uterine masses and assessing malignancy potential (3).

In some patients with uLMS-NOS, serum LDH levels may be elevated, but the sensitivity of this marker is relatively low (17). In a study of different groups, diagnoses of uLMS-NOS were obtained with positive LDH rates ranging from 14.0% to 66.0% (18). The patient in this report had a normal serum LDH level, consistent with the low sensitivity observed of this serum marker.

According to the available literature on this tumor type, the definitive diagnosis of uLMS-NOS is established by pathological analysis of the resected tumor, that is, after surgical excision, and this remains the gold standard (18). Consequently, the differential and conclusive diagnosis between benign and malignant uterine tumors is exclusively post-surgical and relies on histological examination, which is characterized by significant cytological atypia, high mitotic activity, and tumor necrosis (2). Furthermore, they may also show hypercellularity with severe nuclear atypia, tumor cell necrosis, and a high mitotic rate (≥ 10 mitotic figures per 10 high-power fields), as noted in the histological evaluation of the patient in the reported case. However, it is important to note that the diagnosis can be made based on the presence of at least two of these criteria (19).

Ethical considerations and data confidentiality

The patient's legal representative, in this case her mother, consented to the presentation of the clinical case and images for academic purposes. This document does not contain any personal data that could identify the patient.

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uLMS-NOS can vary depending on cell characteristics, as epithelioid tumors are made of polygonal cells with eosinophilic or clear cytoplasm arranged in nested, cordiform, nodular, or diffuse patterns (20). Although rare, this variant is highly aggressive and may exhibit mild atypia, absence of necrosis, and a low mitotic rate. The cytological features of standard uLMS-NOS usually express smooth muscle markers such as desmin, H-caldesmon, smooth muscle actin, and histone deacetylase 8 (HDCA8) (21), which was also consistent with the findings of the immunohistochemical stains applied to the histology slides obtained from the patient, further supporting the fact that the diagnosis in this case was accurate.

The standard of care for patients with medically resectable disease is hysterectomy with complete macroscopic tumor resection, according to the National Comprehensive Cancer Network (NCCN) guidelines. This should include hysterectomy with or without bilateral salpingo-oophorectomy (BSO) and en bloc tumor resection (20). In premenopausal women, ovarian preservation may be an option, given the lack of evidence supporting a difference in outcomes between patients undergoing salpingectomy versus BSO (21). Although these tumors have a poor prognosis, systematic lymphadenectomy and oophorectomy have not shown a clear prognostic benefit, as uLMS-NOS tend to metastasize hematogenously, evidenced by the low incidence of lymph node metastasis (<5.0%) in early-stage clinical disease (22). In any case, markedly enlarged lymph nodes should be surgically removed; however, due to the absence of imaging and intraoperative lymphadenopathy in this patient, lymphadenectomy was not performed, and only hysterectomy with bilateral salpingectomy was prioritized.

Relapses of this condition are common, even in localized disease, so current research focuses on targeted therapies and immunotherapies, although results to date have been limited (3, 21).

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