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Atypical Uterine Leiomyoma: A Case Report and Literature Review

Viridiana Noemí Victoria Acosta[1], Binui Jesús Kumul Canché[2], Alexis Emir Noguera Echeverría[3], María Fernanda Ibarra Guerrero[4], Javier Navarrete Ayora[1], Roselin Arlet May Román[1], Tamara Espinosa Martínez[1], Alejandra Nuñez Ramírez[1], Héctor Alejandro Soto Hernández[5], Minerva Jiménez Reyes[6], Leonardo Jiménez Reyes[7]

[1]Gynecology and Obstetrics Resident. Hospital General de Cancún "Dr. Jesús Kumate Rodriguez", Secretaría de Salud. Facultad de Medicina de la Universidad Autónoma de Yucatán.

[2]Gynecologist and Obstetrician. Hospital General de Cancún "Jesús Kumate Rodríguez". Universidad Autónoma de Nuevo León.

[3]Surgical Resident. Clínica Hospital Mérida, ISSSTE. Facultad de Medicina de la Universidad Autónoma de Yucatán.

[4]Pediatrics Resident. Clínica Hospital Mérida, ISSSTE. Facultad de Medicina de la Universidad Autónoma de Yucatán.

[5]Gynecology and Obstetrics Resident. Hospital General B ISSSTE, Ciudad Juárez, Chihuahua. Universidad Autónoma de Guanajuato.

[6]Medical Intern. Universidad Anáhuac Mayab.

[7]Medical Student. Universidad Anáhuac Mayab.

ABSTRACT

Introduction: Atypical leiomyomas are diagnosed histologically, showing pleomorphic atypical tumor cells with low mitotic counts and no coagulative necrosis. Surgical treatment is indicated when abnormal uterine bleeding or symptoms related to the size of the leiomyomas, infertility, or recurrent gestational loss are presented.

Clinical case: A 40-year-old woman with a history of menstrual changes, abdominal pain, and intermenstrual bleeding was scheduled for surgery. Pathology reported an atypical leiomyoma (23x18x13 cm) with myxoid degeneration.

Discussion: Uterine leiomyomas affect 70-80% of women in their reproductive age. Symptoms vary by size and location, including abnormal bleeding, pain, and pressure-related issues. This patient had a single pedunculated leiomyoma, no endometrial hyperplasia, negative cervical cytology, and uterine adhesions. Myomectomy was chosen to avoid organ damage.

Conclusion: Atypical leiomyomas are rare tumors diagnosed through biopsy and histopathological examination, displaying 10 mitoses per field and no necrosis. Immunohistochemical studies are necessary to distinguish them from leiomyosarcoma. Long-term follow-up after myomectomy is essential to monitor for recurrence.

KEYWORDS: Atypical leiomyomas, myxoid degeneration, bleeding, myomectomy, biopsy, histopathological examination, leiomyosarcoma.

ARTICLE DETAILS

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INTRODUCTION

Leiomyomas or uterine fibroids are the most common solid benign gynecological tumors of the uterus, originating from the clonal expansion of a single myometrium cell. [(1)] The histological study shows an abundant extracellular matrix surrounded by a fine pseudocapsula of connective tissue and muscle fibers, some cases may contain compressed fibronectin and proteoglycans. [(1)]

They may be single or multiple, although the mechanism of their growth remains not fully understood some authors describe normal XX chromosome components with translocations of chromosomes 12 and 14, with involvement of the HMA2 and HMGA1 gene originating on chromosome 6, therefore considered to be hormone-dependent. [(2)]

EPIDEMIOLOGY

The prevalence of leiomyomas varies between different studies and countries (4.5%-68.6%) according to the racial and ethnic demographics of the population studied. [1] It is a common gynecological problem worldwide, and it is estimated that about 60 to 80 percent of women can be affected by the age of 50. [3, 4]

It is estimated that African American women have a prevalence of myomatosis that is 3 to 9 times higher than other groups. At age 35, 60% of these women are affected, and by age 50, up to 80% suffer from it. [2]

On the other hand, in Caucasian women, the frequency is 40% at 35 years, with an increase to 70% at 50 years and with similar reports around the world, including Mexico; it is estimated that about 25% of women with myomatosis in reproductive age may remain asymptomatic. [2]

RISK FACTORS

The cause of these lesions is unclear, but several factors are known to promote growth, with sexual steroids being the most extensively researched. [3] In addition to ethnicity, other risk factors include obesity, nulliparity, hypertension, late menopause, early menarche, family history of fibroids, and old age. [1] The impact of diet, exercise, smoking, alcohol, stress, and other environmental factors on the pathogenesis of uterine fibroids is not yet clear. [1]

ATYPICAL LEIOMYOMA

In addition to the usual uterine leiomyomas and leiomyosarcomas, there is a group of intermediate or borderline tumors. [5] In 1960, Martin et al. first identified an atypical smooth muscle tumor, which has since been referred to by various names: rare leiomyomas, leiomyoblastoma, clear cell leiomyomas, plexiform tumor, symplastic leiomyoma, foreign-core leiomyoma, and atypical leiomyoma with a low risk of recurrence. [5]

The World Health Organization (WHO) adopted the term epithelioid leiomyoma, and it was later Kurman and Norris who proposed the term atypical leiomyoma, which represents less than 1% of uterine neoplasms. [5]

Originally defined by Bell criteria in 1994, it describes a triad of histological features that included moderate to severe cytologic atypia, less than ten mitoses per ten high power fields (HPF), and no coagulative tumor necrosis. [6]

Atypical leiomyomas are diagnosed by histology and characterized by moderately to severely pleomorphic atypical tumor cells with low mitotic counts and no coagulative necrosis of tumor cells. [7] Despite the histological concerns, most tumors have shown benign behavior. [7]

Individual characteristics such as hypercellularity, necrosis, nuclear atypia, mitotic figures, and intravascular growth are suggestive of atypia but should be interpreted with caution because the variants of benign leiomyomas may contain such changes. [8]

Currently, there are no definitive statistics on the epidemiological characteristics of these patients due to a lack of research on the topic. However, in a case series involving tumors that meet the Bell criteria, the mean patient age was found to be 40.5 years, and the average tumor size measured 7.6 cm. [9]

Nowadays, they are classified into types I and II according to their cytological characteristics. Atypical type I leiomyomas are characterized by round or oval nuclei, distinctive smooth nuclear membranes, prominent nucleoli with perinucleolar halos, and open and coarse chromatin. [9] Type II atypical leiomyomas are characterized by elongated or fusiform nuclei, irregular nuclear membrane, point or null nucleoli, and dark and blurred chromatin. [9]

Immunohistochemistry is a valuable resource for differential diagnosis, especially in cases of difficulty or diagnostic doubt, the most used markers are p16, p53, and Ki67. [5] Several studies have shown varying percentages of Ki-67 among cases of atypical leiomyomatosis, ranging from 0 to 30%. [10] The lack of specific biomarkers and some overlapping histological features between atypical leiomyomas and leiomyosarcomas may hinder diagnosis, and research into molecular and genetic alterations specific to these diseases is ongoing. [11]

Surgery is the therapeutic pillar of uterine leiomyomas, and hysterectomy is the definitive procedure. There are alternative treatments, including endometrial ablation, embolization of the uterine arteries, ultrasound-directed surgery by magnetic resonance imaging, and myolysis. [2] Main indications for surgical treatment include abnormal uterine bleeding or symptoms related to the size of the leiomyomas, infertility, or recurrent gestational loss. [2]
In this paper, we present the case of a 40-year-old female patient with changes in menstrual pattern, abdominal pain, and intermenstrual bleeding, in which an abdominal-pelvic ultrasound is diagnosed as a uterine myomatosis of large elements, requiring surgical treatment and reporting in the postoperative findings atypical leiomyomatosis.

CASE REPORT

A 40-year-old female Mexican without chronic diseases or allergies, with a surgical history of two previous caesareans and bilateral tubal occlusion as a family planning method, with a history of changes in the menstrual pattern, abdominal pain, predominantly hypogastrium, related to menstrual bleeding, with little response to analgesics, which increased in the last two months, similarly referred intermenstrual bleeding for six years. It was evaluated in the external consultation of gynecology, where an abdominal tumor of approximately 30 cm was identified, painful to superficial and deep palpation.

An endovaginal ultrasound was performed however, due to the characteristics of the tumor, an abdominopelvic ultrasound was necessary, which reported a uterus of 20x7x6 cm, with irregular contours, with a myoma of large size, FIGO 6, with measures of 17x10.4x16 cm, with central vascularity and an endometrial wall of 7.4 mm, homogeneous; the ovaries were reported without apparent alterations.

The diagnosis of uterine myomatosis of large elements was integrated, and it was decided to perform an elective way, a total abdominal hysterectomy. As part of the surgical protocol, an endometrial biopsy was performed before surgery, which reported stromo-glandular glandular dissociation, negative for malignancy.

Four months after the diagnosis, a total abdominal hysterectomy was scheduled. During the surgery, a tumor approximately 20 cm in diameter was found at the uterine fundus. The tumor exhibited large caliber neovascularity and had formed multiple adhesions to the abdominal wall, omentum, and left ovary **[Image 1]**. Additionally, there was a moderate presence of ascitic fluid in the pelvic cavity.

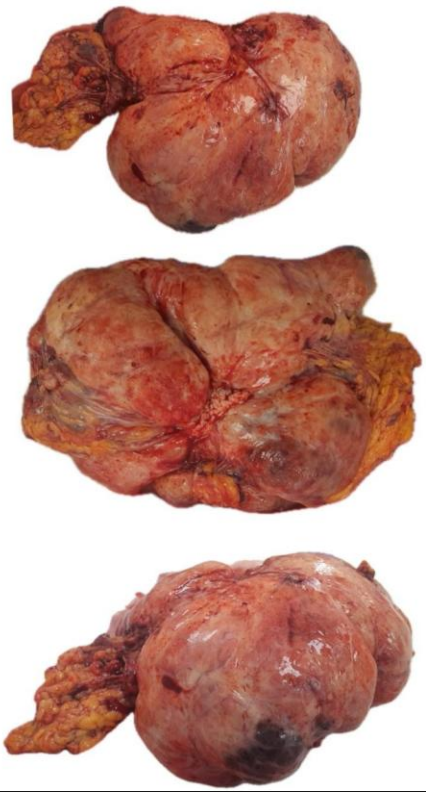

Image 1. Atypical leiomyoma macroscopic structure with adherences and the epiplone observed.

Adhesiolysis and myomectomy were performed, finding a uterus of 6 cm in diameter, without structural alterations, but strongly attached to the abdominal wall and bladder. Therefore, it was decided not to release uterine adhesions due to the high risk of bleeding. Excision of the pelvic tumor was performed with left oophorectomy, reporting a total bleeding of 1700 ml, requiring transoperative blood transfusion. **[Image 2-3]**.

The pathology department reported a tumor of 23x18x13 cm, lobulate, with white-greyish coloration and vinous areas, rubbery to semi-firm in consistency, with a yellowish-white interior and extensive areas of myxoid appearance and focal hemorrhage. The histopathological diagnosis of an atypical leiomyoma with extensive myxoid degeneration and hemorrhage, without necrosis zones, with a mitotic index of 3/10 high-power fields and the presence of congestive omentum attached to the myoma. Suggesting long-term follow-up for the possibility of recurrence or aggressive behavior.

During the patient's hospitalization, analgesic and antibiotic management were indicated. Subsequently, postoperative blood tests were conducted, indicating a hemoglobin level of 8.7 g/dL; therefore, a new blood transfusion was not necessary. The patient continued with adequate general conditions and without early post-surgical complications. The hospital discharge was decided four days after surgery, with outpatient medical follow-up to monitor for possible recurrence or aggressive behavior of atypical myomatosis.

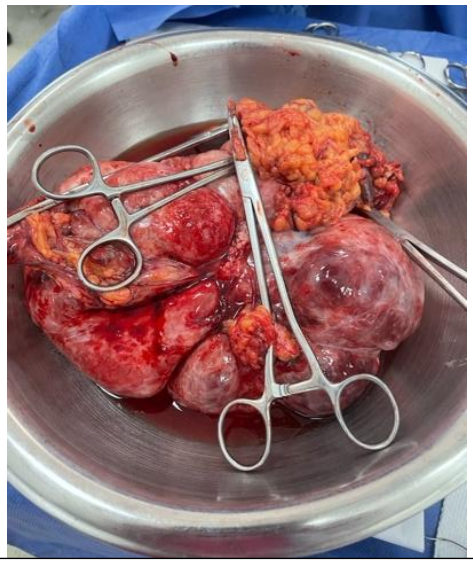

Image 2. Atypical leiomyoma after surgical resection.

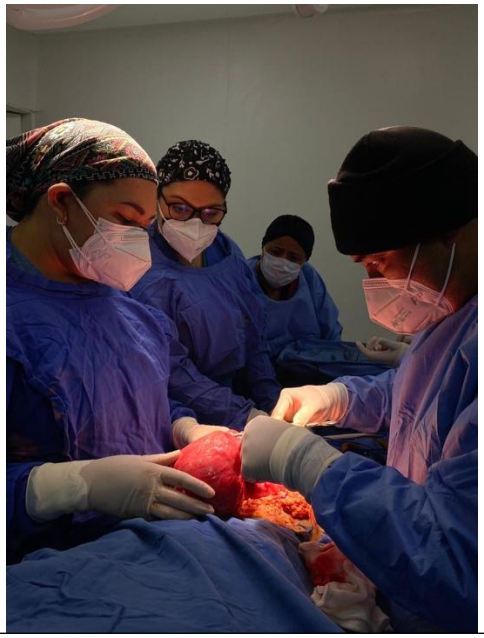

Image 3. Atypical leiomyoma that depended on the uterine fundus.

DISCUSSION

Uterine leiomyomas are one of the most common diseases in women, affecting between 70 to 80% of patients in their reproductive age, the symptoms expressed by patients vary according to their size and location, including abnormal uterine bleeding, dysmenorrhea, symptoms associated with bulking such as pelvic pressure, lumbar or abdominal pain, feeling full, constipation, or increased urinary frequency. [1]

In this case, due to the presence of abnormal uterine bleeding and pelvic pressure, a physical examination needed to be conducted, finding an abdominal mass and a uterine-dependent mass at the bimanual examination. It was decided to use ultrasound as the first line radiological assessment, with a sensitivity of 87% and a specificity of 89% for the diagnosis of uterine myomatosis. [12]

Due to the size of the tumor, an abdominal ultrasound was performed to determine its extent. If the ultrasound is inconclusive, magnetic resonance imaging is the study of choice, with a sensitivity between 86 and 92% and a specificity of 100%. [13] Magnetic resonance imaging (MRI) provides a more accurate assessment of the tumor's location and its relationship with the endometrial cavity. It is also effective in identifying other uterine conditions, such as adenomyosis. [1] However, MRI can be expensive and may not be available in all medical facilities. [1] As a result, this imaging study is typically recommended for women who are considering conservative treatment options. [1]

Magnetic resonance imaging can guide a differential diagnosis between typical leiomyomas, which usually present data suggestive of degeneration, and atypical leiomyomas, according to the cellularity patterns, observing hyperintensity

in T1, T2, or at the contrast application, as opposed to typical leiomyomas which have hypointensity. [13]

Surgery is the basis of treatment for leiomyomatosis, in conjunction with medical follow-up. [5] During the surgical procedure, it is advisable to conduct a transoperative pathology study if macroscopic necrosis or signs of infiltration are present. [5]

In most cases reported in the literature, the diagnosis is made after surgery, and if a myomectomy is performed, long-term medical follow-up is needed, the possibility of recurrence of the tumor. [7]

In this patient's case, only one pedunculated leiomyoma was observed, an endometrial biopsy showed no signs of endometrial hyperplasia, cervical cytology results were negative for malignancy, and multiple uterine adhesions to the abdominal wall and bladder were noted. Given these factors and the risk of damaging nearby organs, it was decided to proceed with a myomectomy. [7]

After the histopathological study, atypical tumor cells with extensive myxoid degeneration and focal hemorrhage were reported without evidence of necrosis zones. These tumors are characterized by moderately to severely pleomorphic atypical tumor cells with low mitotic counts, finding in the patient a 3/10 high-power mitotic index. [7]

Since necrosis or high mitotic counts were not reported, the possibility of a leiosarcoma was not considered, and only long-term follow-up was decided, explaining to the patient the possibility of recurrence of atypical leiomyomatosis.

CONCLUSION

There is limited literature on atypical leiomyomas. The diagnosis is confirmed through biopsy and histopathological analysis, which is characterized by the presence of 10 mitoses per field and the absence of necrosis. If necrosis is observed or if there are fields with suspected mitoses, it is crucial to conduct immunohistochemical studies for differential diagnosis with leiomyosarcoma and to closely monitor for the risk of tumor recurrence. Typically, the diagnosis is made through hysterectomy rather than myomectomy, as described in the case above. Therefore, in instances of myomectomy, long-term follow-up is essential to monitor for any recurrences.

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