

CASE REPORT

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Statement on ethical issues

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Cotyledonoid dissecting leiomyoma of the uterus

Leiomioma disecante cotiledóneo del útero

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ABSTRACT

Benign smooth muscle tumors are the most common tumors in the female genital tract. Cotyledonoid dissecting leiomyoma of the uterus is a rare variant of benign leiomyoma, which presents unusual growth patterns and shares some features with other variants of dissecting leiomyoma. It has an unusual growth pattern characterized by intramural dissection within the myometrium and with similar appearance of its extrauterine components to placental cotyledons. Its sarcomatoid appearance and unusual character give the impression of malignancy. The diagnosis is made by the characteristic histopathologic findings of the lesion. A case of cotyledonoid dissecting leiomyoma of the uterus is presented.

Key words: Cotyledonoid dissecting leiomyoma, Uterine neoplasms, Muscle, smooth.

RESUMEN

Los tumores benignos de músculo liso son los tumores más frecuentes en el aparato genital femenino. El leiomioma disecante cotiledóneo del útero es una variante rara de leiomioma benigno, que presenta patrones de crecimiento inusuales y comparte algunas características con otras variantes de leiomioma disecante. Tiene un patrón de crecimiento inusual caracterizado por disección intramural dentro del miometrio y con apariencia similar de sus componentes extrauterinos a los cotiledones placentarios. Su aspecto sarcomatoide y carácter inusual dan la impresión de malignidad. El diagnóstico se realiza por los hallazgos histopatológicos característicos de la lesión. Se presenta un caso de leiomioma disecante cotiledóneo del útero.

Palabras clave. Leiomioma disecante cotiledonoide, Neoplasias uterinas, Músculo liso.

INTRODUCTION

Benign smooth muscle tumors are common in the female genital tract. Myoma variants account for approximately 10% of cases⁽¹⁾. Leiomyomas with unusual growth pattern include both diffuse, intravenous uterine leiomyomatosis and leiomyoma with vascular invasion, benign metastatic, parasitic, disseminated peritoneal, dissecting, and with perinodular hydropic degeneration⁽²⁾.

Cotyledonoid dissecting leiomyoma (CDL) of the uterus, also known as Sternberg's tumor, is an extremely rare but benign variant of uterine fibroids. It is characterized by the presence of reddish, spongy, bulbous and exophytic extrauterine nodules, similar to the placental maternal face and with a tendency to expand into neighboring structures. Approximately 50 cases have been reported to date⁽²⁻³⁾. Although it is a benign neoplasm, the macroscopic appearance and unusual growth pattern may be similar to malignant lesions⁽⁴⁾. A case of cotyledonoid dissecting leiomyoma of the uterus is presented.

CASE REPORT

This is a 50-year-old female patient, gestation 2, para 2, who was referred to the gynecology office for presenting menorrhagia and bladder urgency accompanied by paresthesia of both lower limbs, of several months' duration. The patient had been diagnosed with uterine myomatosis for 9 years. She reported that both deliveries were spontane-



ous and without complications. She denied the use of hormone replacement therapy, alcohol consumption, smoking, illegal drug use and medical or family history of importance.

On physical examination, the abdomen was soft and depressible, with a soft and mobile tumor. On inspection, the cervix and vagina were normal. Vaginal examination showed the presence of a lesion corresponding to the posterior aspect of the uterus.

Ultrasound found an enlarged uterus with several subserosal uterine fibroids and a heterogeneous, exophytic lesion with irregular margins arising from the posterior aspect of the uterus and measuring 8 x 7 x 6 centimeters, extending towards the right adnexa and fundus of the pouch of Douglas (Figure 1). The cervix was short, the vaginal walls were lax and both adnexa were free. Doppler evaluation assessed low intratumoral resistance index. Abdominopelvic tomography images showed a lobulated mass arising from the right posterolateral uterine wall, with attenuation values similar to soft tissue and extending to the right fornix and right adnexa. There was no evidence of ascites, peritoneal thickening or abdominopelvic lymphadenopathy. The presumptive preoperative diagnosis was gastrointestinal stromal tumor.

Hematology, renal and liver function, electrolytes, urine test and coagulation profile were within normal limits. CA-125 concentrations were 40 IU/L (normal value less than 35 IU/L). The rest of the tumor markers (chorionic gonadotropin, CA19-9, carcinoembryonic antigen and

alpha-fetoprotein) were within normal limits. In view of the findings the case management was discussed with the patient and it was decided to perform surgery.

During the gynecological laparotomy, an enlarged, myomatous uterus was found, together with an extrauterine tumor, soft, mucoid in appearance, with multiple nodules resembling placental cotyledons, originating from the posterior uterine face and extending towards the right broad ligament, pelvic cavity and pouch of Douglas, but without affecting it. The ovaries and bowel loops appeared normal. Frozen section of the lesion suggested smooth muscle neoplasia without cytologic atypia. Total abdominal hysterectomy with bilateral salpingo-oophorectomy, omentectomy and appendectomy were performed.

On anatomopathologic evaluation, the extrauterine tumor measured 8 x 7 x 6 centimeters and protruded from the posterior uterine wall, with multiple deep red nodules that were similar in appearance to the placental maternal face. The nodules were rubbery, reddish, and solid, ranging from 0.5 to 2 centimeters in diameter, and were closely connected by fibrous tissue (Figure 2). Cross sections showed multinodular proliferation of smooth muscle tissue with a cotyledonous architecture.

Histological examination found that the nodules were of variable size, of muscle fascicles with hydropic changes, rich vascularization and marked hyalinized degeneration, separated by abundant intercellular collagen fibers. Smooth muscle cell fascicles showed disorganized and swirling appearance with slightly enlarged nuclei (Figure

FIGURE 1. ULTRASOUND IMAGE OF A HETEROGENEOUS, EXOPHYTIC LESION WITH IRREGULAR MARGINS ARISING FROM THE POSTERIOR ASPECT OF THE UTERUS, EXTENDING TOWARDS THE RIGHT ADNEXA.



FIGURE 2. MACROSCOPIC IMAGE OF THE TUMOR.





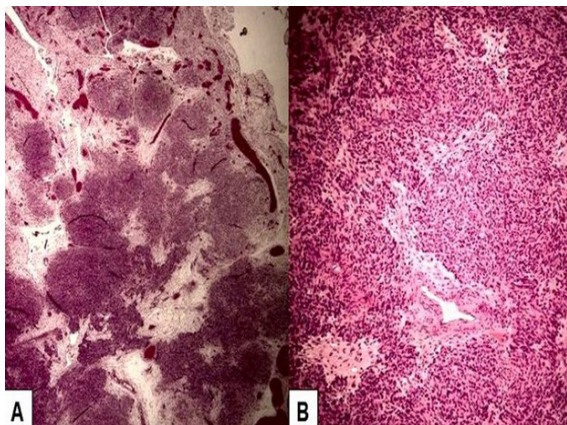
3). Intratumoral vessels were congested and dilated. There was no evidence of atypia, mitosis or necrosis suggestive of malignant changes. On immunohistochemical staining, stromal cells were positive for desmin and smooth muscle actin in tumor cells. Vimentin was positive in intermediate fibrous septa, including vessel walls, and was only focally positive in tumor cells. The Ki67 proliferation index was 2%. The myometrium was diffusely thickened without nodular lesions, and no alterations were found in the fallopian tubes. In view of the findings, the definitive diagnosis was CDL of the uterus without evidence of malignancy.

The patient presented satisfactory recovery during the postoperative period and was discharged three days after surgery. She was monitored with regular postoperative visits at the outpatient clinic, with no evidence of recurrence during the 18 months of postoperative follow-up.

DISCUSSION

Uterine leiomyomas are the most frequent uterine tumors observed between the third and sixth decades of life. Typical uterine fibroids present a typical, well-circumscribed and uniformly expansive growth pattern⁽⁵⁾. CDL (also known as Sternberg's tumor) is an extremely rare variant of dissecting leiomyoma characterized by unusual, exophytic (placental-like), reddish growth pattern with extension into adjacent tissues. To date, less than 50 cases have been described^(5,6).

FIGURE 3. MICROSCOPIC IMAGE OF COTYLEDONOID DISSECTING LEIOMYOMA OF THE UTERUS. A) NODULES OF DIFFERENT SIZES SEPARATED BY LAX FIBROUS TISSUE WITH MULTIPLE BLOOD VESSELS. B) SMOOTH MUSCLE CELLS ARRANGED IN INTERTWINED FASCICLES WITH A DISORGANIZED AND WHIRLING APPEARANCE. HEMATOXYLIN - EOSIN STAINING.



The proposed mechanism of development of CDL is by extension from the uterine wall to the pelvic cavity. Myometrial and stromal glandular cells are trapped in the connective tissue of the intramural component of the tumor, leading to congestion and hydropic changes⁽⁷⁾. There are three types of uterine CDL. The first appears as an exophytic lesion of multinodular tissue protruding from the uterine surface, similar to the placenta. The second type is an intramural dissecting tumor that is confined to the uterus. These two types share similar histopathologic features. The third type is pure cotyledonal leiomyoma, which is not associated with intramural lesion or dissection⁽⁸⁾. The case presented corresponded to the first type.

Clinically, the symptomatology of uterine CDL is abnormal uterine bleeding and abdominal or pelvic lump, the latter usually discovered accidentally. The size can range from 10 to 41 centimeters⁽⁹⁾. Some tumors appear as large fungiform lesions with wide extension into the broad ligaments and pelvic cavity. Due to their low frequency, they can be mistaken for malignant tumors⁽³⁾.

CDL has features similar to typical myoma on imaging studies. On ultrasound, the tumor appears lobulated and more voluminous compared to the typical leiomyoma. However, imaging is not useful for diagnosis⁽¹⁰⁾. Three-dimensional ultrasound can be useful in specifically describing the characteristics of the nodules⁽¹¹⁾. On T2-weighted magnetic resonance images, the tumor is less heterogeneous than sarcomatous lesions, with isointense myometrial-like images on T1-weighted images⁽¹⁰⁾.

Pathologic evaluation shows that the CDL is composed of smooth muscle with a dissecting growth pattern. The extrauterine cotyledonary extension is characterized by nodules of muscle fascicles of variable size surrounded by collagen fibers, with hydropic changes and high vascularization. The neoplastic smooth muscle cells form disorganized fascicles in contrast to the organized pattern of conventional leiomyomas⁽¹²⁾. More than 90% of cases have a dissecting component with infiltrating, sinus-like tongues extending at least 5 millimeters into the surrounding tissue^(1,10).



The vessels are dilated and congested within the extrauterine nodules, while these are round and sometimes hyalinized in the intramural portion. Approximately 20% of cases present vascular invasion; however, it does not change the benign clinical course of the disease⁽⁴⁾. All reported cases have benign behavior despite pathological prognostic factors, such as lymphovascular invasion^(10,13). Unlike other malignant lesions, CDL does not show signs of mitotic activity, nuclear atypia, cellular pleomorphism or necrosis⁽¹³⁾.

Differential diagnoses include hydropic leiomyoma, leiomyoma with perinodular hydropic degeneration, myxoid leiomyoma, intravenous leiomyomatosis, low-grade endometrial stromal sarcoma, perivascular epithelioid cell tumors and leiomyosarcoma⁽¹⁰⁾. Although intravenous leiomyomatosis may be multinodular and involve the broad ligament, the exophytic component is not congestive and intravascular growth is common⁽¹⁴⁾. Multinodularity of the extrauterine component and hydropic connective tissue changes are common in both CDLs and in some leiomyomas with perinodular hydropic degeneration; but the latter does not have a macroscopic appearance similar to placental cotyledons⁽¹⁵⁾.

The standard treatment for CDL of the uterus is hysterectomy, although some cases have been treated conservatively. However, there is one report of tumor recurrence in a single case initially treated conservatively by myomectomy and removal of the extrauterine tumor⁽¹³⁾. To date, no cases of metastasis have been described, which would support the fact of the benign nature of this variant of leiomyoma.

In conclusion, cotyledonoid dissecting leiomyoma of the uterus is a rare variant of leiomyoma. It is a rare benign tumor with multiple placental maternal face-like nodules, different from the typical leiomyoma. Its sarcomatoid appearance and unusual character give the impression of malignancy. The standard treatment is hysterectomy, as conservative treatment may be associated with tumor recurrence.

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