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"The Habit Doesn't Make the Monk" Dissecting Leiomyoma: Report of Two Cases and Literature Review

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Abstract

We report one case of dissecting leiomyoma and one case of cotyledonoid dissecting leiomyoma. Patients were hospitalized for the management of gynecologic bleeding and abdominal pain. The preoperative assessment revealed heterogeneous, fast-growing, possibly malignant, uterine masses. Non-conservative treatment by hysterectomy was performed in both cases. Histopathology of the surgical specimens revealed intramyometrial lesions with dense cellular proliferation, without serous invasion, compatible with dissecting leiomyomas.

We review here the literature and discuss the clinical, radiological and histological aspects of these entities, which can mimic malignant lesions.

Keywords: Leiomyoma; Uterine tumors; Ultrasound; MRI; Hysterectomy; Histopathological analysis

Highlights

- The identification of a leiomyosarcoma before surgery is of utmost importance.
- Rare benign smooth muscle tumors can proliferate in dissecting and extra-uterine growth patterns.
- Differential diagnosis between these entities is challenging to avoid over-treatment.

Introduction

Uterine leiomyomas are the most common benign tumors of the female genital tract. Although usually diagnosed in the preoperative work-up, some types of leiomyoma may mimic malignant forms, and conversely, uterine leiomyosarcoma may be taken as benign conditions [1]. Missing the diagnosis of leiomyosarcoma before surgery can lead to inappropriate surgical management and thus greatly impair patient's outcome [2]. The differential diagnosis between these entities can be challenging. This report describes two cases of dissecting leiomyomas with malignant macroscopic features. Based on available literature data, the preoperative work-up and management of these lesions will be summarized.

Case Series

Case 1

We report the case of a 39-year-old patient, hospitalized in the emergency ward for gynecological hemorrhage. In her medical history, a uterine leiomyoma of 6 cm has been diagnosed six months ago. Clinical examination reveals a large indurated pelvic mass bulging in the pouch of Douglas. Transabdominal and transvaginal ultrasound shows a large heterogeneous mass of presumed uterine origin, with ill-defined margins within the myometrium. Doppler mode shows a rich intralesional vascularization (color score 3). The ovaries are normal and there is no intra-abdominal free fluid. Magnetic resonance imaging identifies a 16 cm × 12 cm, 5 cm × 6 cm uterine mass, heterogeneous on T2-weighted images with hemorrhagic and necrotic components. The upper part of the mass is polynodular with suspicion of uterine serosa invasion (Figure 1a). The lesion involves the uterine cervix but the parametria are free and no lymphadenopathies are visualized. The bladder

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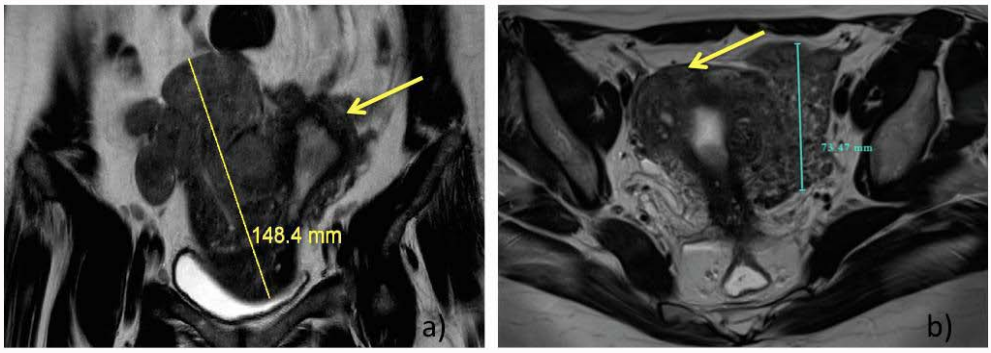

Figure 1: Pelvic MRI on T2-weighted images. a) Case 1: Coronal section, the bulky uterine masse bulges in the pelvic cavity, displacing the uterine body (arrow). b) Case 2: Axial section, the polylobulated uterine masse next to the uterine body (arrow) infiltrates the broad ligament.

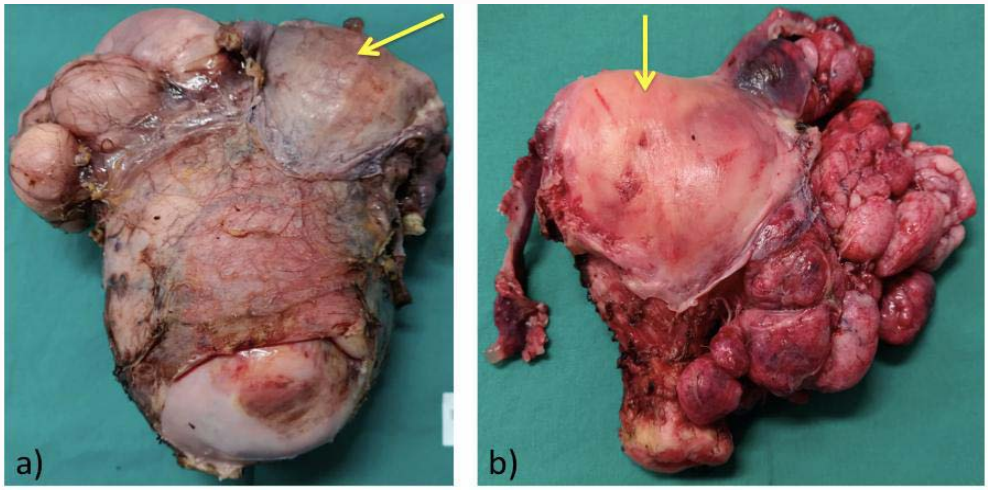

Figure 2: Macroscopic aspects of the specimens showing the polylobulated uterine masses displacing the uterine body (arrows) (a: Case 1; b: Case 2).

and sigmoid are stretched by the mass without any sign of infiltration. The morphological features on MRI and the apparent rapid growth of the lesion are considered compatible with a malignant tumor.

Staging assessment by abdominopelvic CT-scan and chest X-ray shows no metastases. A total radical hysterectomy with pelvic-abdominal exploration was performed by median laparotomy. Macroscopically, the tumor appears polylobulated (Figure 2a) and protruding in the pelvic cavity without involving the adjacent organs. There is no evidence of carcinomatosis. On gross examination, the cervico-uterine tumor appears large multinodular poly-lobulated, fleshy of 12 cm × 9 cm × 8 cm. Histologically, the tumor is composed of pseudo- epithelioid to spindle-shaped smooth muscle cells arranged in interlacing fascicles or swirls and interposed by paucicellular fibrous bands containing vessels. The cellularity is slightly increased, with regular round to ovoid nuclei and complete absence of mitotic figures (Figure 3a). There is no intravascular invasion. Immunohistochemically, the spindle-shaped cells are strongly positive for α-smooth muscle actin, desmin, h-Caldesmon; the Ki-67 labeling index is <1 %.

Case 2

This 42-year-old female is referred for metrorrhagia and pelvic pain. She has been suffering from endometriosis and adenomyosis since several years. Transvaginal ultrasound reveals a polyfibromatous uterus that has doubled in size since the last annual check-up. A pelvic MRI shows an 8.5 cm × 8.7 cm × 4.7 cm subserosal polylobulated uterine mass, located on the left side of the uterus (Figure 1b). The lesion is heterogeneous on T2 and isointense on T1 weighted images, without any sign of restricted diffusion. A laparoscopic total interadnexal hysterectomy is performed. The peroperative aspect is that of a large, exophytic, multinodular "grapelike" tumor extended into the peritoneal cavity with a pedunculated attachment to the uterus in the region of the left broad ligament. This large exophytic mass measures 10 cm × 8 cm × 2.5 cm and is composed of multiple congested bulbous (placenta-like) nodules, varying in size from 0.8 cm to 3 cm (Figure 2b). Some nodules are suspended by threadlike strands. The lesion continues as a white dissecting nodular mass measuring 3 cm × 2 cm × 2 cm in the left lateral myometrial wall.

Microscopy examination reveals benign smooth muscle proliferation in the myometrium that extends beyond the uterus, with an infiltrating growth pattern, a border marked by the dissection of compressive tongues of smooth muscle into the surrounding myometrium with intravascular involvement (Figure 3b). The bulbous processes are composed of nodules of interlacing bundles of bland-looking smooth muscle cells, separated by expanded, edematous, and vascularized stroma. Cellular atypia, mitoses and coagulative necrosis are not detected.

Discussion

Uterine leiomyomas are the most common benign tumors of the

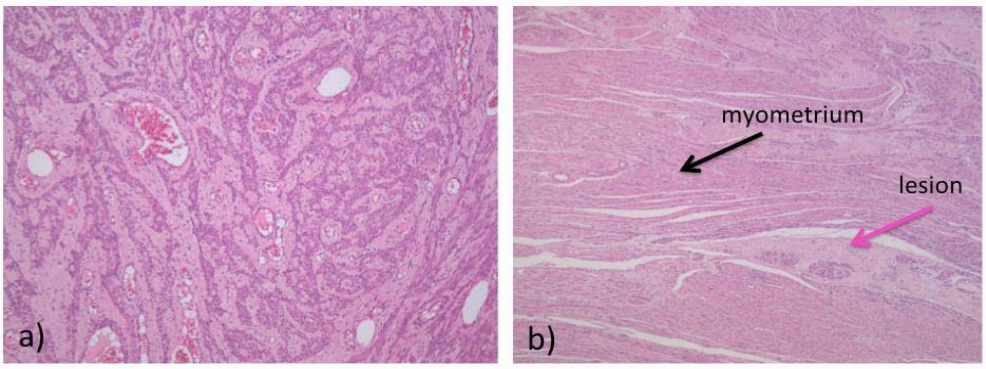

Figure 3: Microscopic examination showing an intramyometrial mesenchymal lesion with an ill-defined border with the adjacent myometrium. Hematoxylin-eosin stain; magnification 50x. (a: Case 1; b: Case 2).

female genital tract. They affect 20% to 40% of women of childbearing age and up to 80% in the peri-menopause. Major non-modifiable risk factors are age, race and genetic factors. Lifestyle risk factors including diet, smoking and physical activity have also been associated with the development of leiomyomas [3]. One third of them are responsible for abnormal gynecological bleeding and/or pelvic pain requiring hospitalization and surgery, which was the case in the two patients presented here. In the majority of the cases, the preoperative evaluation leads to a diagnosis of benign leiomyoma and a fertility-sparing surgical approach is recommended in the reproductive age. However, some forms of leiomyoma may mimic a malignant lesion, leading to a misdiagnosis of a uterine sarcoma and management by radical surgery.

Uterine sarcomas account for 3% of uterine malignancies; leiomyosarcoma is the most common subtype [4]. Their clinical features overlap with those of benign conditions. Imaging is of utmost importance for the differential diagnosis. Ultrasound is often the first-line imaging technique available. Morphological Uterus Sonographic Assessment (MUSA) terms should be used to describe sonographic features of myometrium and uterine masses [5]. Magnetic Resonance Imaging (MRI) is the imaging modality of choice for characterizing atypical uterine masses. Reporting and Data Systems (RADS) criteria have not been yet established for leiomyosarcoma, unlike other neoplasia (breast, prostate and thyroid) [6]. Recently, an MRI predictive scoring system was proposed to differentiate leiomyosarcoma from leiomyoma, based on seven qualitative features [7], but it has yet to be validated by further larger studies. It is of note that all MRI characteristics used for this scoring system were present in case 1 (T2 hyperintensity, projections, necrosis, hemorrhage, T1/T2 heterogeneity, T1 ill-defined borders), suggesting a diagnosis of malignancy.

Cotyledonoid dissecting leiomyoma, also known as Sternberg tumor, is a rare benign uterine tumor that may be confounded macroscopically with a uterine sarcoma. It presents as a heterogeneous bulky mass with irregular margins protruding in the pelvic cavity [8]. Roth, Reed and Sternberg first described this distinct clinicopathological entity in 19969 [9]. Since then, only 94 cases have been described in the literature, with a median age of 44 years [10]. A direct relationship has been found between age and lesion size. The symptomatology is similar to that of typical fibroids: Abdominopelvic masses, gynecological bleeding and/or abdominal pain. Characteristic ultrasound features are a heterogeneous mass with indistinct margins within the myometrium and high vascularization [10]. Pelvic MRI can exclude infiltration of pelvic organs. Typically, cotyledonoid dissecting leiomyomas are isointense on T1 weighted images and hyperintense or heterogeneous on T2 weighted images [10,11]. Usually, diffusion weighted images do not show the presence of restricted diffusion areas in the lesion [12], unlike the images in case 1 suggesting potential malignancy. A non-conservative treatment by hysterectomy is performed in most cases. If a myomectomy is attempted, hormone therapy (GnRH analogues or Ulipristal acetate) can reduce the mass volume before surgery [13]. A radical total hysterectomy was performed in case 1 because of the high suspicion of uterine sarcoma. For case 2, an inter-ovarian hysterectomy was carried out because of abnormal uterine bleeding and the lack of fertility desire. During surgery, cotyledonoid dissecting leiomyomas have a typical placenta-like appearance with multiple cohesive lobules.

Extemporaneous intraoperative examination can be considered to avoid overtreatment [8,10]. The histopathological analysis reveals no signs of malignancy (no atypical cells, no high mitotic activity), but the tumor can have a “dissecting character” extending through the myometrium fascicles [8,10], as found in case 2. Only one case of recurrent cotyledonoid dissecting leiomyoma has been described after a conservative surgical approach by myomectomy [14]. The cotyledonoid variant can exceptionally be seen in its only exophytic form without intra-myometrial continuity; indeed, a case of recurrence of cotyledonoid leiomyoma post-hysterectomy has been described [15].

Conclusion

Uterine leiomyomas are the most common benign mesenchymal tumor in the female tract. Nevertheless, rare benign smooth muscle tumors can proliferate in dissecting and extra-uterine growth patterns, findings that should not be confused with malignant mesenchymal tumors. As these tumors are rare and infrequently encountered, it is imperative that clinicians be aware of this entity. Differential diagnosis with uterine sarcoma is difficult preoperatively, leading to a significant diagnostic and management challenge. These case reports have direct clinical relevance to all operating gynecologists because patients may be inappropriately and inadvertently over-treated for an essentially benign condition.

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