Cotyledonoid dissecting leiomyoma of the uterus with infiltration of tumor cells within intestinal wall: the first Polish case report of a rare uterine tumor and review of the literature

Guz Stenberga macicy z komórkami guza infiltrującymi ścianę jelita – pierwszy polski opis przypadku wraz z przeglądem piśmiennictwa

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Słowa kluczowe: mięśniak, opis przypadku, guz Sternberga.

Abstract

Cotyledonoid dissecting leiomyoma (CDL, "Sternberg tumor") is an unusual type of benign uterine smooth-muscle tumor with a distinctive gross appearance. Although this tumor is benign clinically and pathologically, the appearance and growth pattern are unusual; thus it may be misdiagnosed as a malignancy. A 41-year-old woman was admitted to the hospital with suspected fibroid uterus. The gynecological examination revealed a tumor corresponding to the size of approximately 18 weeks of pregnancy. The transvaginal ultrasound scan showed a 22 × 18 cm sized regular myoma-like contoured mass. She underwent laparotomy. In view of the alarming bizarre appearance, a provisional diagnosis of sarcoma was rendered, and total hysterectomy and bilateral salpingoophorectomy were performed. Simultaneously part of the small intestine adhering closely to the uterine tumor was removed with part of the mesentery. The definite pathologic examination result was cotyledonoid dissecting leiomyoma of the uterus with infiltration of tumor cells within the intestinal wall. The worrying appearance of the gross specimen is often mistaken for sarcoma or non-uterine lesions that may result in overtreatment. It is important to be aware of this entity to prevent overly aggressive treatment and preserve the fertility in young women.

Streszczenie

Guz Sternberga (*cotylenoid dissecting leyomyoma* – CDL) jest rzadko spotykanym guzem mięśni gładkich macicy o charakterystycznym wyglądzie makroskopowym. Mimo że guz jest łagodny klinicznie i histopatologicznie, to wygląd makroskopowy sugeruje złośliwy charakter. Pacjentkę 41-letnią przyjęto do kliniki z podejrzeniem macicy mięśniakowej. W badaniu ginekologicznym stwierdzono guz odpowiadający wielkości ok. 18. tygodnia ciąży. Przezpochwowe badanie ultrasonograficzne wykazało regularny guz o wymiarach 22 × 18 cm, o gładkim obrysie i ultrasonograficznej morfologii mięśniaka. Pacjentkę zakwalifikowano do laparotomii. W związku z niepokojącym, nietypowym wyglądem makroskopowym postawiono wstępną diagnozę mięsaka macicy. Przeprowadzono całkowitą histerektomię i obustronną salpingooforektomię, jednocześnie wykonując resekcję części jelita cienkiego ściśle przylegającego do guza wraz z fragmentem krezki. Ostateczny wynik badania histopatologicznego: *cotylenoid dissecting leyomyoma* z infiltracją komórek guza w ścianie jelita cienkiego. Niepokojący wygląd makroskopowy zdiagnozowanego guza macicy prowadzi często do wstępnej diagnozy mięsaka lub guza o innym niż macica punkcie wyjścia, może to skutkować zbyt radykalnym leczeniem przed otrzymaniem ostatecznego wyniku histopatologicznego. Świadomość występowania tego nowotworu jest ważna w przypadku zachowania płodności u pacjentek w wieku prokreacyjnym.

Introduction

The cotyledonoid dissecting leiomyoma (CDL) of the uterus is a rare variant of uterine leiomyoma. Showing an unusual growth pattern, it was first reported in 1996 by Roth *et al.* [1]. Sometimes it is described as "grape-like leiomyoma" because it resembles cotyledons of the placenta and because of its dissecting myometrial component [1, 2]. There is a variant of CDL called cotyledonoid leiomyoma (CL), which lacked an intramural component with only exophytic component [3]. Approximately 48 cases of CDL/CL have been reported in the English language literature [4]. There is a lack of reports on CDL/CL in the Polish literature.

Case report

A 41-year-old Caucasian woman was referred to the Department of Obstetrics and Gynecology of the Provincial Combined Hospital in Kielce due to pelvic tumor and vaginal bleeding. She had no other systemic disease except dilated cardiomyopathy. Her gynecological examination revealed a tumor filling the small pelvis and lower abdomen, with limited mobility, a size corresponding to an 18-week pregnancy and bleeding from the cervical canal.

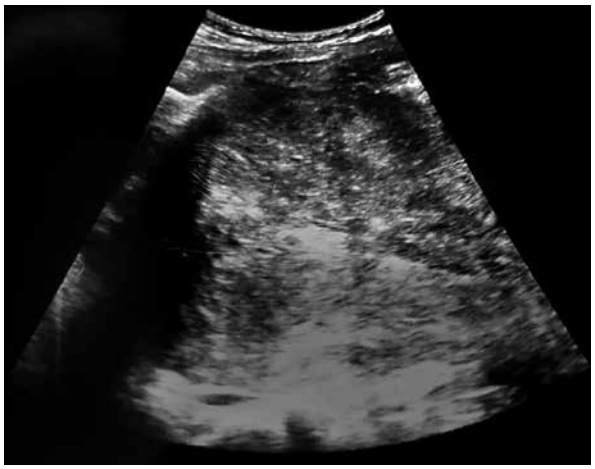

Figure 1. Transabdominal ultrasonographic scan of CDL

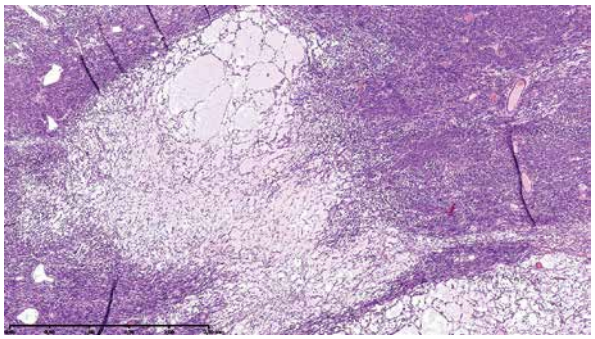

Figure 2. Microscopic features of the tumor composed of fascicles and nodules of bland smooth muscle cells with prominent hydropic degeneration. Original magnification 200×

The transvaginal ultrasound scan showed a 22 × 18 cm hypoechogenic, solid regular myoma-like mass with hyperechogenic regions probably with an origin point from the uterus (Figure 1). The ultrasound did not show the ovaries. The cervical smear result of the patient was normal and endometrial biopsy proved presence of normal endometrial cells.

Laboratory tests revealed mild normocytic anemia (Hb = 10.6 g/dl, Hct 31.3%, MCV = 95.1 fl), leukocytosis (WBC – 20.1 thousand/μl) normal concentration of C-reactive protein (CRP = 1.03 mg/l). The Ca 125 level was 154.3 U/ml. Other laboratory parameters are normal. She underwent laparotomy with a medial incision up to 5 cm above the navel. After opening the peritoneal cavity, the uterus enlarged entirely with a tumor with a tuberous surface with macroscopic grapelike appearance was visualized in close adhesion of about 20 cm of the small intestine. Ovaries and fallopian tubes on both sides were without macroscopic changes. A preliminary diagnosis of uterine sarcoma was made. Total hysterectomy and bilateral salpingoophorectomy were performed. Approximately 12 cm of intestine was removed due to macroscopic infiltration of the serosa. The postoperative period was complicated by anemia (hemoglobin 6.6 g/dl); 2 units of red blood cells and plasma concentrate were transfused. The patient was discharged on the ninth day, postoperatively in good general condition.

Two materials were sent to the Department of Pathology. One of them consisted of a part of small intestine – macroscopically with a thickened serosa on an area of 7 × 5 cm after cutting. The second preparation consisted of a 4.5 cm long cervix and a tumor-changed uterine body measuring 20 × 17 × 15 cm together with fallopian tubes and ovaries. Macroscopically the surface of the uterus was multinodular and lobular. On the cross section the gray tumor was lobularly protruding subserosa; small cysts up to 0.5 cm in diameter were visible. The endometrium was 0.4 cm thick.

On microscopic examination the tumor was composed of fascicles and nodules of smooth muscle cells with prominent hydropic degeneration (Figure 2). It infiltrated within the wall of the removed small intestine (Figure 3). No necrosis and no atypia were found. Mitotic figures were sporadic. Immunohistochemical stains were performed showing strong, positive SMA (smooth muscle actin) (+++) (Figure 4), and desmin (+++) reactions, while Ki-67 was very low, less than 5% (Figure 5). The final diagnosis was "cotyledonoid dissecting leiomyoma" (CDL). Both ovaries and fallopian tubes had normal appearance. The small intestine, except for leiomyoma infiltrates, was unchanged.

During the 4-year follow-up, no relapse was observed in the patient.

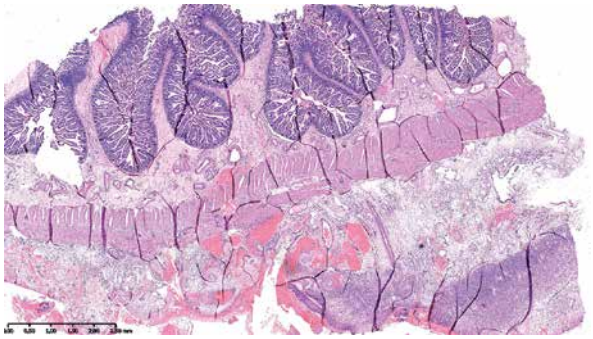

Figure 3. Infiltration of tumor cells within intestinal wall. Original magnification 12.5×

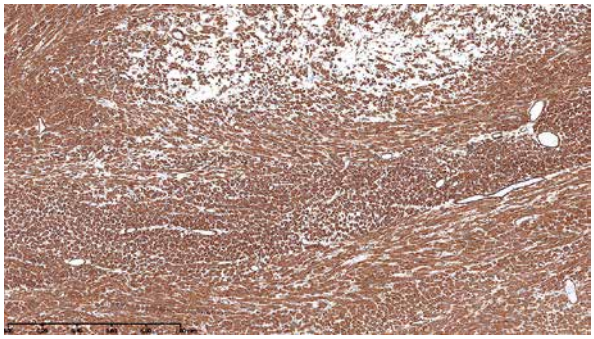

Figure 4. Strong, positive SMA (+++) staining. Original magnification 25×

Discussion

Cotyledonoid dissecting leiomyoma of the uterus is a rare variant of the most common uterine neoplasm – leiomyoma. The most common symptoms are a pelvic tumor and bleeding from the reproductive tract. Pain in the abdominal cavity is less common. In several cases the tumor was also asymptomatic, diagnosed incidentally in cases of imaging of other conditions [4].

This type of tumor is usually large with an average dimension of 17.7 cm (tumors have ranged in size from 6.0 to 41 cm). In most cases the onset of the tumor is located on the back wall of the uterus or around the horn [4, 5]. Imaging and macroscopic appearance of the tumor may raise suspicion of malignancy. Although macroscopically the tumor is usually bulky, reddish and exophytic, and grape-like, which can clinically imitate uterine sarcoma, it has a benign histologic nature without atypia and necrosis. Its pre-operative sonographic view may also imitate a malignant condition [6]. Ultrasonographically, it is frequently seen as an irregular bordered, bulky, solid-cystic mass at the pelvic cavity, descending towards the Douglas pouch, rectum and pressing upwards to the bladder and gastrointestinal tract [7]. 3D/4D imaging can be a helpful tool in ultrasound diagnostics. In 3D reconstructions of the uterus, it can be seen as a lobulated mass with many partially confluent patchy nodules of different sizes, delimited by an irregular capsule [8].

Contrast-enhanced dynamic magnetic resonance imaging can be another useful tool for differential diagnostics. In MRI examination in T1-weighted sequences, the tumor is intense (in relation to the surrounding myometrium), in T2-weighted sequences, it is enhanced in the images obtained after contrast. T2 signal characteristics can be more variable and heterogenic. The signal is increased compared with adjacent leiomyomas and decreased compared to the myometrium. This MRI finding may be characteristic for CDL. Being familiar with the MR imaging diagnosis of this disorder may help determine the precise management strategy, including fertility preservation surgery [9].

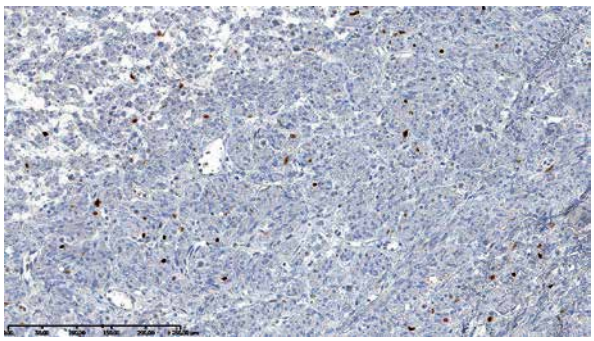

Figure 5. Low index of proliferation, Ki-67, less than 5%. Original magnification 20×

Clinically CDL most often occurs in women in reproductive age [10] with nonspecific clinical presentation. Treatment typically includes: total abdominal hysterectomy with bilateral salpingo-oophorectomy. No cases of recurrence have been reported in the literature. In one published case excision was incomplete, but no recurrence was detected. This finding suggests that the increasing awareness of this tumor will prevent unnecessary surgical procedures, for example total abdominal hysterectomy and bilateral salpingo-oophorectomy during the reproductive period.

Conclusions

CDL is a rare variant of benign leiomyoma with unusual growth patterns, the gross appearance of which leads to a mistaken diagnosis of a malignant tumor. Therefore, both clinicians and pathologists should be well aware of such cases. It is important to recognize that this tumor is a benign and unusual variant of leiomyoma to prevent aggressive surgery, especially if a patient desires to stay fertile.

Conflict of interest

The authors declare no conflict of interest.

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