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Case Report

Uterine Leiomyosarcoma Delivered through the Cervix Cauliflower Appearance, a Case Report, Review of the Literature

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Abstract

Cervical leiomyosarcoma is a rare malignant tumor that develops from the smooth muscle cells of the lining of the cervix. It usually forms in middle-aged women and can cause symptoms such as abnormal vaginal bleeding, pelvic pain, and an increase in the size of the uterus. We report a case of cervically delivered leiomyosarcoma initially diagnosed by anatomic pathology examination of a biopsy of the process delivered through the cervix carried out in an outpatient setting on a 70-year-old woman. This is a special case as the diagnosis is often difficult to make because it can be confused with other forms of cervical cancer or uterine fibroids. Imaging tests such as ultrasound and CT scan can be used to assess the size and the location of the tumor, while a biopsy can confirm the diagnosis. Treatment for leiomyosarcoma of the cervix may include surgery to remove the tumor and, in some cases, the uterus. Radiation therapy and chemotherapy may also be used to reduce the risk of cancer recurring or spreading.

Keywords: *Leiomyosarcoma; Metrorrhagia; Diagnosis and Surgical Management*

Introduction

Uterine leiomyosarcomas are rare uterine cancers and represent between 2 and 6% of malignant tumors of the body of the uterus [1,2]. These are tumors with a poor prognosis characterized by great heterogeneity on the anatomopathological level. Although it is accepted that the standard treatment for uterine sarcomas is surgical, the place of adjuvant treatments remains debated [1,3,4]. The prognosis is poor with recurrence, most often metastatic, in 50 to 78% of cases depending on studies [1-4].

The exact cause of the development of leiomyosarcomas is unknown but certain risk factors have been identified such as previous pelvic irradiation, tamoxifen, age and certain genetic factors (congenital retinoblastoma, Li Fraumeni syndrome) [5-8].

The authors report a case of leiomyosarcoma delivered through the cervix, specifying its clinical, radiological and histopathological characteristics and discuss the diagnostic and therapeutic difficulties.

Case Study

Ms. Naima aged 70 years, G7P7, without history of notable pathologies, of low socio-economic level, was admitted to our department in December 2023 for a mass delivered through the cervix associated with metrorrhagia without an anemic syndrome or urinary or digestive signs. The clinical examination found a patient in good general condition, her conjunctivas well colored, abdominal palpation without particularity and on gynecological examination the vulva was macroscopically healthy, we noted the presence of a whitish cauliflower mass exteriorized by the vulva (Figure 1), delivered through the cervix invading the external orifice of the cervix, vaginal examination found the uterus of normal size with atrophic soft consistency without lateral uterine mass at the level of the Douglas pouch (Figure 2); this mass is distant from the urethral meatus (Figure 3). The remainder of the general examination was unremarkable. Pelvic ultrasound found the atrophic uterus of normal size, homogeneous myometrium, thin endometrium, interrupted at the cervico-isthmic level by an echogenic thickening of 19*14 mm in relation to the base of implantation of the cauliflower process delivered by the ectocervix and externalized at the level of the vulva, the appendages were without abnormalities. A biopsy of the mass was carried out with anatomic pathological results in favor of spindle cell tumor proliferation with a sarcomatous appearance which was completed by immunohistochemistry of P40, CK5/6 which returned in favor of a uterine leiomyosarcoma. We completed with an abdominopelvic scan which came back in favor of a tissue process originating through the cervix and exteriorized through the vagina which seems to be inserted at the level of the right posterolateral part of the endocervix, with a pedicle extended over approximately 6.5 cm and measuring 15 mm thick at its distal end. This process measuring 5*5.5 cm, seems to have extensions at the level of the vaginal fornices. It is attached to the vaginal and vulvar walls, and comes into contact with the urethral meatus, without separating fatty borders. The hemogram showed hemoglobin at 13 g/dl normochromic normocytic. The rest of the preoperative assessment was without abnormalities. On surgical exploration the uterus was atrophic measuring 5*3 cm. The left ovary and right appendix appear healthy. A total hysterectomy with bilateral adnexectomy is performed. The postoperative course was unremarkable. The histological study was in favor of a grade I uterine leiomyosarcoma with healthy adnexa. Faced with this result, a decision to add therapeutic radiotherapy was made. The patient underwent radiation therapy with success. She is currently under surveillance. The chest CT scan was normal.

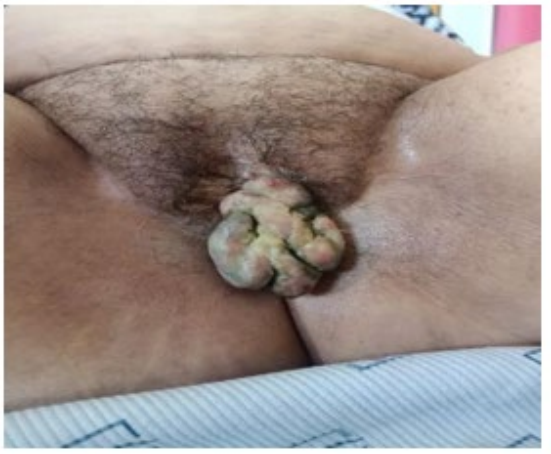

Figure 1

Figure 2

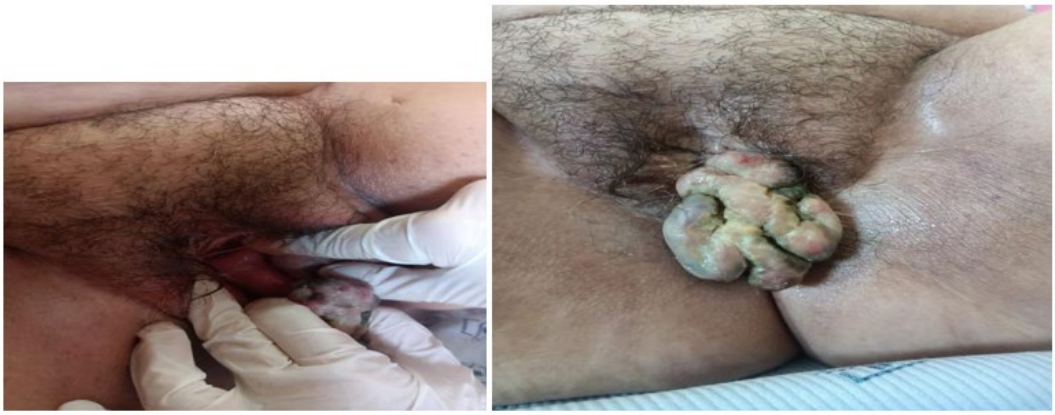

Figure 3

Discussions

Uterine leiomyosarcoma is a rare malignant tumor of a connective nature, developed at the expense of the mesenchymal elements of the myometrium [5]. The relative frequency is 1.3% of all uterine cancers [6] and corresponds to 40 to 50% of uterine sarcomas. Statistics show, there are 2 leiomyosarcomas per 1,000 uterine fibroids [6,7]. They are characterized by great heterogeneity on the anatomopathological level. These tumors have a poor prognosis since the five-year survival is approximately 30% [1-8]. Their diagnosis must be early, because patient survival is correlated with tumor stage [1]. The average age of onset varies from 45 to 55 years [9]. The disease most often occurs in patients who have been postmenopausal for 6 - 7 years. Our patient is 70 years old and is in menopause. The clinic presentation remains not very specific. The two most frequent signs found in our study as in the literature are: genital hemorrhages and pelvic mass [5,7,9]. Preoperative diagnosis is rarely made; leiomyosarcoma most often presents in the form of a banal or necrobiosis myoma. The diagnosis is most often made on a hysterectomy specimen except in our patient the biopsy allowed us to confirm the diagnosis. This is the point of our study. Classically, the rapid increase in volume of a leiomyoma and its softening should raise suspicion of the diagnosis [6]. Rapid growth of a leiomyoma only turns out to be leiomyosarcoma in 0.27% of cases. Ultrasound is not very specific

[6]; leiomyosarcoma is most often limited to a single mass, there is no preferential location and the base is broad or pedunculated. A rapid increase in fibroid size and an image of a necrobiotic myoma can be misleading. Several studies have analyzed the contribution of ultrasound and color Doppler, but have not found characteristics allowing the differentiation of a fibroid and a uterine sarcoma. Other authors comparing indexes and color Doppler did not find morphologic differences between fibromas, leiomyosarcomas, and carcinosarcomas or differences regarding Doppler indexes between fibromas and leiomyosarcomas. On CT scan [10], the lesions appear in the form of large areas of necrosis or cystic transformation, although not specific, should suggest the diagnosis. The CT scan also makes it possible to look for metastases in the lung, mesentery, omentum, retroperitoneal lymph nodes and spleen. These metastatic lesions often have a necrotic center. On MRI, the lesions present a heterogeneous signal on T2 imaging with areas of hypersignal. On T1 imaging, they are iso- or hypo intense compared to the signal from the myometrium. They take on the contrast intensely in the arterial phase of the injection; this contrast enhancement is often heterogeneous. At a later time, it is possible to assess whether there is tumor necrosis [10]. The differential diagnosis is represented by fibroids which are T2-imaging hypointense and homogeneous after injection; the kinetics of contrast enhancement of fibroids is superimposable to that of the myometrium. On the other hand, in the case of a histologically proven lesion, MRI allows the best assessment of local and locoregional extension. On the anatomopathological level, the suggestive appearance of a leiomyosarcoma is the soft, friable consistency, the white color, the size often bulky (on average 10 cm in diameter), the site of necrotic and hemorrhagic changes with sometimes frank invasion of the myometrium and of the peritoneal serosa. The histological criteria used by most authors [5-7] for the diagnosis of leiomyosarcoma are those of Hendrickson and Kempson, taken up by Zaloudek and Norris. The diagnosis is made when there are more than 10 mitoses per field at objective 10 or if there is a number of mitoses between 5 and 9 per field at objective 10 associated with cellular atypia or metastases. Immunostaining with anti-desmin, anti-vimentin and anti-1-anti-trypsin are useful to confirm the smooth muscle nature of the proliferation. Extension [11] occurs contiguously towards the vagina, pelvis and abdomen. Metastases affect, in order of frequency, the lung, liver, bones and brain. Lymphatic extension [11] is towards the pelvic, para-aortic, mesenteric, mediastinal, hiliary and supra-clavicular lymph nodes. The dominant prognostic factor is the mitotic activity of the tumor [5,12], the prognosis is even worse when the mitotic activity is high. The stage of the disease clearly influences the progression of the cancer [12]. The absence of necrosis and peritumoral hyalinization are good prognostic factors. Young age and pre-menopause are good prognosis factors [5,7,12].

All stages combined, 5-year survival is estimated between 25 and 40% [13]. The presence of metastases clearly influences survival. The recurrence rate of leiomyosarcomas varies from 35 to 70% depending on the authors, most often affecting the pelvis. They most often occur within two years of diagnosis. Treatment is essentially surgical and must be complete from the outset [9,12]. The first stage of the intervention consists of peritoneal cytology and exploration of the abdomen. Most authors perform a non-conservative hysterectomy, although some have shown that conservation of the ovaries does not modify survival. Additional procedures depend on the exploration: visceral excision depending on the extension and pelvic dissection if lymphadenopathy is palpated. Radiotherapy reduces the incidence of pelvic recurrences but does not improve overall survival [9]: its place must be discussed on a case-by-case basis.

Chemotherapy may be offered before surgery if the tumor is deemed unresectable from the outset or after surgery if the tumor resection was not optimal or finally in the event of distant metastases. Depending on the histological risk of uterine rupture, postoperative radiotherapy may be proposed.

Conclusion

Leiomyosarcomas are rare diseases with a poor prognosis whose diagnostic and therapeutic management must be provided in a multidisciplinary manner by reference centers with systematic review by an expert pathologist. Treatment is dominated by surgery. Radiotherapy only reduces local recurrences without modifying survival and chemotherapy has not proven its effectiveness.

Conflicts of Interest

The authors declare no conflict of interest.

Author Contributions

All authors contributed to the conduct of this work. All authors also declare having read and approved the final version of the manuscript.

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