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Low-grade sarcoma of the cervix: an unusual diagnosis

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Abstract: Introduction: low-grade endometrial stromal sarcoma is a tumor that represents less than one percent of all uterine malignancies. It is mostly diagnosed between 40 and 55 years of age, it has slow growth, with extrauterine dissemination in 40 and up to 50% of cases at the time of diagnosis. Patient information: 72-year-old woman, with a history of high blood pressure, type 2 diabetes mellitus and surgery for fibroid, along with necrobiosis, who had to undergo total abdominal hysterectomy with double adnexectomy. As a result of the biopsy, submucosal and subserosal intramural leiomyomas with areas of necrosis and hemorrhages were detected. Subsequently, in August 2022, she presented scant bleeding, so she was sent to the Neck Pathology Consultation. She underwent video colposcopy with an exophytic lesion in the vaginal vault. The patient returned to the center due to a tumor measuring ±6 to 8 cm, violaceous, protruding through the vulvar introitus, coming from the vaginal vault and accompanied by bleeding, and friable. The biopsy result determined a low-grade sarcoma-type malignant mesenchymal tumor with two foci of necrosis. Conclusions: uterine cancer is the most common gynecological malignancy. On the other hand, low-grade endometrial stromal sarcoma is a very unusual tumor that ranks second among uterine mesenchymal tumors.

Key words: uterine cervical neoplasms; diagnosis differential; biopsy

1 Introduction

The cervix protrudes into the vaginal vault. The lowest opening is the external os, where the endocervix begins as a narrow channel extending to the internal os or isthmus, from which the endometrial cavity begins. The endocervix is lined with columnar mucinous epithelium and is continuous with the vaginal mucosa [1].

The region in the cervix where squamous epithelium transitions to columnar epithelium is the transformation zone. Most cervical epithelial neoplasms and their precursors occur at the squamocolumnar junction or within the transformation zone [2]. One study found that uterine cancer is the most common gynecologic malignancy, accounting for approximately 50% of all gynecologic malignancies [3].

Low-grade endometrial stromal sarcoma (LG-ESS) is a very rare tumor with low malignant potential. It is characterized by the proliferation of short spindle cells with low nuclear atypia and a low mitotic index. These tumors represent less than one percent of all uterine malignancies and are the second most common uterine mesenchymal tumor

after leiomyosarcoma [4,5].

The five-year disease-specific survival rates are 90% for stage I or II disease and 50% for stage III or IV disease. LG-ESS has an indolent clinical course, although late recurrence or distant metastasis can occur after curative surgery. It is mostly diagnosed between 40 and 55 years of age, with slow growth and extrauterine spread in 40% to 50% of cases at diagnosis [6,7].

We report a case of low-grade cervical sarcoma that presented three years after a total abdominal hysterectomy. Given the low incidence of this neoplasm, it is of interest to describe the diagnostic course of low-grade endometrial stromal sarcoma.

2 Patient information

A 72-year-old patient is presented, with a history of chronic arterial hypertension, treated with atenolol (¼ tablet per day), nifedipine (one tablet per day) and hydrochlorothiazide (one tablet per day); of type 2 diabetes mellitus, controlled with diet and having been surgically intervened at the Provincial University Oncology Hospital "María Curie" of the City of Camagüey, of the province of the same name, in September 2019, for fibroma and necrobiosis; she underwent a total abdominal hysterectomy with double adnexectomy and samples were taken for biopsy (based on the result, submucosal and subserosal intramural leiomyomas were diagnosed with areas of necrosis and hemorrhages, the largest being 8cm in diameter).

Subsequently, in August 2022, she experienced scant bleeding and was evaluated at the Cervical Pathology Clinic of the same hospital. A videocolposcopy was performed, revealing an exophytic lesion in the vaginal vault. Two biopsy samples were taken, with inconclusive results; these were repeated in September with the same result, so an extended biopsy was performed two months later.

One month later, the patient returned to the "María Curie" Hospital with a purplish, approximately 6 to 8 cm mass protruding through the vulvar introitus, originating from the vaginal vault, accompanied by bleeding, and friable (Figure 1). The bleeding led to anemia, for which she received a transfusion of 500 ml of packed red blood cells. Oral tranexamic acid was administered to control the hemorrhage.

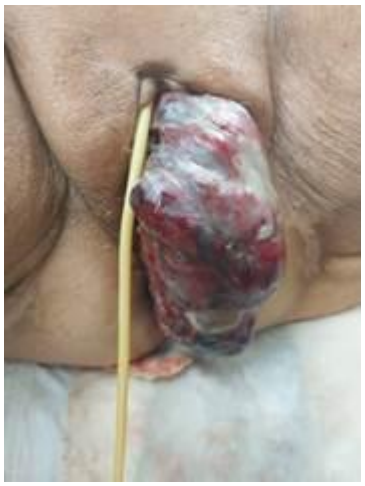

Figure 1. Photograph of the patient showing a tumor protruding through the vulvar introitus

The physical examination revealed moist, hypocolored mucous membranes, non-infiltrated subcutaneous cellular tissue, no dyspnea, with a respiratory rate of 18 full breaths per minute, rhythmic heart sounds of good tone, and blood pressure of 130/80 mmHg.

Abdominal ultrasound imaging studies were performed, which showed a liver with marked, diffusely increased

echogenicity, of normal size, without biliary tract dilation; normal pancreas and spleen; right kidney measuring 115x83 mm, with a parenchyma of 12.3 mm containing a 9.5 mm stone in the mid-calyceal group causing marked dilation of the renal pelvis and slight dilation of the proximal third of the ipsilateral ureter; left kidney measuring 110x83 mm, with a parenchyma of 12.7 mm with a good sinus-parenchyma ratio, without stones or ectasia; empty bladder, with a balloon catheter in place, no free fluid in the abdominal cavity, and no intra-abdominal lymphadenopathy. No solid lesion occupying the intra-abdominal space was demonstrated.

The biopsy results showed a malignant mesenchymal tumor, a low-grade sarcoma, with two foci of necrosis. Follow-up and treatment were decided upon.

3 Discussion

Low-grade endometrial stromal sarcoma (LG-ESS) is one of the rarest subtypes of uterine sarcomas and is indolent in nature; few studies have established definitive treatment guidelines for it. Distinguishing LG-ESS from other tumor types, such as leiomyoma with degeneration or cellular leiomyoma, using imaging studies is difficult [7,8].

LG-ESS is usually identified by postoperative pathological diagnosis after a hysterectomy or lumpectomy for the preoperative diagnosis of uterine leiomyoma; there are reports of LG-ESS being diagnosed during or after laparoscopic surgery [8,9].

Hormonal therapy is not a standard adjuvant treatment for LG-ESS; previous studies [10] indicated that patients with advanced or metastatic LG-ESS might benefit from hormonal therapy, including megestrol acetate or medroxyprogesterone acetate. One study in a patient with metastatic LG-ESS, who responded well to hormonal therapy, showed a very indolent course for a considerable period of more than 15 years, even without any treatment [11].

Surgical intervention is the most important procedure in the management of patients with endometrial stromal sarcoma. Chemotherapy has not had a significant impact on survival outcomes, but it may benefit patients undergoing surgical treatment. The role of adjuvant therapy lies in the prevention and treatment of recurrent, residual, and metastatic lesions; however, the administration of adjuvant radiotherapy or chemotherapy is not routinely used at the LG-ESS, and its role is still debated [12].

Uterine cancer is the most common gynecological malignancy. Low-grade endometrial stromal sarcoma is a very rare tumor, ranking second among uterine mesenchymal tumors. It is important to understand its characteristics to include it in the differential diagnosis of other diseases.

Conflicts of Interest

The author declares no conflicts of interest regarding the publication of this paper.

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MJGB: conceptualization, data curation, research, visualization, drafting, and editing.

GJCC: research, writing (review and editing).

DCP: original draft writing, data curation.