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Leiomyosarcoma of the vagina with invasion of the rectum as cause of abdominal sepsis. A case report

Leiomiosarcoma de vagina invasor a recto como causa de sepsis abdominal. Reporte de un caso

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Palabras clave:
Sarcoma de vagina, recto, invasor, alto grado, displasia.

ABSTRACT

Cancer of the vagina is the least common gynecological tumor, representing only a 2%. The most common of these tumors is the squamous type (75-90%), and the least common, the sarcoma (< 3%), of them being the leiomyosarcoma the most common. We present a case report of a 40-year-old female who came to the emergency room with abdominal tenderness, constipation, an unquantified weight loss, fecaloid transvaginal discharge and hyperthermia. An IV contrasted CAT scan was performed, showing changes in the peri-rectal fat and free fluid. A laparotomy was done, during which free inflammatory fluid, and a mass that involved rectum and cervix were found. A hysterectomy was done, which included the upper two thirds of the vagina. A transoperative biopsy reported vaginal sarcoma, thus leading to the decision of escalating the procedure to an abdomino-perineal resection. Due to the low incidence and lack of information regarding this rare tumor (less than 150 cases have been reported in the last 50 years), we decided to add this new case to the literature.

RESUMEN

El cáncer de vagina es la tumoración ginecológica menos común: representa tan sólo 2% de los tipos de cáncer. El tipo histológico más común es el escamoso (75-90%), y el menos frecuente, el sarcoma (< 3%). Presentamos el caso clínico de una paciente de 40 años que acudió por aumento de volumen en el mesogastrio, acompañado de dolor tipo cólico intenso, pérdida de peso no cuantificada, leucorrea fecaloide e hipertermia. Se solicitó una tomografía axial computarizada con contraste intravenoso, que reportó cambios grasos perirrectales y abundante líquido libre. Se realizó una laparotomía exploradora, en la que se encontró líquido libre inflamatorio hacia el hueco pélvico, así como una masa que involucraba el recto y el cérvix. Se realizó la resección de la vagina hasta el tercio distal e histerectomía total, que se envió a biopsia transoperatoria. Ésta reportó sarcoma de la vagina, por lo que se llevó a cabo una resección abdominoperineal. Debido a la poca incidencia de los sarcomas de vagina (existen menos de 150 casos reportados en los últimos 50 años), no hay consenso con respecto al mejor tratamiento, por ello decidimos añadir un caso más a la literatura existente.

Abbreviations:
CT = Computerized axial tomography scan.
HR = Heart rate.
BP = Blood pressure.
PhE = Physical examination.
Hb = Hemoglobine.

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INTRODUCTION

Vaginal cancer is the least common, it represents only 2% of the gynecological tumors. No more than four thousand cases are reported per year worldwide.[1,2] Eighty-four percent are usually secondary carcinomas; of these, 32% comes from the cervix, 18% from the endometrium, 9% from the colon and rectum, and 6% from the ovary.[3] Of the primary tumors, the most common histological type is the squamous (75-90%), followed by adenocarcinoma (5-10%), melanoma (3%), and sarcoma (< 3%), of various histological varieties, such as endometrial stroma, angiosarcoma, adenosarcoma, and leiomyosarcoma.[4]

Leiomyosarcoma of the vagina represents the 0.62% of malignant neoplasms of the female reproductive tract.[3] Morphological types mentioned are myxoid, epithelioid, and conventional. It is

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characterized by cellular atypia, with more than 5 to 10 mitoses per field, areas of necrosis and a diameter greater than three centimeters, which is also a prognostic factor for recurrence.[5] It is usually spread by local and hematogenous invasion, having a lung metastatic predominance.

According to its incidence, the average age of diagnosis prevails in young women, with an age range from 18 to 86 years and a mean of 50 years.[1] They tend to vary according to race; African-Americans are more likely, with a risk twice as high as in the Caucasian population.[6]

Its clinical presentation is variable and ambiguous; it usually presents as an asymptomatic vaginal mass; it may or may not be accompanied by leucorrhoea, transvaginal bleeding, intermittent pain and, in advanced stages, weight loss.[7] On physical examination, they tend to be towards the posterior vaginal septum, unlike leiomyomas, which usually appear in the anterior portion; in lower tumor locations they may resemble a Bartholin's cyst.[8] There is no specific imaging method; however, on MRI they usually appear as a heterogeneous, hyperdense mass, with irregular reinforcement during the T1 phase after administration of the contrast medium.[9] However, there are other malignancies that could resemble such a finding, which are detailed as differential diagnosis *(Table 1)*.[10]

Table 1: Differential diagnoses.

Vaginal masses	Gartner's cyst
	Granuloma
	Epithelial inclusion cyst
	Neurofibroma
	Rhabdomyoma
	Capillary hemangioma
Malignant primary tumors	Squamous epithelial carcinoma
	Adenocarcinoma
	Rhabdomyosarcoma
	Melanoma
	Small cell carcinoma
	Mixed Müllerian sarcoma
	Trophoblastic gestational tumor
	Endometrial cancer
	Metastatic

Taken from: Mills AM, Longacre TA. *Smooth muscle tumors of the female genital tract*. Surg Pathol Clin. 2009; 2: 625-677.

Regarding treatment, there is little consensus on which is the best therapy; however, surgery is considered fundamental.[1,2,4,11] In the discussion we detail various approach options.

The object of this paper is to document one of the first cases of sarcoma of the vagina invasive to the rectum reported in the Mexican literature, as well as a brief review of the literature concerning management and its outcome.

PRESENTATION OF THE CASE

A 40-year-old female patient with no significant family history and chronic degenerative gynecological history; menarche at age 14, three gestations, three Csections. Date of last menstrual period: October 2015. Last cytological analysis, normal. Complaint started three months earlier with an increased volume of the mesogastrium, accompanied by intermittent colic pain of mild intensity, unquantified weight loss and alteration of bowel movement habits. It progressed seven days prior to admission, with an increase in pain intensity, transvaginal bleeding, fecaloid leucorrhea and unquantified hyperthermia, for which she requested clinical evaluation. Upon arrival, HR was 110 beats per minute, BP 100/70 mmHg, RR: 18 breaths per minute, temperature 38.5 ºC. On physical examination she was found well oriented, cooperative, with pain facies; head, neck, and chest with not significant findings, a globose abdomen, with no peristalsis; muscular rigidity and tenderness in the mesogastrium, hypogastrium, and left iliac fossa. A plastron was palpated on the left iliac fossa and the mesogastrium. Laboratory data showed a hemoglobin of 11.8 g/dl, of 20,000 leukocytes/mm^3, 95% of neutrophils, 150,000 platelets/μl. CT showed inflammatory changes in the perirectal fat and free fluid in the posterior cul-de-sac. With these signs of abdominal sepsis, the protocol for an exploratory laparotomy was completed. A supra-infraumbilical incision of 12 centimeters was made; planes dissected to reach the

abdominal cavity; free inflammatory fluid was observed towards the pelvic cavity (100 cm^3) —not sent for cytochemical or cytological study— as well as a mass involving the rectum and cervix *(Figure 1)*. A circumferential dissection of the tumor was started, until delimited; it was evidenced that it came from the posterior septum of the vagina, with extension towards the rectum. It was decided to perform a resection of the vagina down to the distal third and radical hysterectomy. The piece was sent for an intraoperative biopsy, which reported a vaginal sarcoma. An abdominoperineal resection was then decided, with a terminal colostomy of the descending colon *(Figure 2)*. A closed Blake 19 Fr drainage was placed,

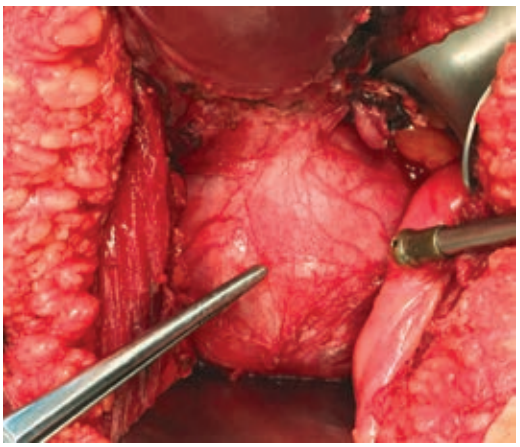

Figure 1: *Tumor mass dependent on the cervix, with invasion of the rectum.*

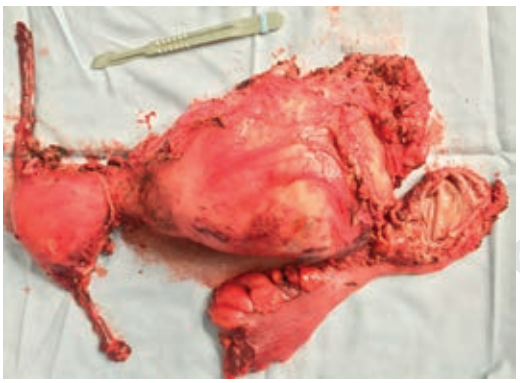

Figure 2: *En bloc resection: radical hysterectomy, vaginectomy down to the distal third, plus rectosigmoidectomy.*

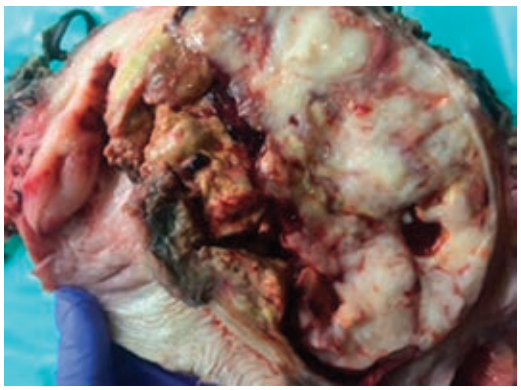

Figure 3: *Tumor mass, macroscopic section.*

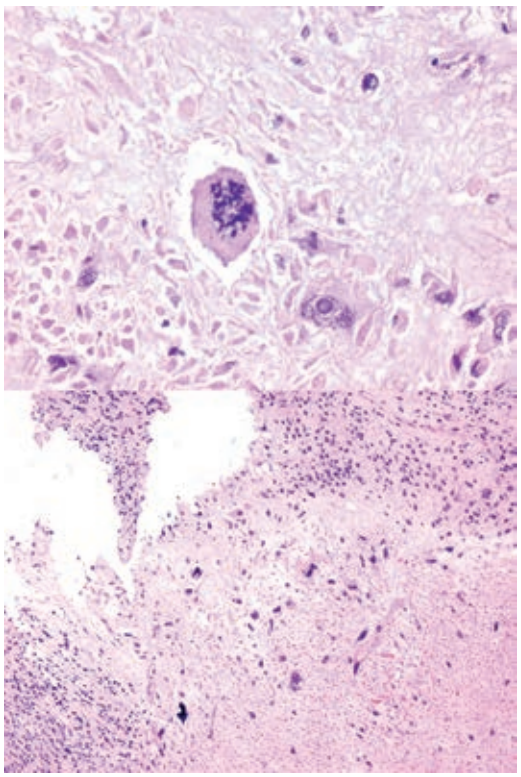

Figure 4: *Tumor microphotograph stained with hematoxylin and eosin (H and E), which demonstrates grade III nuclear atypia, necrosis, and multiple atypical mitoses.*

fixed to the abdominal wall with 2-0 Prolene® (polypropylene) and closure done started by planes: fascia with a continuous suture with 0 Monocryl® (polydioxanone), the subcutaneous cellular tissue with 2-0 Vicryl® (polyglactin 910) simple inverted points, and the skin with

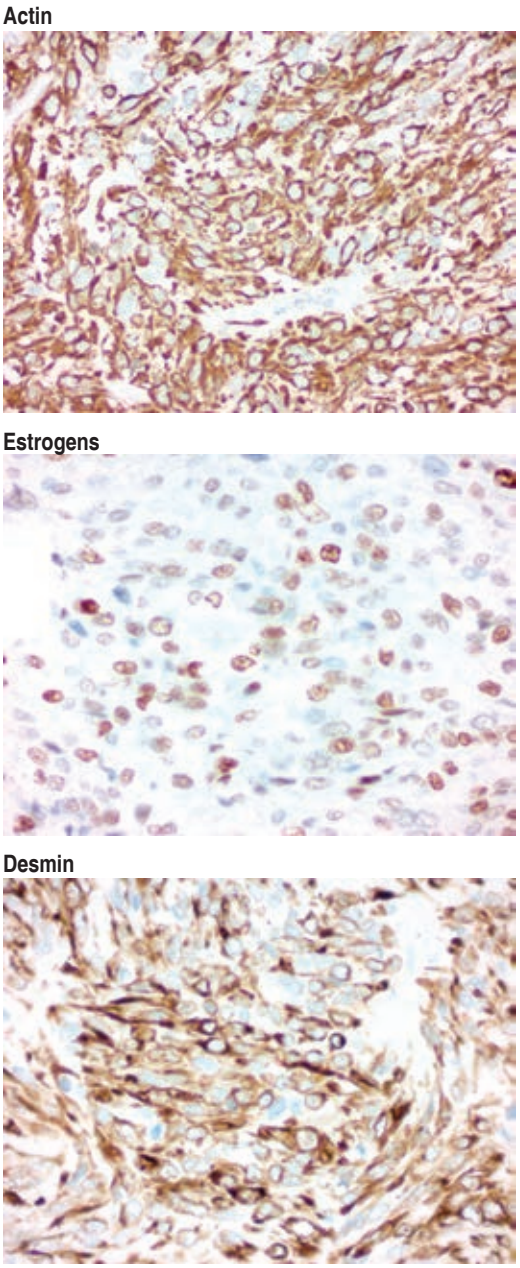

Figure 5: *Positive immunohistochemistry.*

Prolene® 3-0 simple stitches, to finish the surgical procedure. The patient was admitted to intermediate therapy, she had a good clinical evolution. Her transfer to the general ward was decided on the fourth day, and he was discharged on the tenth day, without drainage. The pathology report which reported macroscopically a high-grade leiomyosarcoma of 16 cm, with a free surgical distal limit of two centimeters *(Figure 3)*. Microscopically, a grade III nuclear atypia, necrosis and multiple atypical mitoses *(Figure 4)*, as well as positive immunohistochemistry for muscle-specific actin, estrogens, desmin and Ki-67 *(Figure 5)*. During her post-surgical follow-up, she reported no symptoms, and a functional colostomy. She was readmitted three months after surgery for the placement of a permanent subcutaneous central venous reservoir (Port-a-Cath®) for chemotherapy and a first surgical time of vaginoplasty, performed without complications; however, the patient decided not to attend chemotherapy sessions. She was readmitted 11 months after the first surgical procedure with fecaluria, pelvic recurrence, respiratory distress secondary to lung metastasis, and general deterioration. She was kept in the hospital for 48 hours with conservative treatment. Her relatives then decided to transfer her home for palliative treatment.

DISCUSSION

Leiomyosarcoma of the vagina is the most common vaginal sarcoma; however, only 150 cases have been reported in the world literature in the last 40 years; therefore, there is no well-established treatment. According to Kohsla and his collaborators, the primary treatment lies in complete resection with free margins. Resection with 1 to 2 cm margins is recommended, and in high-grade tumors, greater than 3 cm. Peters et al. demonstrated in their study that patients treated with pelvic exenteration had a better long-term prognosis, so they recommend this procedure as the first choice. In the case series of Ciaravino et al. 66 patients with leiomyosarcoma where they reported, they determined a 5-year survival of 43%; the main predictor of survival was the degree of tumor differentiation, as well as the clinical stage. Stage I has a five-year survival of 55%; stage II, 44%, and stages III and IV, 25% to 18 months. Age, likewise, affects the prognostic factor, since patients younger than 40 years have a 5-year survival of 51%, while those older than 50 years have a survival of 26%.

The use of adjuvant with chemotherapy and radiotherapy remains an obscure terrain,

unlike the case of uterine sarcoma, where there is more information. Various studies have postulated that adjuvant radiation therapy should be considered in high-grade sarcomas, recurrent sarcomas, and resections with positive margins; however, in a series of 17 cases (10 of which were leiomyosarcomas), 35% received radiotherapy; with no difference between those who did not receive radiotherapy. As far as chemotherapy is concerned, equally controversial; no specific guide exists as for the most suitable drugs. It has been used primarily to combat a high risk of systemic recurrence. In the latest meta-analysis by Pervaiz et al. where 18 cohort studies of 1,953 patients were analyzed, it is mentioned that a combination of ifosfamide and adriamycin reduces mortality from 41 to 30%. They recommend this scheme in patients with lowgrade tumors, without dissemination.

In the Ciaravino study, patients treated with adjuvant chemo/radiation therapy survived more than two years; however, those who received primary surgical treatment had a survival rate of 57% at five years. According to the guidelines of Mastrangelo and his collaborators, the surgical management of sarcomas should include a resection margin of 2 cm; however, it has not been uniformly established. They recommend that for high-grade injuries, the margin should exceed 3 cm; likewise, they mention that treatment without adjuvant is acceptable for tumors smaller than 3 cm and superficial tumors with a low degree of differentiation.

There is no set time for follow-up; however, it is recommended to do it every three months during the first three years and every six months in the subsequent years until the fifth post-surgical year. A stable or controlled disease is defined as one where absence of metastasis or residual tumor can be documented.

CONCLUSIONS

Vaginal sarcoma is a rare entity. We lack protocols that determine the best therapy to prolong the survival of our patients; however, we must consider surgical resection as fundamental and individualize each patient to receive or not adjuvant chemotherapy and/or radiotherapy.

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