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Ovarian leiomyosarcoma: A case report

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Abstract

Introduction: Ovarian leiomyosarcomas are extremely rare neoplasms and are part of the subgroup of smooth muscle tumors that constitute less than 1% of ovarian tumors; within it are classified as carcinosarcoma, angiosarcoma, fibrosarcoma, and leiomyosarcoma. According to the reviewed literature, 63 cases of leiomyosarcoma have been reported to date.

Clinical Case: We present the case of a 53-year-old female patient. She was admitted to the emergency area due to severe abdominal pain accompanied by abdominal distension and orthopnea. Physical examination revealed a large, slightly mobile mass. The tomography showed the presence of an abdominopelvic mass that extended between the intestinal loop and the root of the mesentery and compressed the bladder, uterus, and ovaries. She underwent exeresis hysterectomy with right salpingo -oophorectomy and left oophorectomy through exploratory laparotomy. Histopathological studies yielded the results of a malignant neoplasm compatible with leiomyosarcoma; she was treated with surgery and adjuvant chemotherapy.

Keywords:
MESH: ovarian leiomyosarcoma, malignant neoplasm; ovarian neoplasm; case reports.

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Introduction

Ovarian leiomyosarcoma is a rare oncological pathology, with a prevalence of less than 1% of all malignant ovarian tumors. Carcinosarcoma, angiosarcoma, fibrosarcoma, and leiomyosarcoma are classified within it. According to the literature reviewed, 63 cases of leiomyosarcoma have been reported to date, all published in the international literature [1-2].

Ovarian leiomyosarcoma is seriously related to uterine leiomyomatosis; its origin is likely from the smooth muscle of the vascular walls (ovarian vein, ovarian ligaments, smooth muscle of the follicles, corpus luteum, ovarian totipotent stem cells) [3-4].

It generally occurs in postmenopausal patients and is unilateral; however, some exceptions can occur in young women and bilaterally [1-3-5].

In most cases, the behavior is aggressive and has a poor prognosis for life within two years after starting specific treatment.

This review is of significant relevance, given the scarcity of cases reported in the country's literature, intending to provide valuable data for the epidemiological characterization and behavior of this oncological disease.

Clinical case

This case corresponds to a 53-year-old female patient, postmenopausal, with no significant pathological history. She referred to six pregnancies as gynecological-obstetric antecedents, of which three were deliveries, three were abortions, three were living children, and the date of her last menstruation was 48 years. She was hospitalized for abdominal-pelvic pain one month before the emergency assessment at this institution. The pain was intense 8/10 on the VAS scale, undefined type, accompanied by diaphoresis and paleness. The possibility of a diagnosis of uterine myoma was raised, and the evolution of the clinical case is indicated in Figure 1.

The patient went to the emergency service due to severe abdominal pain at the level of the hypogastrium of 6/10 according to the VAS Scale, accompanied by abdominal distension, diaphoresis, and generalized paleness. Physical examination revealed a distended abdomen with dullness; A round, slightly mobile mass was palpable that occupied the four quadrants.

Diagnostic workshop

In the extension tests, he presented leukocytosis at the expense of neutrophils (13,520 u/µL), normocytic, normochromic anemia (HB 11 g/dL, Htco 35%), elevated lactic dehydrogenase (1077 IU/L) and tumor markers (alpha-fetoprotein, carcinoembryonic antigen, and Ca-125) within normal parameters. Figure 1

The tomography determined that the mass depended on the left ovary and had a diameter of 44x25x22 cm. An exploratory laparotomy was performed on the hospitalized patient seven days after the assessment by the oncology surgery service; a large tumor that covered the abdomen with a smooth surface and adhered to the sigmoid colon from the left ovary was found.

Through digitoclasy and blunt dissection, its release and complete exeresis are achieved. Additionally, an ascitic fluid sample was taken, revealing the presence of malignant cells. It was complemented with cytoreductive surgery with hysterectomy; no complications were recorded during the procedure. Figure 3.

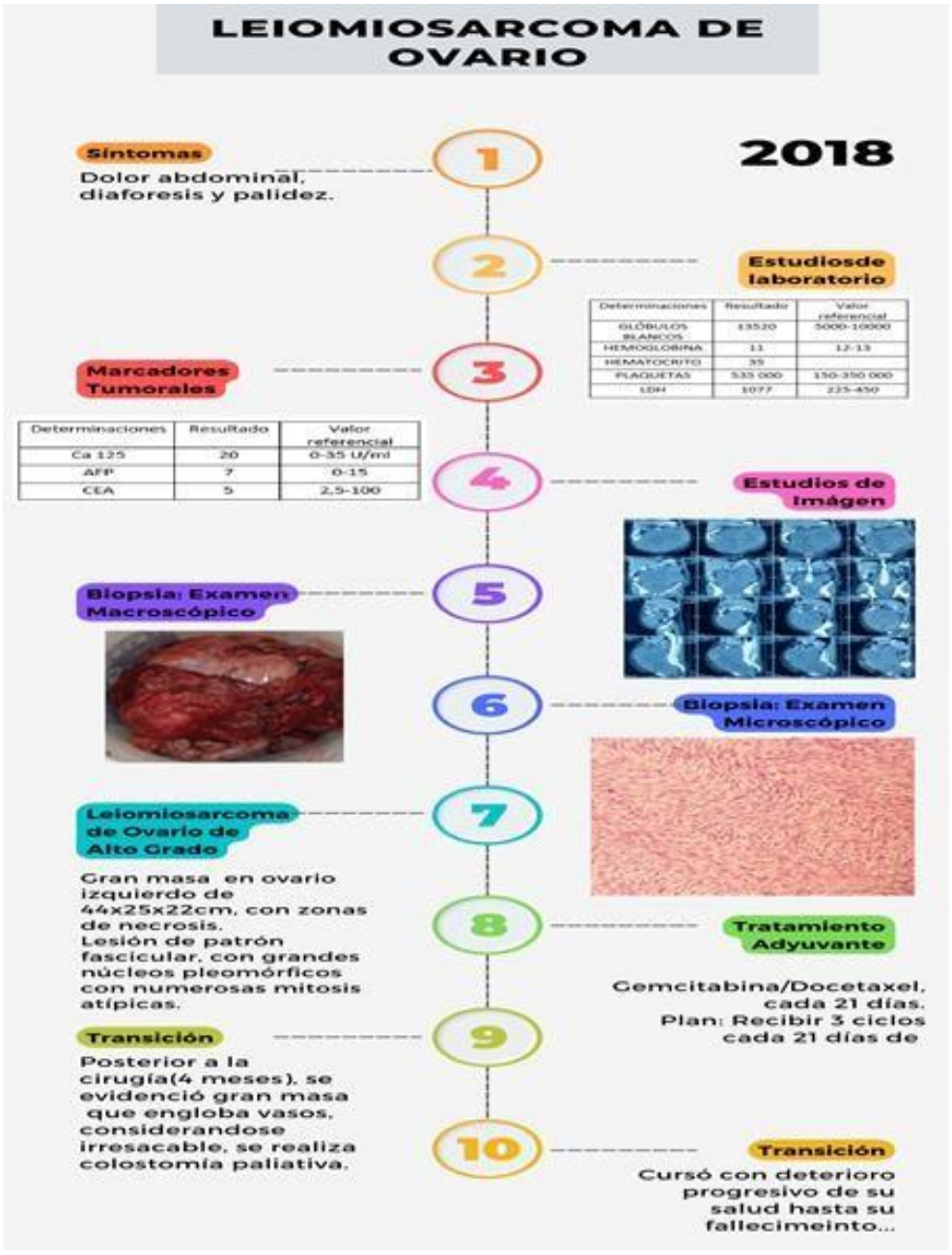

Figure 1. Timeline of the evolution of the case, according to the CARE guidelines.

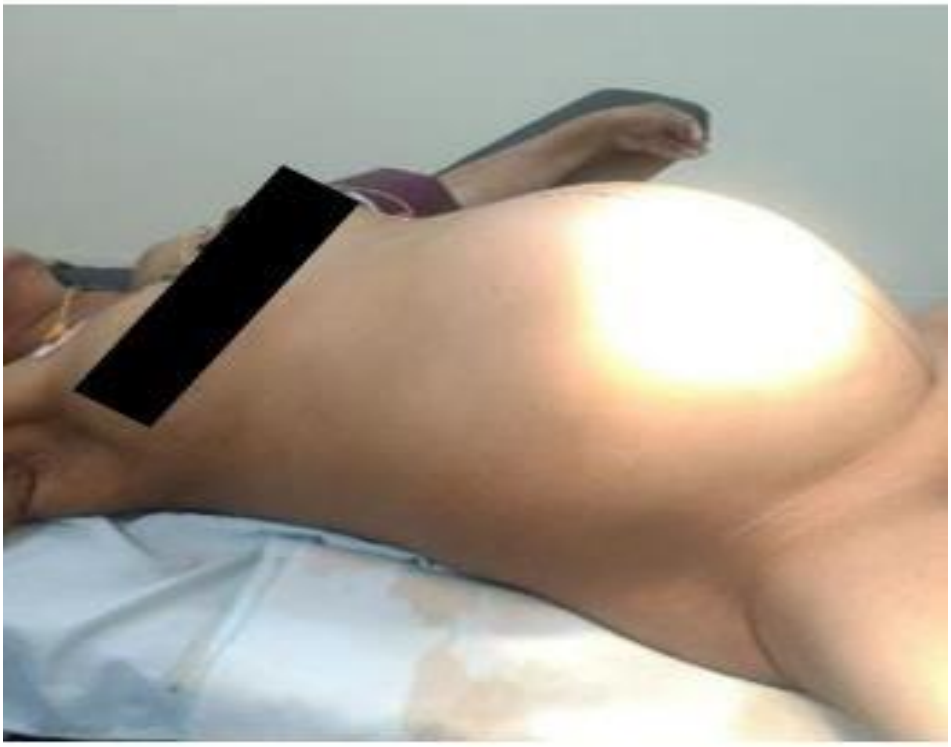

Figure 2. Patient with a distended abdomen due to the presence of a tumor mass

Among the macroscopic characteristics, a left ovary of 8,500 gr, 44x25x22 cm was found; partially encapsulated, solid throughout; white with areas of necrosis in approximately 30% and areas of hemorrhage.

Microscopically, the lesion presented a fascicular pattern with large, pleomorphic, hyperchromatic nuclei, numerous atypical mitoses, and extensive necrosis and hemorrhage areas. Ascitic fluid cytology showed abundant cells with loss of the nucleus/cytoplasm relationship, large, hyperchromatic, ovoid nuclei, histiocytes, and lymphocytes against a hemorrhagic background, being positive for malignant neoplastic cells.

After the surgical intervention, the gemcitabine/docetaxel scheme was scheduled as an oncospecific adjuvant treatment every 21 days, receiving three cycles. Four months after surgery, the patient went to the emergency service, reporting diffuse abdominal pain of 8/10 according to the VAS scale, abdominal distension, and constipation; in the tomographic study, a retroperitoneal mass was identified, which encompassed large vessels, so it was considered unresectable, and palliative loop colostomy was performed. During the second intervention, there were no complications either; however, he progressed with progressive deterioration until his death.

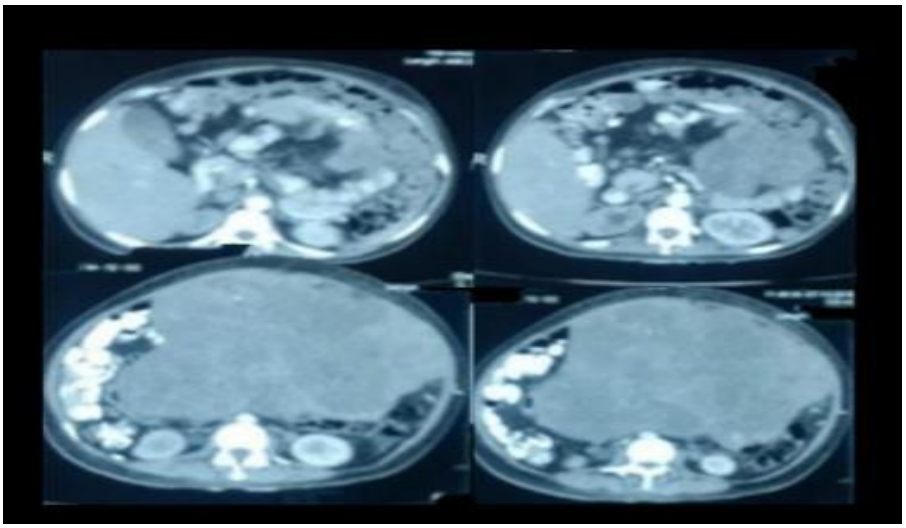

Figure 3. Coronal computed tomography of the abdomen shows a mass of 44x25x22 cm.

Discussion

Ovarian leiomyosarcoma is a rare oncological pathology, with a prevalence of less than 1% of all malignant ovarian tumors; carcinosarcoma, angiosarcoma, fibrosarcoma, and leiomyosarcoma are classified, although only 63 cases have been reported worldwide to date [1 - 2 - 6 .] Regarding its origin, it is presumed that it comes from the smooth muscle of the vascular walls, such as the ovarian vein, ovarian ligaments, smooth muscle of the follicles, corpus luteum, totipotent stem cells of the ovary, remains of the Wolffian duct and smooth muscle metaplasia of a focus of ovarian endometriosis from cells of a teratoma that has differentiated; even according to Taskin et al. [7-8], they mention an etiological possibility concerning radiotherapy and previous neoplasia since there are 2 cases of 12-year-old girls with a history of medulloblastoma [1-8-9]. Zygouris calculated the mean age of presentation, which was 52.6 years; on the other hand, He et al. reported that the average age was 54.9 years, which is the age data that is fulfilled in the patient, since the same age was 53 years old [10 - 11].

Most studies express no significant elevation of tumor markers, as occurs in the patient in question; however, some studies manifest the presence of estrogen and progesterone receptors [1-12-13]. Clinically, it is characterized by abdominal pain, metrorrhagia, tenesmus, and urinary retention, as long as they are large masses, similar to our patient; however, when the tumor is of moderate or small size, they are asymptomatic and are even identified incidentally; it is an entity of rapid and progressive growth, they are more likely to occur in postmenopausal women, and unilaterally, however, there is evidence of having presented in young women and bilaterally; it is also seriously related to a history of uterine leiomyomatosis [2-8].

Macroscopically, they are characterized as tumors of variable size and pale and lobulated surfaces, which when fragmented present solid areas with foci of necrosis and hemorrhage; microscopically, they are indistinguishable from their counterpart, uterine leiomyosarcoma , for which the same histopathological diagnostic criteria are established; according to Lerwill

[12-14] to differentiate between a malignant leiomyoma and a benign leiomyoma, it is based on the presence of 5 mitoses per 10 high-power fields of cytonuclear atypia [1-13].

Ovarian leiomyosarcoma (LMO) is made up of intertwined bundles of fusiform cellularity that variably express desmin, vimentin, and smooth muscle actin; the same conditions will prove positive in distinguishing between the verses as mentioned above fibrosarcomas, rhabdomyosarcoma, thecomas, and extradigestive stromal tumors [1–15].

Leiomyosarcoma is considered a neoplasm with an inferior prognosis unfavorable for the life of the human being. Monk et al., in their review study [16], stated that most of the cases had a recurrence within a year and that they also died within two years after the initial treatment, similar to what happened in this case, so much so much so that in the study proposed by Mayerhofer, who studied predictive factors of tumor aggressiveness, reported that matrix metalloproteinases MMP 1 and MMP 2, angiogenic factors (VEGF) are positive in the pathological entity; however, since they are not routine paraclinical tests, there are no results of these in the case [17].

Management is strictly based on surgical resection accompanied by adjuvant treatment . Since there is a high rate of probability of occult metastatic disease, according to little scientific evidence, it is recommended to continue with 4-6 cycles of chemotherapy based on the DOXORROBUCIN (ADRIAMYCIN)/GEMCITABINE scheme; in the present case, it was used of the GEMCITABINE/DOCETAXEL scheme for three cycles [1 - 2].

We can conclude that we are dealing with an LMO, a rare tumor that is difficult to diagnose and aggressive in behavior, where the pathologist continues to be the protagonist in the diagnosis of tumors. Radical surgical treatment is recommended through a radical cytoreductive hysterectomy. To date, there are no studies that assess the effectiveness of any therapy. Due to the few cases studied, the benefits of chemotherapy and radiotherapy have not been established, thus being a wide and open field for future research.

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Abbreviations

LM O: Ovarian leiomyosarcoma.

Administrative information

Additional Files

The authors declare none.

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10. Writing - revision and editing: Blas Luciano Mendez Orellana, Bryan Ariel Valarezo

RomeroAll authors read and approved the final version of the manuscript.

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Availability of data and materials

Data availability is available upon request to the corresponding author. No other materials were reported.

Statements

Ethics committee approval

It does not apply to observational studies with a review of databases or medical records.

Consent to publication

The authors have the corresponding publication permission for this clinical case.

Conflicts of interest

The authors declare that they have no conflict of interest or competence.

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