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Case Report

Giant leiomyosarcoma of the anterior abdominal wall: case report and review of the literature

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ABSTRACT

Smooth muscle tumors are rare tumors with a low worldwide incidence, which is a diagnostic and therapeutic challenge for doctors, within these, leiomyosarcomas are tumors that occur more frequently in the uterus or abdominal cavity, however, another presentation called leiomyosarcomas peripheral which are subdivided into cutaneous or subcutaneous which tend to appear in the extremities of the lower limbs. The presentation of a clinical case of a patient of the eighth decade of life who presents a giant tumor of the ulcerative abdominal wall and hemorrhage that conditions anemia syndrome and hemorrhagic shock is made, for which an emergency surgical resection is performed. The treatment of leiomyosarcoma consists of surgical removal, however the margins are not clearly defined and regularly require a multidisciplinary team that includes an oncological surgeon and a plastic and reconstructive surgeon. In advanced cases and selected patients, medical treatment with chemotherapy can be granted.

Keywords: Leiomyosarcoma, Surgery, Abdominal wall

INTRODUCTION

Leiomyosarcomas are rare and rare mesenchymal neoplasms. Aggressive, arising from smooth muscles. They represent 3 to 7% of the soft tissue sarcomas, incidence rates have been estimated at 0.2/100,000/year, which is an added difficulty in diagnosis and treatment. It is the third most common cutaneous sarcoma, after dermatofibrosarcoma and spinal sarcoma. from kaposi.[2] Leiomyosarcomas are divided into those involving deep soft tissue sites such as the retroperitoneum and those that involve peripheral soft tissue sites. tissue leomyosarcomas peripheral soft leiomyosarcomas, also called superficial leiomyosarcomas, are divided into cutaneous or dermal leiomyosarcomas originating from muscles hair follicle erectors and on the other hand leiomyosarcomas subcutaneous where its origin is from the smooth muscle of the arteries or veins, the different origins of these two entities they confer a different clinical appearance and prognosis.[1,2]

Subcutaneous leiomyosarcomas are tumors rare accounting for 1% to 2% of all malignancies superficial soft tissue usually occurs in patients between 50 and 70 years, with a male predominance that varies from 2:1 to 3:1 being the factors Most common predisposing factors reported were trauma and radiation.

Because subcutaneous leiomyosarcomas tend to be more aggressive tumors and are usually diagnosed at a more advanced stage, they are usually larger than cutaneous leiomyosarcomas.[2]

CASE REPORT

This is a 77-year-old male patient with a history of benign prostatic hyperplasia and epilepsy. Surgical history with exploratory laparotomy secondary to blunt abdominal trauma where intestinal resection and splenectomy were performed 17 years ago, presenting as a late complication an incisional hernia in the midline without surgical management (Figure 1).

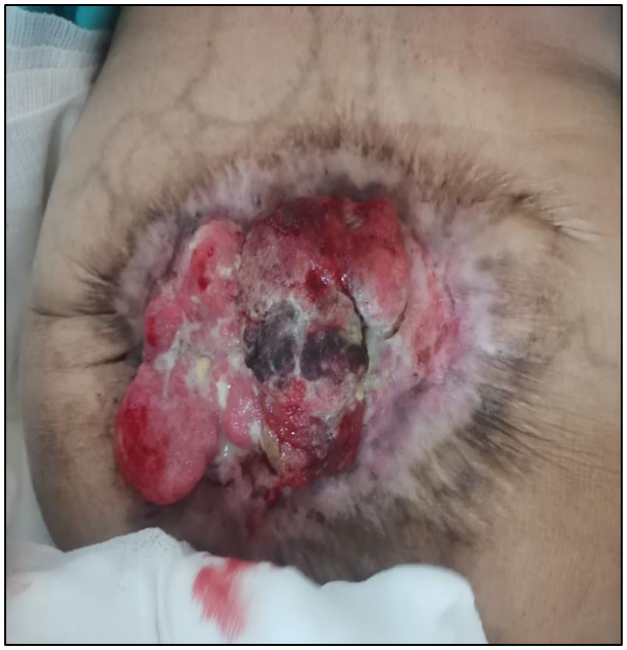

Figure 1: Abdominal wall tumor.

His condition began in October 2021 with the appearance of a progressively growing polypoid lesion in the mesogastrium, a study protocol was initiated with an incisional biopsy with a report of malignant neoplasm with an infiltrating epithelial appearance with focal tumor necrosis, suggesting immunohistochemical studies with differential diagnoses urothelial carcinoma , acinar adenocarcinoma of the prostate, gastrointestinal stromal tumor and abdominal tomography where no communication to the abdominal cavity or other lesions suggestive of tumor activity are observed, in addition a colonoscopy is performed ruling out intraluminal tumor. It is referred to an oncology hospital to continue the diagnostic protocol and therapeutic, however, the patient goes to the emergency department of a tertiary hospital due to hemorrhage due to a tumor accompanied by asthenia, adynamia.

On admission with the presence of a tumor in the anterior wall of the abdomen on the midline of approximately 18x20 cm, cerebroid appearance with discharge of fetid and hematic purulent material with areas of necrosis. Computed tomography was performed with a report of an ovoid morphology tumor in the anterior wall of the abdomen, lobulated edges, defined with a dorsoventral growth vector, exophytic, heterogeneous density of 37 HU, predominantly hypodense with peripheral areas isodense to air upon administration of medium. contrast, shows heterogeneous nodular enhancement 92 HU, stomach with normal morphology and situation, small intestine at the level of the ileum, diffuse thickening of the mucosa loses interface with mesenteric fat and with the anterior wall of the abdomen, mesentery and peritoneum with little free fluid in space anterior subhepatic, aortic and para-caval lymph node growths with inflammatory characteristics.

An emergency surgical intervention was performed with a wide resection of the anterior abdominal wall tumor + intestinal resection with an entire lateral lateral mechanical anastomosis+wall plasty with component separation technique + application of a split-thickness skin graft with the findings of an aspect tumor. Cerebroid, friable, prone to bleeding approximately 15x15x8 cm, with vascularity increased in its periphery, tumor infiltration into the loop of the small intestine 200 cm from the fixed loop and 20 cm from the ileocecal valve, resecting approximately 20 cm with mechanical side-to-side anastomosis with total bleeding of 1800 cc (Figure 2 and 3).

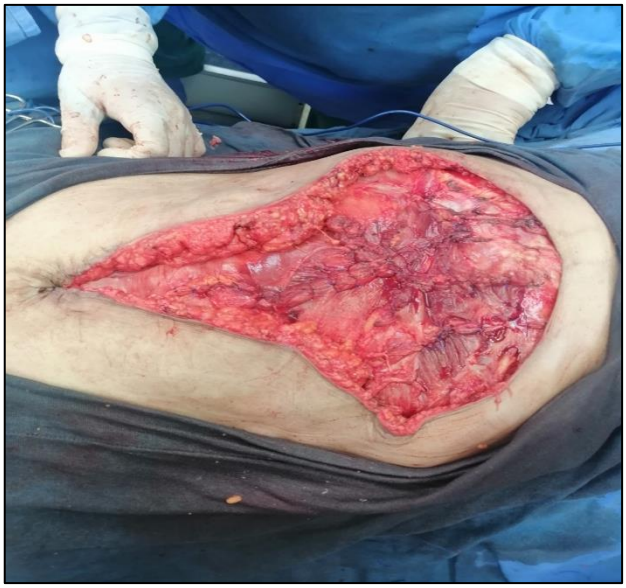

Figure 2: Tumor resection.

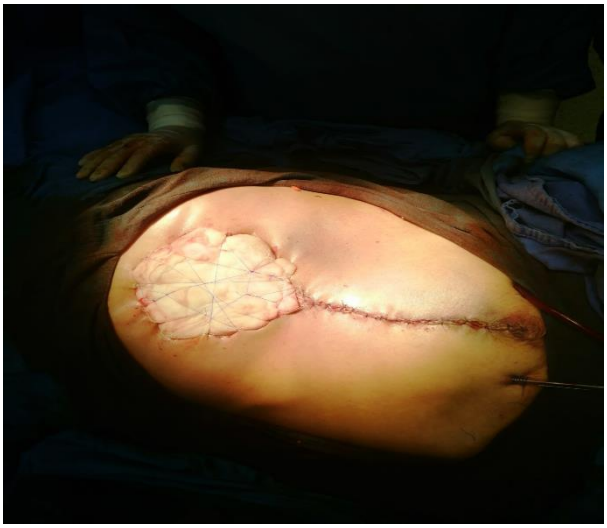

Figure 3: Abdominal wall reconstruction.

Surgical sample sent to cm pathology service with histopathological diagnostic report of high-grade leiomyosarcoma, conventional type and ulcerated and abscessed pleomorphic, histological grade 3, mitotic

index of 3 mitoses in 10 high-power fields, 20% necrosis, lymphovascular invasion unidentified, surgical margins with deep edge with infiltration to muscles of the pars and soft tissues, distant metastasis undetermined, (Figure 4-7). Pathological stage pT2, pNX, pMX, with positive immunohistochemical results for H CALDOSMON, AML; negative for SOX 10, DESMIN, k167 10%. Patient is discharged from the operating room with hemorrhagic shock and later dies.

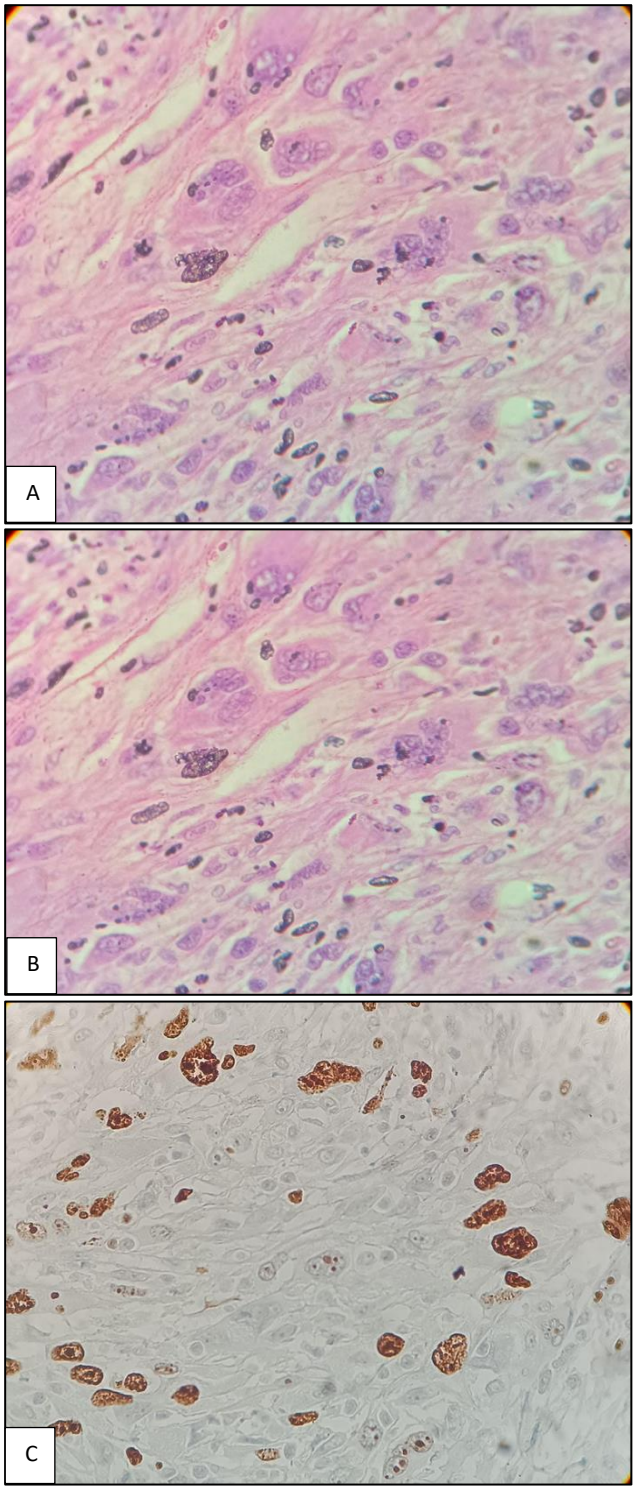

Figure 4 (A-C): Histopatology.

DISCUSSION

There is little bibliographic information on this pathology being the largest studies two retrospective studies in electronic file reviews including 200 patients in 20 years taking into account all those classified as cutaneous leiomyosarcoma, however, in both the clinical and prognostic importance of the adequate differentiation of subcutaneous leiomyosarcomas is concluded due to their greater morbidity and mortality

Neelam et al in their analysis of the database SEER of 40 years. identified 208 patients diagnosed with leiomyosarcoma with important demographics like a median age of 62 years, with a marked predisposition for the male sex, with the location more frequent in the lower limbs and hip with a size greater than 3 cm which coincides with that published in smaller series of clinical cases.[3]

Subcutaneous leiomyosarcomas derive from smooth muscle fibers of the middle layer of the vascular wall of arteries and veins. Unlike the superficial or dermal variant, they are characterized by a high rate of locoregional recurrence (37%) and distant metastases (21-62%), so they are considered to have a less favorable prognosis.[2,3]

The etiology of these tumors is relatively unknown, although the predisposing factors for leiomyosarcoma are the precancer and previous traumatic injury or radiation exposure to the site of onset of leiomyosarcoma. Cutaneous leiomyosarcomas are usually asymptomatic, but patients complain of pain, itching, burning sensation and bleeding, with pain being the most common symptom in 80% to 95% of patients, however the patient presented was asymptomatic and presented a large tumor with data of active bleeding from ulcerations.[4]

As the clinical presentation can be nonspecific, diagnosis is based on histologic findings and examination immunohistochemical. It is usually done on a skin biopsy being the analysis of the surgical removal the definitive diagnosis.[5]

Histological examination of cutaneous leiomyosarcoma usually reveals a poorly demarcated lesion that occupies the entire thickness of the dermis and sometimes extends into the subcutaneous tissue, the cells are spindle-shaped and have elongated nuclei with blunt ends, a nondescript nucleolus and eosinophilic fibrillar cytoplasm.[6]

Several variants have been described histopathology of leiomyosarcoma, including leiomyosarcoma epithelioid, giant cell leiomyosarcoma multinucleated, granular cell leiomyosarcoma, and sclerotic leiomyosarcoma. Considerable desmoplasia has also been described that can complicate the diagnosis and reporting of myxoid and pleomorphic variants of subcutaneous leiomyosarcoma.[6]

Before making a definitive diagnosis of a primary cutaneous leiomyosarcoma, the possibility of a metastatic cutaneous leiomyosarcoma of a deep tissue or visceral leiomyosarcoma must be ruled out, given its clinical manifestation, epidermal cysts and cutaneous metastases are important differential diagnoses.[6]

The immunohistochemical study is a essential tool for the proper differential diagnosis between leiomyosarcoma and other spindle cell tumors with similar histological characteristics. Well -differentiated leiomyosarcoma is positive for vimentin, desmin, h-caldesmone , muscle-specific actin, alpha-smooth muscle actin, and muscle myosin. smooth in more undifferentiated lesions and subcutaneous leiomyosarcomas, desmin is frequently negative.[6]

In cases of diagnostic uncertainty, other immunohistochemical markers should be used for confirmation; at least two positive muscle markers are recommended to confirm the diagnosis.[6]

Complete surgical excision is the treatment of choice for cutaneous leiomyosarcoma. The classic recommendations suggested aggressive surgeries with margins of 3 to 5 cm, the deep margin should reach the fascia, and in the most infiltrative cases, the muscle should be included.[7] Devene et al standardized margins for patients with diagnosis of leiomyosarcoma in a period of 5 years of 1-2 cm obtaining only 3% of recurrences, however only patients were considered in the study with cutaneous leiomyosarcoma.[7,8]

In unresectable cases, radiation with or without chemotherapy is recommended, although success rates are mixed. Radiotherapy can also be used in local palliative control in cases with metastases.[9,10]

Regarding prognostic factors, there are few studies in which only the size of the tumor was shown to be an independent prognostic factor in relation to decreased survival. Factors such as a tumor size equal to or greater than 5 cm, deep location with involvement of the fascia, and a high histological grade were correlated with lower survival.[10]

CONCLUSION

Leiomyosarcoma -type soft tissue tumor in an unusual location with very few case reports in the literature, which results in a difficult diagnosis in hospital centers that do not have an oncology and pathology system experienced in this area. type of tumors. The definitive treatment is the surgical removal of the tumor. However, the best management of giant tumors that compromise a large abdominal surface and intra-abdominal organs has not been determined, which often requires the intervention of a multidisciplinary surgical team. It is vitally important to continue reporting leiomyosarcoma cases in order to eventually create an international consensus for the management of this pathology.

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