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SARCOMAS SCOPING REVIEW

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SUMMARY

Introduction: *Soft tissue sarcomas are a heterogeneous and relatively uncommon group of tumors that arise from mesenchymal tissues and form almost anywhere in the body. The mainstay of treatment is surgery; wide excision for low-grade lesions and wide or radical (compartmental) resection for high-grade tumors. Adjuvant preoperative and/or postoperative radiotherapy improves the likelihood of local control and maintains function when adequate margins cannot be achieved.*
Objective: *to detail current information related to sarcomas, description, etiology, epidemiology, presentation, evaluation and treatment.*
Methodology: *a total of 47 articles were analyzed in this review, including review and original articles, as well as clinical cases, of which 32 bibliographies were used because the other articles were not relevant to this study. The sources of information were PubMed, Google Scholar and Cochrane; the terms used to search for information in Spanish, Portuguese and English were: sarcomas, soft tissue tumor, oncologic surgery, cancer, osteosarcomas, brachytherapy, radiotherapy.*
Results: *The rate of progression and the probability of hematogenous spread, usually to the lung, are determined mostly by the grade of the tumor. The probability of regional spread is low. Prognosis is determined by multiple factors, such as: age, tumor size, histologic grade, depth, histologic subtype and site. About 90% and 98% of recurrences are seen between 5 and 10 years, respectively. Overall 5-year survival rates are between 60% and 80%. When combining systemic treatment with chemotherapy and*

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control of the primary tumor by surgery and/or radiation, survival rates for localized disease are between 70% and 75%. In the extremities, they are most frequently found in the proximal limb, with the thigh being the most common site 44% of the time.
Conclusions: *It is important to know how to recognize, evaluate and adequately treat soft tissue sarcomas, these sometimes represent a challenge, both diagnostically and therapeutically due to their special character, the infrequency of their occurrence and the difficulties in predicting outcomes. A proper differential diagnosis of the pathology must be made in order to better direct management. Developments in imaging, improvements in surgical techniques and complementary treatment methods such as radiotherapy, brachytherapy, among others, have enhanced the treatment of individuals affected with these rare disorders. A diligent preoperative study is required, consisting of an MRI to show the extent of the tumor. The current surgical approach for sarcoma resections consists of a wide en bloc resection. Surgery alone for high-grade sarcomas has a recurrence rate of 33% at 5 years, so adjuvant radiotherapy is recommended.*
KEY WORDS: *sarcomas, tumor, treatment, cancer.*

INTRODUCTION

Soft tissue sarcomas are a heterogeneous and relatively rare group of tumors that arise from mesenchymal tissues and form almost anywhere in the body. Sarcoma is a cancer that originates from cells of mesenchymal origin, such as bone, cartilage, muscle, adipose, vascular or hematopoietic tissue. It is rare with more than 50 histological subtypes(1).

Appropriate imaging, predictive immunological and genetic studies, optimized surgery and newer adjuvant and neoadjuvant treatment methods should lead to optimized outcomes for individuals with these tumors(2).

The rate of progression and the likelihood of hematogenous spread, usually to the lung, are determined mostly by the grade of the tumor. The likelihood of regional spread is low. Evaluation prior to treatment includes CT scan of the primary site, chest and MRI of the primary tumor. The mainstay of treatment is surgery; wide excision for low-grade lesions and wide or radical (compartmental) resection for high-grade tumors. Occasionally the procedure cannot be performed because of the location and extent of the sarcoma or the anticipated functional deficit. Adjuvant preoperative and/or postoperative radiation therapy improves the likelihood of local control and maintains function when adequate margins cannot be achieved. The role of adjuvant chemotherapy presents data suggest that doxorubicin-containing regimens may improve the likelihood of cure of high-grade lesions. Prognosis is determined by multiple factors including: age, tumor size, histologic grade, depth, histologic subtype and site. About 90% and 98% of recurrences are seen between 5 and 10 years, respectively. Overall 5-year survival rates are between 60% and 80%(3).

METHODOLOGY

A total of 47 articles were analyzed in this review, including review and original articles, as well as cases and clinical trials, of which 32 bibliographies were used because the information collected was not important enough to be included in this study. The sources of information were Cochrane, PubMed and Google Scholar; the terms used to search for information in Spanish, Portuguese and English were: sarcomas, soft tissue tumor, oncological surgery, cancer, osteosarcomas, brachytherapy, radiotherapy.

The choice of bibliography exposes elements related to sarcomas, description, etiology, epidemiology, presentation, evaluation and treatment.

DEVELOPMENT

Malignant bone tumors and soft tissue sarcomas make up about 14% of childhood malignancies. Successful treatment of individuals with sarcoma depends on a multidisciplinary therapeutic approach, with collaboration of various teams such as oncology, surgery, orthopedics, radiation oncology, radiology, pathology, and physiatry. By combining systemic treatment with chemotherapy and control of the primary tumor by surgery and/or radiation, survival rates for localized disease are between 70% and 75%. Children with metastatic or recurrent disease have dismal outcomes. A better understanding of the underlying biology of bone and soft tissue sarcomas is needed to optimize outcomes in infants with these tumors(4).

Soft tissue sarcomas (STS) are a group of about 60 neoplasms that can occur anywhere in the human body and affect individuals at extreme ages. They can involve skeletal muscle tissue, adipose tissue, connective tissue, blood, lymphatics and peripheral nerves. They can be found in various forms from benign lipomas to aggressive metastatic angiosarcomas(5-7).

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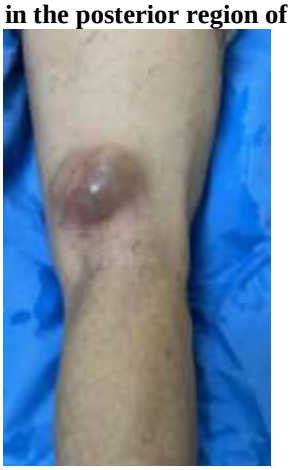
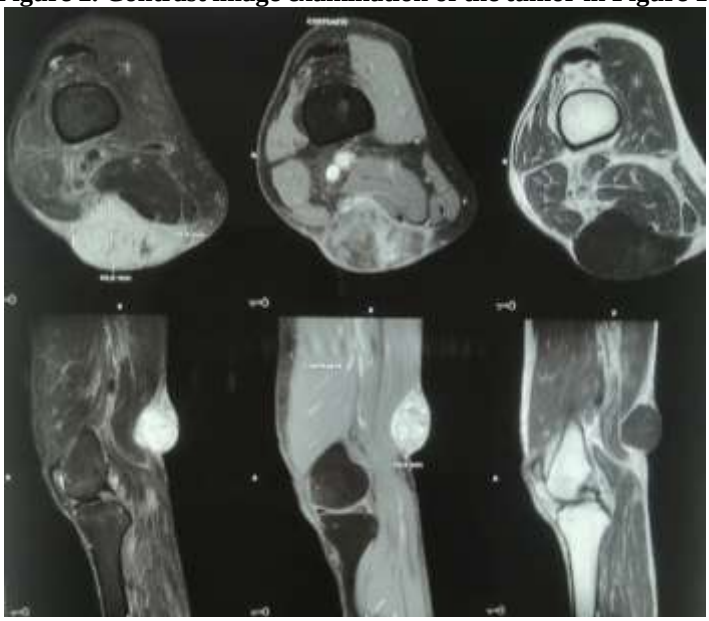
Figure 1. Tumor in the posterior region of the lower limb.



Source: The Authors.

Figure 2. Contrast image examination of the tumor in Figure 1.



Source: The Authors.

Etiology

There are multiple causative factors.

Germline Mutations.

1. Neurofibromatosis type 1 (NF1) Von Recklinghausen's disease.
- Autosomal dominant condition, encodes a protein called neurofibromin.
- A tumor suppressor of the ras oncogene signaling pathway.
- Mutations in the NF1 gene trigger various cutaneous neurofibromas.

2. Li-Fraumeni syndrome
- A rare AD disorder caused by mutations in the TP53 gene (17p13.1), which encodes p53. Some individuals will present with rhabdomyosarcoma by the age of 4 years.

3. Familial adenomatous polyposis (FAP)
- AD disorder with a mutation in the APC gene (5q21-q22).
- Tumor suppressor gene, which inhibits the localization of B-catenin in the nucleus.
- The mutant protein fails to inhibit generating an uncontrolled cell cycle and proliferation.
- It presents innumerable colonic polyps or extracolonic manifestations such as epidermoid cysts, osteomas and desmoid tumors.

4. Radiation
- Significantly contributes to a patient's long-term risk of developing STS.
- Effects are dose-dependent.
- Children who develop post-radiation STS do so on average 11.8 years later and in a dose-dependent manner.

5. Carcinogens
- Arsenic
- Thorotrast.
- Polyvinyl chloride.

6. Chronic lymphedema
- Chronic lymphatic blockage is thought to stimulate proliferation of lymphatic vessels and lymphatic vessels or lead to local immunodeficiency leading to the development of malignant disease. Common post radical mastectomy, especially post radiotherapy.

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- Individuals with parasitic nematode infections, such as filariasis, may also present with these malignant neoplasms.
- High risk of angiosarcoma formation.
- Stewart-Treves syndrome.

Epidemiology

Soft tissue sarcomas are infrequent, in 2014 in the United States 12,020 new cases and 4740 deaths were reported, representing about 1% of all cancer incidence in this country, as well as accounting for 2% of cancer-associated deaths. These are divided as to their location within the body and extremities, STS of the trunk being more common than intraperitoneal and retroperitoneal STS. In the extremities, they are most frequently found in the proximal limb, with the thigh being the most common site 44% of the time. Age at diagnosis and histologic subtype are usually related to rhabdomyosarcoma, hemangioma, neurofibroma and alveolar sarcoma, which are more common in children and young adults(8).

Pathophysiology.

Sarcomas are connective tissue tumors that may involve bone, cartilage, fat, muscle, vascular or hematopoietic tissues. Sarcomas are more infrequent compared to carcinomas. They usually grow locally and invade adjacent tissues, presenting as a hard mass or as pain due to pressure on nerves and soft tissues.

Evaluation.

Indications for preoperative imaging and biopsy assess the extent of the mass on physical examination and the expected neurovascular involvement. The possibility of lymph node involvement or distant metastasis should be considered, as well as relative resectability and functional deficits(5).

Imaging such as MRI is usually found to be the most informative for STS of the trunk and extremities. Chest CT with contrast is used in cases with high metastatic potential. The use of PET/CT has not yet become the standard because it has not shown value in distinguishing between benign and malignant disease, however, it has shown promise in assessing response to neoadjuvant chemotherapy(9,10).

When performing a biopsy, the choice is a core needle biopsy, if there is no favorable response with the diagnosis, an incisional biopsy can be performed. There is evidence that about 74% of individuals who undergo unplanned resection of sarcoma of the trunk or extremities have residual disease at the time of the next resection.

Close postoperative surveillance is appropriate due to the high risk of recurrence. A physical examination should be performed every 3 to 6 months for 2 to 3 years and then every 6 months for the next two years and finally once a year. Complementary examinations such as chest, abdominal and pelvic X-rays, as well as indications for follow-up MRI are based on the peculiarities of each affected individual in addition to those of the tumor(5).

Treatment

- **Leiomyosarcoma:** the second most common subtype of soft tissue sarcoma. It is a malignant smooth muscle tumor that can originate anywhere in the body and is usually found between the sixth and seventh decades. It is frequent in the retroperitoneum and uterus in females and in other locations in males. They are heterogeneous and well circumscribed tumors with cystic or necrotic central areas, they present positive staining for desmin and smooth muscle actin and their first line of treatment is surgical resection with negative margins(11).
- **Desmoid tumors:** infrequent forms of fibroblastic tumors, usually sporadic or related to familial adenomatous polyposis (FAP). Sporadic cases are related to pregnancy and previous trauma. They are more common in women and between 30 and 40 years of age. They can be seen in the extremities, intraperitoneal space, abdomen and/or chest wall, slow growing but aggressive. They use the WNT signaling pathway. In radiographs they are seen as homogeneous and solid in appearance with a distinct or infiltrative border(12).
- **Gastrointestinal stromal tumor (GIST):** they are the most frequent visceral soft tissue sarcoma, usually sporadic. They originate in the interstitial cells of Cajal within the gastrointestinal myenteric plexus. The most common location is the stomach, small intestine and rectum. GIST tumors have a marker for CD117, which encodes a transmembrane tyrosine kinase receptor called c-kit. Their clinical manifestations vary from being asymptomatic to symptomatic with pain, nausea, hematemesis and gastrointestinal blood loss. Metastases are frequent, the common sites being the liver and peritoneal surface(13).
- **Angiosarcoma:** a malignant tumor arising from the endothelial lining of blood vessels and can occur anywhere in the body. It usually occurs between the seventh and eighth decade of life and is most common on the scalp, head, neck and viscera. Tumors larger than 5 cm and with evidence of epithelium are considered indicators of poor prognosis(14).
- **Lipomatous tumors:** benign adipocytic tumors can arise in any area of the body and cause symptoms due to mass effect. They are usually encapsulated, homogeneous with no evidence of nodules or septa, and may contain calcifications or hemorrhage resulting from trauma. Excision beyond the tumor capsule should be performed. There is some degree of clinical overlap with the potentially malignant form and the most common soft tissue sarcoma; liposarcoma. Liposarcomas are tumors larger than 10 cm in size, with thick internal septa and lesions that typically have less than 75% adipose tissue. The treatment of liposarcoma is surgical resection with wide margins and local recurrence is frequent. Well-differentiated liposarcomas usually present a low risk of distant metastasis(15).
- **Retroperitoneal and visceral sarcomas:** they represent 15% of all soft tissue sarcomas. The average age of presentation is 54 years, with a great diversity of clinical presentations, sometimes being asymptomatic or presenting abdominal pain, weight loss, early satiety, nausea, vomiting, back or side pain, paresthesia and weakness. Gross resection is the treatment of choice(16).

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- **Sarcoma of the trunk and extremities:** several types are chemoresistant and there are conflicting results regarding the usefulness of neoadjuvant and adjuvant chemotherapy. Evidence supports a surgical margin of 1 cm with respect to adequate resection(17).

Differential Diagnosis

It is extensive, ranging from benign changes to metastatic disease. Usually, benign lesions tend to be more superficial in the dermal or subcutaneous soft tissue. Possible differential diagnoses for a soft tissue mass include:

- **Benign:** acrochordons, myositis ossificans (look for history of physical trauma), lipoma, dermoid cyst, neurofibromas, hemangioma, keloids, ganglion cyst,, pilonidal cyst, angiofibroma, rickets, among others.
- **Infectious:** abscess, cellulitis.
- **Malignant:** melanoma, keratoacanthoma, squamous cell carcinoma, basal cell carcinoma, Merkel cell carcinoma, cutaneous metastasis, Ewing's sarcoma, soft tissue sarcoma, cutaneous lymphoma, Kaposi's sarcoma, non-rhabdomyosarcoma soft tissue sarcoma, pediatric neuroblastoma, pediatric osteomyelitis, pediatric osteosarcoma, pediatric rhabdomyosarcoma, pediatric non-Hodgkin's lymphoma, among others.

Treatment

Medical Oncology.

The treatment of metastatic STS has changed over time, previously the standard first line treatment for individuals with metastatic STS was doxorubicin with ifosfamide, single agent doxorubicin or gemcitabine plus docetaxel and in those with contraindications to anthracycline treatment a gemcitabine based chemotherapy regimen was considered. Those with poor performance status or multiple comorbidities can be treated with pegylated liposomal doxorubicin or single agent gemcitabine(15).

The use of systemic chemotherapy in the treatment of non-pediatric extremity and trunk sarcomas has been extensively studied, systemic therapy can be used in neoadjuvant, adjuvant, metastatic and recurrent settings. The most common adjuvant/neoadjuvant regimens are AIM (doxorubicin/ifosfamide/mesna) and gemcitabine/docetaxel. There is a meta-analysis of adjuvant chemotherapy that showed improvement in local and distant recurrence. It is thought that the benefit of chemotherapy is likely to be small. Temozolomide, an alkylating agent, has been used as a single agent in patients with advanced, pretreated STS. Trabectedin, eribulin and dacarbazine are other therapies used(5,17,18).

Currently, trials using targeted agents and immunotherapy have been conducted. Pazopanib, an oral multi-targeted tyrosine kinase inhibitor, showed single agent activity in non-lipogenic STS. Other tyrosine kinase inhibitors, such as sunitinib and crizotinib, have shown efficacy in solitary fibrous tumors and inflammatory myofibroblastic tumors. Pembrolizumab, an anti-PD-1 antibody, shows encouraging activity in undifferentiated sarcomas(5,19).

Isolated limb perfusion (ILP) is a technique that allows the administration of significantly higher doses of chemotherapy in individuals with primary or recurrent STS who would otherwise require amputation. Agents used include melphalan, dacarbazine, doxorubicin and tumor necrosis factor (TNF), showing treatment response rates of 76%, as well as 3-year limb preservation of 71%. Concurrent chemoradiotherapy using three cycles of preoperative MAID (mesna, doxorubicin, dacarbazine and ifosfamide) with 44 Gy of split radiotherapy interdigitated with chemotherapy has also been studied showing superior overall survival, distant metastasis and disease-free survival, but at the cost of significant hematologic toxicity(20).

Figure 3. Image After Mass Excision.

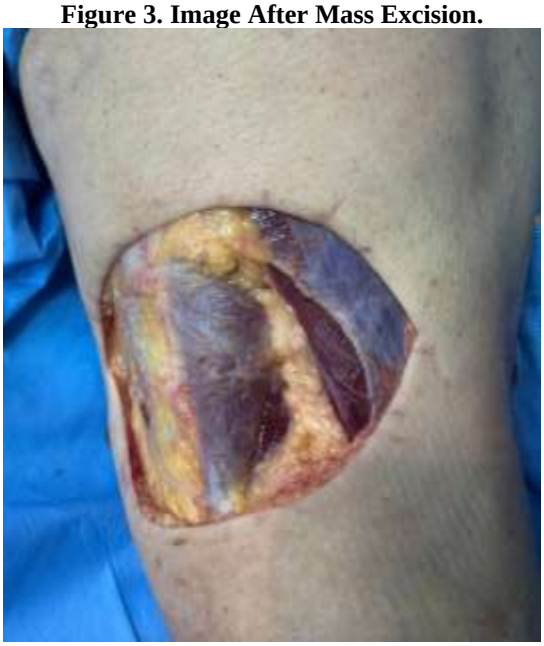

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Figure 4.Tumor Removed by Surgery.

Source: The Authors.

Surgical Oncology

Surgical resection is critical in sarcoma; currently, amputations account for <5% of all sarcoma surgeries. A diligent preoperative workup is required, usually an MRI to show the extent of the tumor. The current surgical approach for sarcoma resections consists of a wide en bloc resection to leave at least a 1 cm margin of unaffected tissue in all directions. A 2 cm margin may be considered for tumors with infiltrative borders. If the tumor is close to or displaces neurovascular structures, it is not necessary to resect them if the adventitia and/or perineurium are removed. Bone is rarely involved, resection may be required if it is involved. The margins are classified as intralesional, marginal, wide or radical. Intralesional margins go through a tumor and leave residual tumor. Marginal margins have a surgical plane through the reactive zone and a high recurrence rate. Wide margins have surrounding normal tissue but remain within the same compartment. Radicals remove the tumor and the affected compartment. The skin surrounding the biopsy site must be completely removed. Keep in mind that if the tissue defect is expected to be large, a myocutaneous flap will sometimes be necessary(21,22).

Figure 5. Marking at the site where the subsequent flap is to be cut.

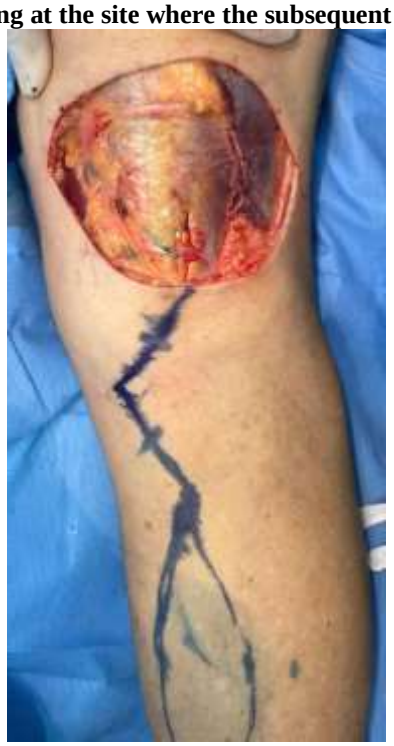

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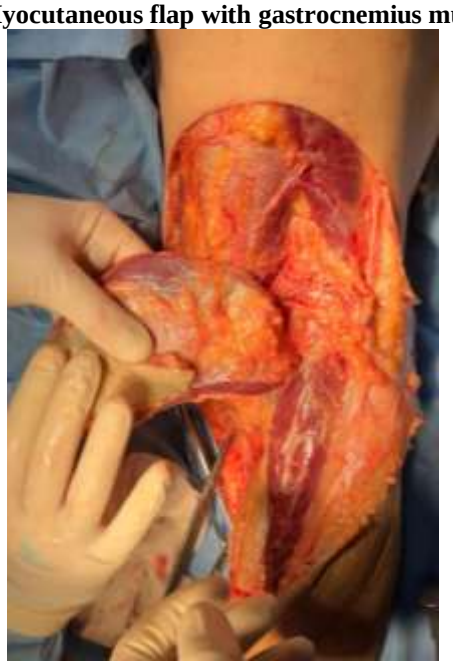
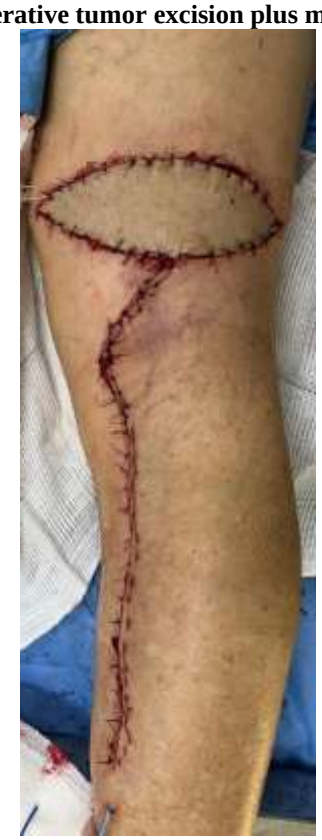
Figure 6. Myocutaneous flap with gastrocnemius musculature.



Source: The Authors.

For locally recurrent tumors, surgery may be an alternative with wide excision or amputation. Studies show that in wide excision, local recurrence-free survival at 5 and 10 years was 66% and 50%, conversely in amputations, the recurrence rate is less than 10%. Local control rates for low grade sarcomas are around 5% and can usually be treated with surgery alone. Surgery alone for high-grade sarcomas has a recurrence rate of 33% at 5 years, so adjuvant radiotherapy is recommended. Affected individuals will require significant post-treatment follow-up and ongoing physiotherapy(5,23,24).

Figure 7.Postoperative tumor excision plus myocutaneous flap.



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Nutritional Intervention

A proactive assessment of the clinical changes that occur in cancer is paramount to select the right nutritional intervention with the best possible impact on nutritional status, body composition, efficacy of management and reduction of complications, as well as optimizing survival and quality of life. To decrease the response to surgery-related metabolic stress and catabolism in malnourished individuals, the enhanced post-surgical recovery program is advised for all individuals with cancer undergoing curative or palliative surgery. In oncologic surgical individuals with moderate to severe nutritional risk, nutritional support is indicated before and after surgery. When there is significant malnutrition, the delay of surgery should be considered. When the affected individual undergoes major surgery, adequate nutritional support should be routinely provided, with special emphasis on individuals with sarcopenia for many years(25-27).

Radiation Oncology

Radiation

As limb-sparing soft tissue sarcoma resections began to replace amputations, radiation therapy was seen as a means to maintain high rates of local control while maintaining limb function. Radiation therapy can be incorporated into any phase of trunk and extremity STS treatment. Indications for radiotherapy include high-grade disease, stage II+, inoperable, recurrent disease or positive margins. The most commonly used techniques are external beam radiotherapy and brachytherapy. Preoperative radiotherapy has become the preferred form of treatment when possible. Postoperative radiotherapy can be useful in the case of positive margins, either as a boost after preoperative treatment or alone(5,28).

External beam radiation therapy (EBRT)

It is frequently used to treat sarcomas. It can be administered definitively, preoperatively or postoperatively by 3D conformal or intensity-modulated radiation therapy (IMRT). Preoperative radiotherapy is the preferred method because it allows easier target delineation, simple set-up, lower radiation doses, smaller radiation field size and better functional results. However, adjuvant therapy may be required when there are positive margins or a high-grade tumor where preoperative treatment was not planned. The dose of radiotherapy correlates with margin status, resectability and timing: individuals with resectable high-grade and intermediate-grade sarcomas where preoperative radiotherapy is planned. Usually a dose of 50 Gy is administered in 25 fractions. For preoperative radiotherapy, target delineation by CT/MRI fusion is essential(17,29,30).

Brachytherapy

Brachytherapy (BRT) is used post-surgically as monotherapy combined with external beam radiotherapy as a boost, as well as for recurrent disease. It is highly conformal, thus limiting dose to adjacent structures by optimizing dwell time positions. It is usually given at a high dose rate (HDR - > 12 Gy/h), a low dose rate (LDR - 0.4 to 2 Gy/h) or a pulsed dose rate (PDR). The HDR technique, using a high-energy radionuclide Iridium-192 with a remote afterloader, can be given on an outpatient basis, unlike previous brachytherapy techniques such as LDR implants, however it is underutilized compared to EBRT.

Individuals with excised high- and intermediate-grade sarcomas managed with adjuvant LDR BRT monotherapy have superior local control rates compared to resection alone. HDR brachytherapy showed local control rates between 78% and 90%. Brachytherapy offers highly conformal radiation plans that can help minimize dose to many previously irradiated tissues.The dose of brachytherapy will depend on the technique (LDR or HDR), margin status and the use of EBRT. As monotherapy for high/intermediate grade sarcomas with negative margins, the dose is 45 to 50 Gy for LDR and 30 to 54 Gy in 2 to 4.5 Gy/fraction given twice daily. As a boost, the preoperative EBRT dose is considered. The HDR dose ranges from 12 to 20 Gy for 2 to 3 days every 12 hours, while LDR ranges from 15 to 25 Gy. High-quality imaging is critical for radiation therapy planning, especially in individuals with sarcoma, especially MRI.

In brachytherapy, the patient will have catheters placed along the surgical site, ideally with a spacing of 1 to 1.5 cm. Catheters should not be placed over critical structures, cross catheters, or displace tissue. Catheters are implanted parallel to each other; it is also possible to have multiplane implants. Treatment usually begins 2 to 4 days after surgery. The clinical target volume (CTV) should encompass the entire tumor bed with a longitudinal margin $\geq$ 2 cm and a radial margin $\geq$ 1 cm. For each treatment, the site and integrity of the catheters should be examined. For organs at risk, various dose constraints can be used(12,31,32).

CONCLUSIONS

It is important to know how to recognize, evaluate and adequately treat soft tissue sarcomas, these sometimes represent a challenge, both diagnostically and therapeutically due to their special character, the infrequency of their occurrence and the difficulties in predicting outcomes. A proper differential diagnosis of the pathology must be made in order to better direct management. Developments in imaging, improvements in surgical techniques and complementary treatment methods such as radiotherapy, brachytherapy, among others, have enhanced the treatment of individuals affected with these rare disorders. A diligent preoperative study is required, consisting of an MRI to show the extent of the tumor. The current surgical approach for sarcoma resections consists of a wide en bloc resection. Surgery alone for high-grade sarcomas has a recurrence rate of 33% at 5 years, so adjuvant radiotherapy is recommended.

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Conflict of Interest Statement

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