EFFECTIVENESS OF SURGERY FOR MYXOID LIPOSARCOMA OF THE THIGH (CASE REPORT)

Shalva Giuashvili, Tamaz Chkhikvadze, Temur Mosiava, Tatiana Mermanishvili, Mirza Mikava, Mari Molashvili

[1]*Aleksandre Aladashvili Clinic (Tbilisi, Georgia),* [2]*Ivane Javakhishvili Tbilisi State University, (Tbilisi, Georgia),* [3]*East-West University (Tbilisi,Georgia)*

Contact person: Mari Molashvili, mrmlshvl@gmail.com

DOI: https://doi.org/10.48412/GTBGS.2024.12.39-44

Rezume

Soft tissue sarcomas (STS) account for about 1% of all adult malignancies. The prevalence in the United States is less than 20% of all soft tissue sarcomas. The most common histological subtypes were well-dediferenciated tumors (33% and 31%), followed by dediferenciated (20% in both), myxoid (19% in both) and pleomorphic (7% and 8%) tumors. Most tumors were found in the extremities (39%-41%) and retroperitoneum (21%-22%), with other areas of the body accounting for 39%. The average age of diagnosis is 50 years, although case reports of childhood-onset have been noted. According to the American Cancer Society, liposarcoma is the most common soft tissue sarcoma worldwide. No other predilection with race or gender. A slight male predominance has been reported in some studies. We report a case of a giant, recurrent myxoid liposarcoma, with necrotic skin disruption, causing immobility, significantly affecting patient's quality of life.

A 50 year-old Caucasian female approaches the clinic with complains: discomfort, pain, progressively increasing mass in the medial aspect of the right thigh, with the necrotic breakdown of the covering skin, causing significantly declared quality of life. Patient was practically unable to walk. Initially, she was diagnosed with lipoma 1 year ago, complete surgical resection was performed but patient disagreed to perform recommended immunohistochemichal investigation. Within 6 months postoperatively, recurrent liposarcoma was diagnosed and confirmed histomorphologically. Under the general anesthesia, complete resection was performed by multidisciplinary surgical team without any intraoperative or postoperative complications. She went through the one complete course of radiation therapy. After 6 months, follow-up scans were completely clear.

This case is described, to underline how important is to make an early diagnosis and resect the tumor completely, despite its' recurrent nature, giant size and close contact with the surrounding important anatomical structures and give the patient's chance to return to their normal life.

Key words: Recurrent liposarcoma, total resection, RT, cancer treatment

Introduction

Liposarcomas arise from lipocytes. The relative frequency at different body sites is dependent on the tumor subtypes. They can be located in any parts of the body, usually weights a few gram and are usually less than 2cm. For example, dedifferentiated liposarcoma is much more common in retroperitoneal locations, while myxoid liposarcoma occurs in the lower extremities. Histologically it can be divided into five subtypes: well-differentiated, myxoid, round cell, dedifferentiated and pleomorphic. Myxoid liposarcoma is characterised by a prominent myxoid matrix and round to oval-shaped cells with lipid vacuoles, constitutes approximately 5% of all adult soft tissue sarcomas and 20-30% of all liposarcomas, predominantly affecting adults between the ages of 40 and 60. More than half of cases are seen in the thigh muscles, where it is typically found in the deep soft tissue of the extremities. Specific chromosomal translocation t(12;16)(q13;p11) or less commonly t(12;22)(q13;q12) plays a key role in the pathogenesis, resulting in the fusion of the DDIT3 gene with either FUS or EWSR1. Many people with MRCLS do not have symptoms when the cancer first starts. Later, when the tumor gets larger, symptoms can include: a visible lump under the skin, pain, tiredness, nausea, unintentional weight loss. The tumor typically appears as well define mass on MRI. The primary treatment is surgical resection with clear margins. Adjuvant radiation therapy may be considered, especially if complete surgical removal is challenging or if there is a risk of local recurrence. For advanced or metastatic cases chemotherapy may be considered. We report a case of a recurrent, giant, myxoid liposarcoma of the right thigh, which was completely resected successfully.

Case presentation

50 years-old Caucasian female, known to have right lower limb recurrent sarcoma, presented at our clinic complaining of a pain and discomfort in medial aspect of the

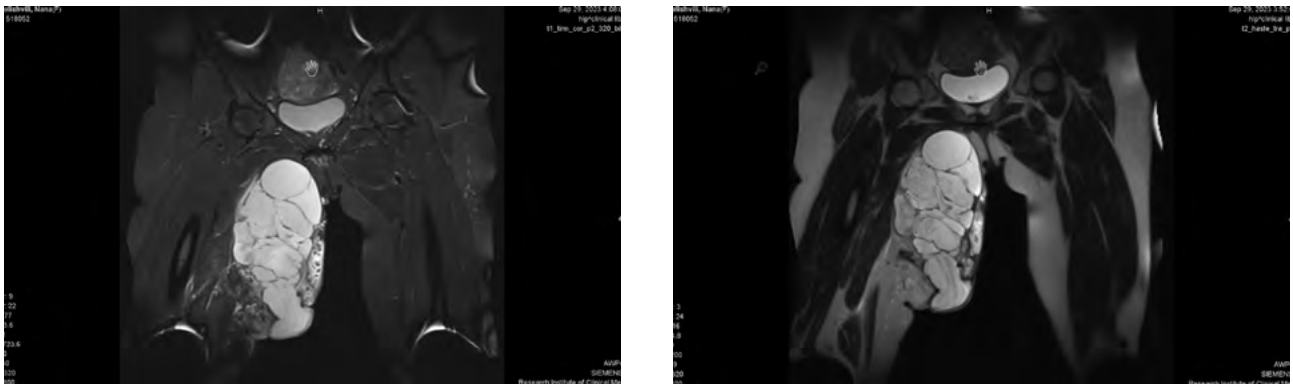


Figure1. Preoperative MRI scan

right thigh, necrotic breakdown of the skin extending from the public area to the lower third of the thigh, restriction of movement, inability to walk, severe decrease in working capacity. 1 year ago she was diagnosed with the lipoma of the right thigh, which was surgically resected. Based on pathomorphologic results, patient was advised to perform immunohistochemichal investigation, to rule out liposarcoma, but she dismissed the recommendation. In 6 months postoperatively, she noticed a recurrent slow growing soft tissue structure, swelling, skin necrosis and bleeding from surgical site. Treatment was continued in Italy, where biopsy was taken and based on pathomorphologic and immunohistochemistry results, recurrent liposarcoma was diagnosed. Because of the giant size, recurrent nature and close contact to the important anatomical structures, case was assessed as inoperable and patient was unable to find a surgeon, who would have agreed to operate on her. She returned back to Georgia, took a consultation from oncologist, underwent additional investigating studies (chest, pelvic and abdominal CT scan with IV contrast, MRI of both femur in T1, T2 modes, saggital and transverse slices, biopsy). MRI showed 22.8x14.2x10.2sm irregular cystic lesion, characteristic for liposarcoma. CT scan results were unremarkable (detection of distant metastases), biopsy results confirmed T4N1M0 myxoid liposarcoma. She was referred to our clinic for surgical consultation. Physical exam showed average built woman of normal health, without any systemic diseases. Local examination showed a hard, non-movable tumor of 25x18cm in the medial aspect of right thigh, covering all the surface from the pubic region to the lower one-third of the medial thigh, with the necrotic breakdown of the covering skin. The range of motion was disrupted, walking was practically impossible. Tumor was assessed as operable, but risk factors and possible complications were explained to the patient. Despite high risk of the surgery, patient agreed to undergo the procedure and at least, have a chance to return to her normal lifestyle again. Elective surgery was planned [fig.1]

Figure 2. Specimen from en-block resection of the tumor

Table 1. Follow-up guidelines for liposarcoma

Follou-Up	Fequency of Visits	
	in Years 1-3	in Years 4-5
ESMO	Every 3-4 months	Every 6 months
NCCN	Every 3-6 months	High grade - Every 6 months
		Low grade - annually
Local guideline	Every 4 months	Every 6 months

Multidisciplinary surgical team (oncosurgery, angiosurgery) performed the operation. Intraoperatively, two semi-oval incisions were made on the medial surface of the femur, necrotic skin, including previous surgical scar, was resected and encapsulated tumor was seen near the great saphenous vein, superficial femoral artery, growing deeply subfascially, closely attached to the muscles. With technical difficulties, en-block resection of the tumor was performed – part of the Sartorius, vastus medialis muscles were resected, tumor mobilized and finally, resected along with all the lymphatic collectors, without disturbing capsular integrity. Surrounding nerves and arteries were saves, so the normal leg function was maintained. The resected specimen, weighting 7kg, including necrotic skin and lymphoid tissues, was sent to morphology [fig.2]. The final pathologic diagnosis, as the preoperative one, was T4N1M0 myxoid liposarcoma, confirmed that the tumor was completely resected with 1cm clear surgical margins.

The patient tolerated operation well, had an uneventful postoperative recovery and was discharged home on postoperative day 5. She went through the one complete course of radiation therapy. She has been followed-up and screened continuously.

Relapse following primary treatment occurs frequently, with 40–50% of STS patients developing either local or distant disease recurrence. Therefore, routine follow-up is designed to detect disease recurrence as early as possible because early treatment improves prognosis. Current ESMO-EURACAN (European Society for Medical Oncology- European Reference Network for rare adult solid cancers) and NCCN (National Comprehensive Cancer Network) consensus guidelines recommend follow-up visits every 3 to 6 months in the first three years, then twice a year up to the fifth year and annually thereafter, with NCCN guidelines, distinguishing between low- and high-grade STS [Table 1]. As guidelines recommend, our patient follows up in every 3 months.

First follow up MRI scan, 3 months postoperatively - Seroma (postoperative fluid collection, as reaction to a surgical exposure) [fig.3]

Second follow-up scan, 6 months postoperatively, was absolutely clear, indicating complete remission. [fig.4]

Discussion

Liposarcomas appear to originate from primitive mesenchymal cells, rather than mature adipose tissue and most commonly occurs in the extremities accounting for 52%, followed by retroperitoneum (19%) and inguinal (12%) regions. WHO classifies liposarcomas into five histologic types: well-differentiated, myxoid, round cell, pleomorphic, dedifferentiated. It frequently occurs in the deep soft tissues of proximal extremities and is usually painless, grows slowly to reach a large size.

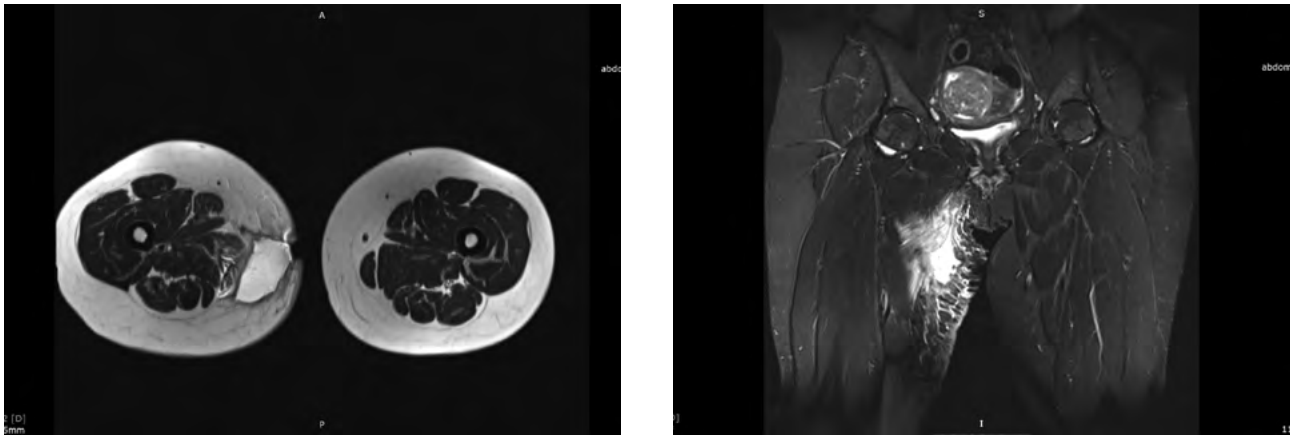


Figure 3. First postoperative MRI scan – seroma

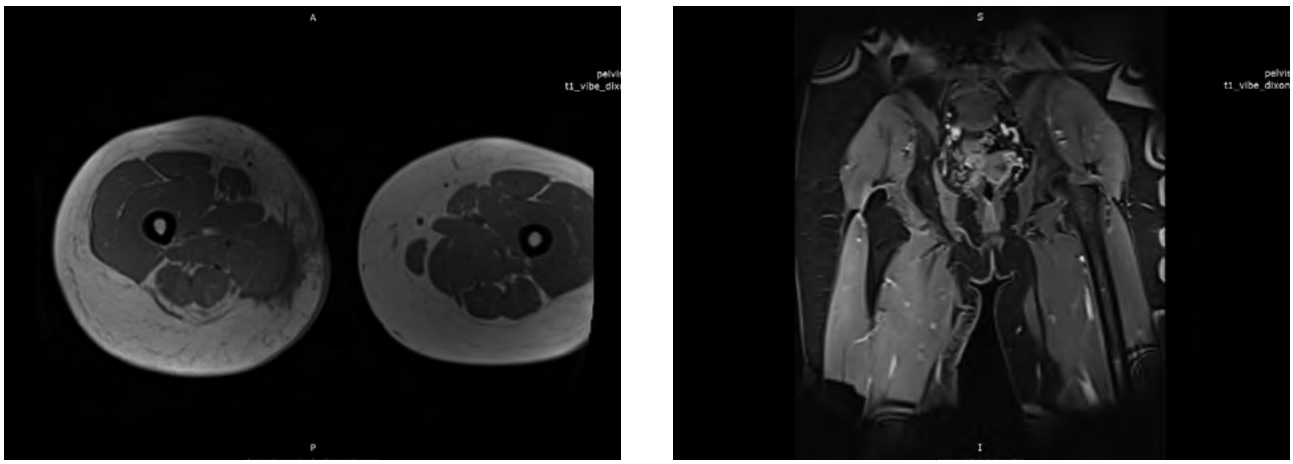


Figure 3. 6 months postoperatively follow-up MRI scan – complete remission

Myxoid liposarcoma is characterised by a prominent myxoid matrix and round to oval-shaped cells with lipid vacuoles, constitutes approximately 5% of all adult soft tissue sarcomas and 20-30% of all liposarcomas, predominantly affecting adults between the ages of 40 and 60. More than half of cases are seen in the thigh muscles, where it is typically found in the deep soft tissue of the extremities. Specific chromosomal translocation t(12;16) (q13;p11) or less commonly t(12;22) (q13;q12) plays a key role in the pathogenesis, resulting in the fusion of the DDIT3 gene with either FUS or EWSR1.

Many people with MRCLS do not have symptoms when the cancer first starts. Later, when the tumor gets larger, symptoms can include: a visible lump under the skin, pain, tiredness, nausea, unintentional weight loss.

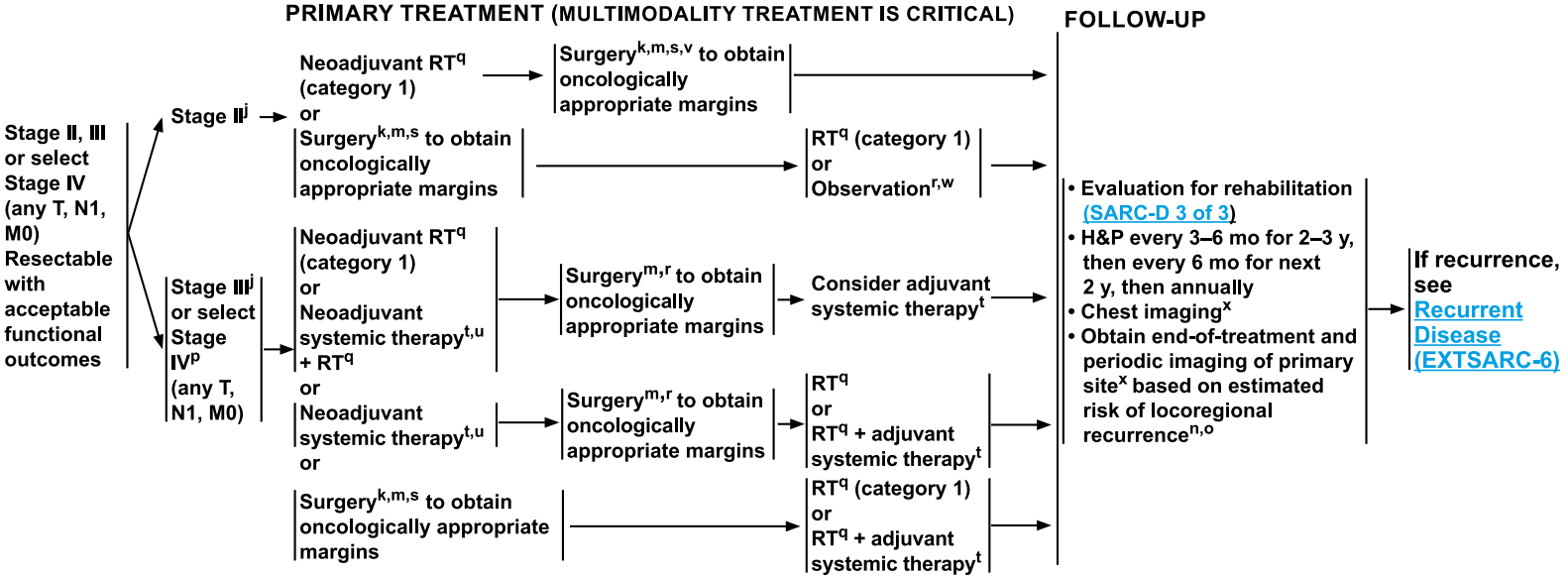
The tumor typically appears as well define mass on MRI. The primary treatment is surgical resection with clear margins. Lesions that are located in the extremities have favorable prognosis among young candidates and adjuvant radiotherapy yields greater relapse-free survival in those over 30 years of age. The primary treatment for high risk patients is surgical resection and local control with adjuvant radiotherapy [fig.5]. As usual, surgery is limb sparing, with or without radio- and/or chemotherapy. Adequate clinical and radiological assessment is important to assess the resectability of the lesion. Larger tumors may need second look excision surgery to clear positive margins.

Conclusion

In this case, despite difficult surgical characteristics (close attachment to the great saphenous vein, superficial femoral artery, muscles) of the tumor, recurrent, giant, myxoid lyposarcoma, weighting 7kg, causing necrotic disruption of the covering skin, was successfully resected without intra- or postoperative complications. This case underlines the importance of early pathomorphologic and immunohistochemical diagnosisis of liposarcoma, giving an example, that even in high risk patients, experienced surgeon can perform successful limb sparing surgery as the first choice of treatment and give a patient chance to return and maintain normal quality of life.

ლიტერატურა:
References:

1. *Liposarcoma, Rabia Zafar; Yurong Wheeler StatPearls Publishing; 2024 Jan*
2. *Liposarcoma: Advances in Cellular and Molecular Genetics Alterations and Corresponding Clinical Treatment, Lingge Yang,1,2 Shiqi Chen,1,2 Peng Luo,1,2 Wangjun Yan,1,2,and Chunmeng Wang, PMCID: PMC6930414 PMID: 31892977*
3. *Myxoid Liposarcoma: Treatment Outcomes from Chemotherapy and Radiation Therapy, Varun Chowdhry, 1 Saveli Goldberg, 2 Thomas F. DeLaney, 2 Gregory M. Cote, 3 Ivan Chebib, 4 Jason Kim, 5 Santiago A. Lozano-Calderon, 5 and Karen De Amorim Bernstein , PMCID: PMC6236966 PMID: 30515050*
4. *Management of myxoid liposarcoma of the extremity, Yehia Tfayli,1 Ahmad Baydoun,2 Ahmad Salaheddine Naja,1 and Said Saghieh1, PMCID: PMC8228380 PMID: 34188698*
5. *Myxoid Liposarcomas: Systemic Treatment Options, Elise F Nassif 1 2 3, Emily Z Keung 3, Prapassorn Thirasastr 1, Neeta Somaiah 4, doi: 10.1007/s11864-023-01057-4. Epub 2023 Feb 28*
6. *Follow-Up after Curative Surgical Treatment of Soft-Tissue Sarcoma for Early Detection of Recurrence, Anouk A. Kruiswijk,1,2,*† Laurien S. Kuhrij,1,† Desiree M. J. Dorleijn,2 Michiel A. J. van de Sande,2 Leti van Bodegom-Vos,1 and Perla J. Marang-van de Mheen, Cancers (Basel). 2023 Sep, PMCID: PMC10527323, PMID: 37760585*



[j] See AJCC Staging, 8th Edition (ST-5 and ST-6).
[k] Principles of Surgery (SARC-D).
[m] See SARC-D for either a previous unplanned excision or a failure to obtain oncologically appropriate margins with planned sarcoma surgery.
[n] In situations where the area is easily followed by physical examination, imaging may not be required.
[o] After 10 years, the likelihood of developing a recurrence is small and follow-up should be individualized.
[p] For management of a primary sarcoma with synchronous regional nodal metastatic disease, see above for treatment of the primary tumor and refer to EXTSARC-6 for management of nodal disease.
[q] See Principles of Radiation Therapy (SARC-E) for discussion of adjuvant versus neoadjuvant therapy.
[r] A prospective study demonstrated low rates of local recurrence with surgery alone in carefully selected patients with high-grade tumors <5 cm (Pisters PW, et al. Ann Surg 2007;246:675-681). Consider omission of RT for tumors <5 cm resected with wide margins if a repeat resection would be feasible with low morbidity in the case of a recurrence.
[s] In selected cases when margin status is uncertain, consultation with a radiation oncologist is recommended. Re-resection, if feasible, may be necessary to render margins >1.0 cm.
[t] Systemic Therapy Agents and Regimens with Activity in Soft Tissue Sarcoma Subtypes and Aggressive Soft Tissue Neoplasms (SARC-G).
[u] FDG-PET/CT may be useful in determining response to systemic therapy (Schuetze SM, et al. Cancer 2005;103:339-348).
[v] Re-image using MRI with and without contrast (preferred for extremity imaging) or CT with contrast to assess primary tumor and rule out metastatic disease. See Principles of Imaging (SARC-A).
[w] Resections with wide negative margins may be considered for observation alone if the risk of radiation is unacceptable.
[x] See Principles of Imaging (SARC-A).

Note: All recommendations are category 2A unless otherwise indicated.

Figure 5. Liposarcoma – treatment guidelines.

ბარძაყის მიქსოიდური ლიპოსარკომის ოპერაციის ეფექტურობა (კლინიკური შემთხვევა)

შალვა გიუაშვილი[1,2,3], თამაზ ჩხიკვაძე[1], თემურ მოსიავა[1,3], ტატიანა მერმანიშვილი[1,3], მირზა მიქავა[1], მარი მოლაშვილი[1]

[1]შპს „ალექსანდრე ალადაშვილის კლინიკა", [2]ივანე ჯავახიშვილის სახელობის თბილისის სახელმწიფო უნივერსიტეტი, [3]აღმოსავლეთ-დასავლეთ უნივერსიტეტი

პასუხისმგებელი პირი: მარი მოლაშვილი, mrmlshvl@gmail.com

DOI: https://doi.org/10.48412/GTBGS.2024.13.39-44

რეზიუმე

რბილი ქსოვილების სარკომა მოიცავს ზრდასრულთა სიმსივნეების 1%-ს. გავრცელება აშშ-ში - რბილი ქსოვილების სარკომის 20%-ზე ნაკლები. ლიპოსარკომა წარმოადგენს ყველაზე გავრცელებულ ტიპს, რომელსაც გამოეყოფა რამოდენიმე მორფოლოგიური ქვეტიპი: მაღალი-დედიფერენცირებული (20%), მიქსოიდური (19%), პლეიომორფული (7%-8%). სიმსივნეების უმეტესობა გვხვდება კიდურებში (39%-41%) და რეტროპერიტონეუმში (21%-22%), ხოლო სხეულის დანარჩენი ნაწილები მოიცავს სიმსივნეების 39%-ს. დიაგნოსტირებისას პაციენტების საშუალო ასაკია 50 წელი, თუმცაღა აღწერილია სიმსივნეები ბავშვთა ასაკშიც. ამერიკის ონკოლოგთა ასოციაციის თანახმად, ლიპოსარკომა წარმოადგენს მსოფლიოში ყველაზე გავრცელებულ რბილქსოვილოვან სარკომას. დამახასიათებელია მამრობითი სქესის პოპულაციის არამკვეთრი მიდრეკილება სიმსივნისაკენ. ქვემოთაღწერილი შემთხვევა წარმოადგენს გიგანტური ზომის, რეციდიულ, მიქსოიდური ტიპის ლიპოსარკომას, კანის საფარველის ნეკროზული ცვლილებებით, დაჩირქებით. სიმსივნე იწვევდა პაციენტის პრაქტიკულად სრულ იმობილიზაციას და ცხოვრების ხარისხის მკვეთს გაუარესებას.

ალექსანდრე ალადაშვილის სახელობის კლინიკას მომართა 50 წლის ქალბატონმა, რომელიც უჩიოდა ტკივილს, დიდ ზომის, რეციდიული სიმსივნური წარმონაქმნის არსებობას მარჯვენა ბარძაყის მედიალურ ზედაპირზე, კანის საფარველის ნეკროზული რღვევით. ჩატარებული ლაბორატორიულ-ინსტრუმენტული და მორფოლოგიური კვლევების საფუძველზე დიაგნოსტირდა მარჯვენა ბარძაყის მიქსოიდური ლიპოსარკომა T4N1M0. სიმსივნური წარმონაქმნი, ლიმფურ კოლექტორებთან ერთად, ერთ ბლოკად იქნა ამოკვეთილი. ჰოსპიტალიზაციამ ჩაიარა გართულებების გარეშე. 5 სტაციონარული დღის შემდეგ, პაციენტი დამაკმაყოფილებელ მდგომარეობაში გაეწერა ბინაზე. ჩაიტარა სხივური თერაპიის 1 სრული კურსი. 2 თვის შემდგომ, საკონტროლო MRI კვლევაზე, სიმსივნის სარეცელის მიდამოში აღინიშნებოდა სერომა. ოპერაციიდან 6 თვის შემდეგ, მეორე საკონტროლო კვლევაზე პათოლოგიური ცვლილება არ იქნა ნანახი, პაციენტი იმყოფება სრულ რემისიაში.

ჩვენს ქეისში, მიუხედავად სიმსივნის უზარმაზარი ზომისა, რეციდიული ხასიათისა, ირგვლივმდებარე კუნთებთან და სისხლძარვებთან მჭიდრო კავშირისა და ნეკროზულ-ჩირქოვანი ცვლილებებისა, წარმატებით მოხერხდა სიმსივნური წარმონაქმნისა და ლიმფური კოლექტორების ამოკვეთა ერთ ბლოკად, რას წარმოადგენს მაგალითს, რომ სრულყოფილ ქირურგიულ ჩარევას გადამწყვეტი მნიშვნელობა აქვს ლიპოსარკომების მკურნალობაში.

საკვანძო სიტყვები: რეციდივული ლიპოსარკომა, ტოტალური რეზექცია,