კლინიკური შემთხვევა

PRIMARY LEIOMYOSARCOMA OF THE BONE (CASE REPORT)

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Resume The paper presents a rare clinical case of bone leiomyosarcoma (musculoskeletal system malignant neoplasm) surgical treatment performed by a group of authors. Described is a contemporary method of surgical treatment (reconstruction of the proximal tibia with onco prosthesis) of this lesion with its complications and outcomes. It describes the correct management of this disease and the need of multidisciplinary approach.

Key words: leiomyosarcoma, malignant bone tumor, chemotherapy

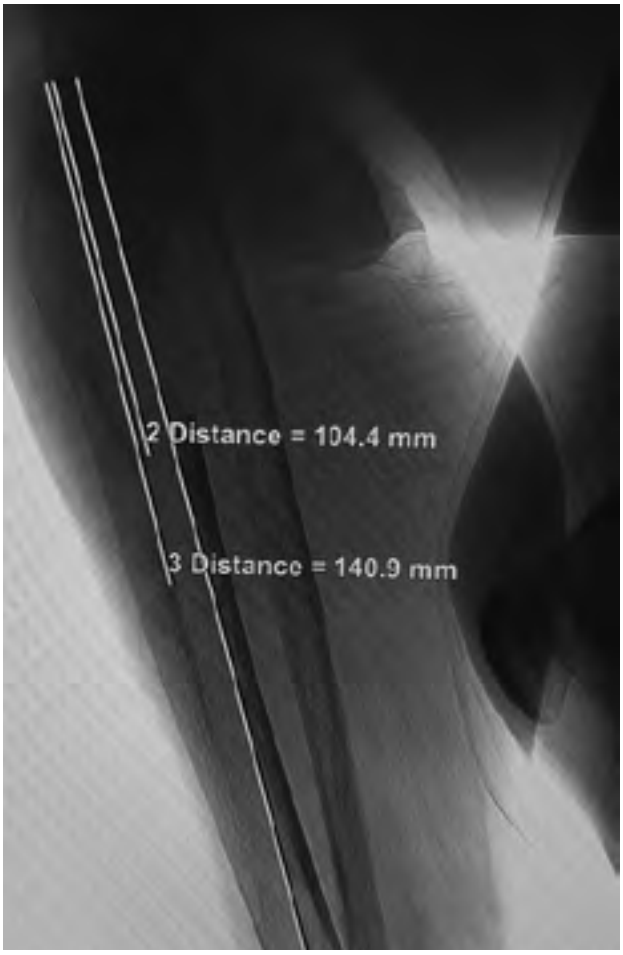
INTRODUCTION

Leiomyosarcoma of bone is a very rare primary malignant bone tumor affecting a predominantly older population, accounting for <0.7% of all primary malignant bone tumor. They were first described in 1965 by Evans and Sanerkin [1] in the proximal tibia of a 73-year-old man and have since been slowly accepted in published data as a unique classification described in the literature by series of small case studies. Little is known about the biology and clinical behavior of bone LMS due to its extreme rarity. No data are available about chemotherapy in neoadjuvant, adjuvant, or advanced settings. Surgery with a curative intent is the cornerstone of treatment of localized disease. Further research is needed to identify more effective therapies.

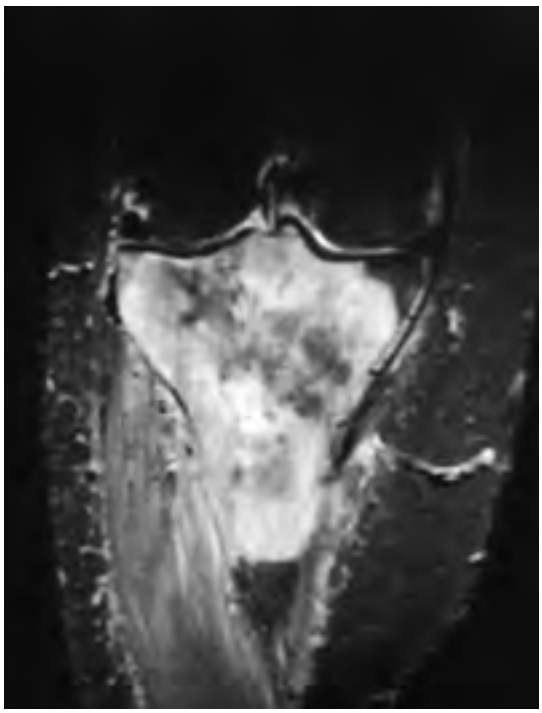
CASE REPORT

Patient 67 year old woman. Complains of progressive pain and restriction of motion in her right knee for 9 months. His past medical history was positive for diabetes mellitus treated with oral hypoglycemic agents. There was no history of smoking or alcohol consumption. On x-ray the solitary osteolytic lesion with focal cortical destruction in the right proximal tibia. The formation gradually increased in size with a worsening of the pain. A subsequent MRI scan of the right tibia showed the presence of osteolytic lesions with a focal cortical destruction possibly correlated to pathologic fracture. The lesion was located predominantly in the medullary cavity and presented a soft tissue extension from the bone with indistinct tumor margins. The patient could not weight bear on the right lower extremity and had a severe Varus deformity.

The patient was evaluated at our Institute by an Multidisciplinary Team, composed of an orthopedist, an oncologist, a radiologist, a pathologist. The morphology and immunohistochemistry studies reviled the primary leyomiosarcoma of the bone. The biopsy was performed on the line of future surgical incision, which revealed the presence of spindle cells with positive reaction for SMA and desmin at immunohistochemical analysis, without the



Picture 1. X-ray the solitary osteolytic lesion with focal cortical destruction in the right proximal tibia



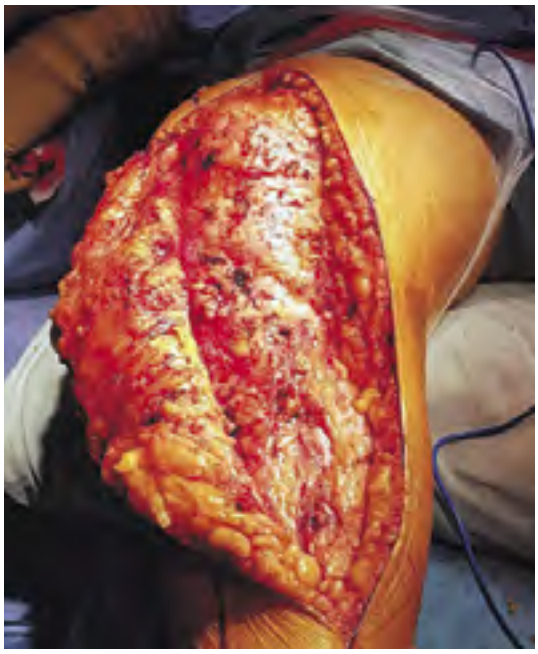
Picture 2. MRI scan of the right tibia

presence of cartilage or bone matrix. These results were consistent with a high-grade malignant LMS arising from the bone. The multidisciplinary team reviewed the patient clinical condition, the radiographic imaging, and the results of the biopsy, suggesting surgery of the bone LMS. Subsequently, the patient underwent surgery for removing the bone lesion and reconstruction of the proximal tibia with modular silver coated proximal tibia prosthesis.

After Adequate preoperative planning surgical team decided the length and extent of resection (the maximum tumor dimensions may be either intramedullary, periosteal or in terms of the associated soft tissue component). Both, the magnetic resonance imaging (MRI) and the plain radiograph were carefully evaluated. A 2-3 cm marrow margin as calculated on the T1-weighted MRI image was considered an adequate resection margin.

The antero medial surgical approach was performed for easier access to the popliteal vessels, fibula and medial gastrocnemius muscle. The biopsy tract was removed with skin and specimen to avoid local recurrences. The patellar tendon was sectioned proximal to its attachment on the tibia during resection. The popliteus muscle remains on the specimen due to oncologic margins. The vascular bundle was dissected and distal perfusion was controlled during whole surgery to avoid vascular complications. The tibio fibular joint was free from tumor so fibular resection was not performed. Prior to this the lateral popliteal nerve was carefully dissected free to prevent injury to it.

We use elective medial gastrocnemius flap in proximal tibial resections. The flap is preferably dissected out after resecting the proximal tibia but prior to implanting the prosthesis. This is technically easier than after the prosthesis is implanted. Care must be taken not to carry the dissection too far proximally to avoid injury to the vascular pedicle of the medial gastrocnemius muscle (the medial sural artery, the main pedicle of the medial gastrocnemius muscle, arises off the popliteal artery 1-2 cm below the joint line). The muscle flap is brought forward anteriorly to cover the prosthesis. It serves the dual purpose of providing a layer of muscle cover to the subcutaneous prosthesis and also providing a biological anchorage to the patellar tendon which is sutured to it with absorbable sutures. In certain situations where excess skin may need to be excised at the time of resection (because of an improper biopsy), the gastrocnemius muscle flap also serves as a good



Picture 3. The antero medial surgical approach

Picture 4. The stage of mobilization and excision

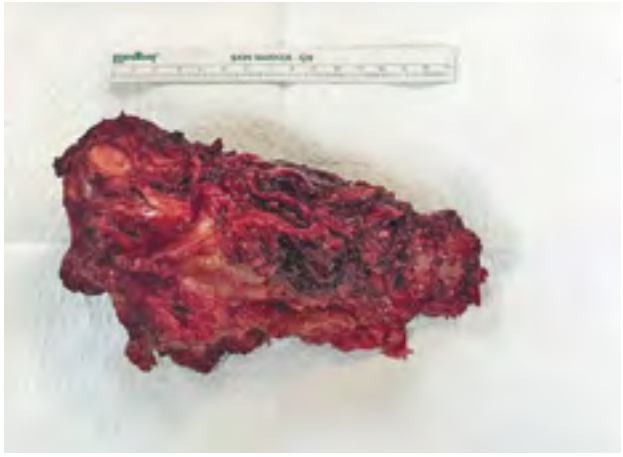
Picture 5. The stage of mobilization and excision

Picture 6. Excised bone tumor

bed for a split thickness graft if primary skin closure is not possible. In current case there was no need to use skin flap, the wound was closed with primary sutures. Postoperative care was similar as the primary knee replacement. No prolonged antibiotics were used. Patient received low molecular heparin for 5 weeks and the Brace in full extension was used for 6 weeks to allow the patella tendon healing to the gastrocnemius flap. After 6 weeks the knee flexion was started by CPM machine. No local recurrence on 9 months postoperatively.

DISCUSSION:

Bone lesions are primary diagnosed by X-ray. The most common symptoms are pain, swelling, and, occasionally, a palpable mass.The bone tumors have a predilection to age and location. In about 15% of cases, patients present with a pathological fracture. In elder patients >50 years the most common bone tumors are: Multiple myeloma, Metastasis, Chondrosarcoma. Major differential diagnoses

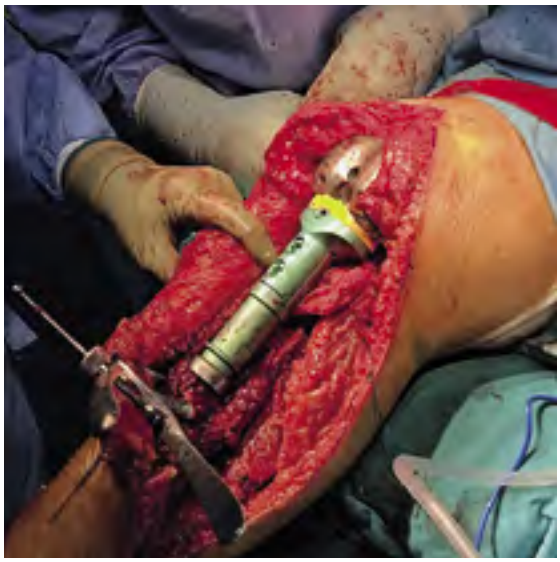


Picture 7. მიღებული მასალა ჰისტომორფოლოგიური კვლევისთვის

of the leiomyosarcoma of bone are metastatic carcinoma of unknown primary origin, lymphoma, and osteosclerotic myeloma. It is important to differentiate between leiomyoma and leiomyosarcoma especially when a previous history of benign leiomyoma exists [10] so the the bone biopsy and tumor grading is mandatory. Adelani et al., (2009) [1] found the median age affected to be 47 years, with a range from 9 to 87 years, while the median age in our series was 46, ranging from 9 to 88 years. Generally, it is felt that there is an equal distribution between the sexes for primary leiomyosarcomas [2, 5, 6] Many of the case series published have reflected a poor prognosis, although no one has successfully stratified the risk according to stage/grade. Antonescu et al. produced the largest series to our knowledge [2], presenting 33 patients with primary leiomyosarcoma between 1977 and 1996. They had an average follow-up period of 30 months and found no significant differences in disease free of overall survival rates between low- and high-grade tumors. In that series, only 21% had neoadjuvant chemotherapy, and there was no clear difference in survival between those treated with chemotherapy and those without. More recently, Rekhi et al. [8] presented a series of 8 cases, none of whom had chemotherapy. All the patients went on to develop metastases within 12 months following the original diagnosis leading the authors to conclude that it was a dismal condition to have. The most important prognostic factors associated with a decrease in survival were age >40 years, size >8cm, the presence of a pathological fracture, amputation, involved margins, and a poor response to preoperative chemotherapy.[7,11]

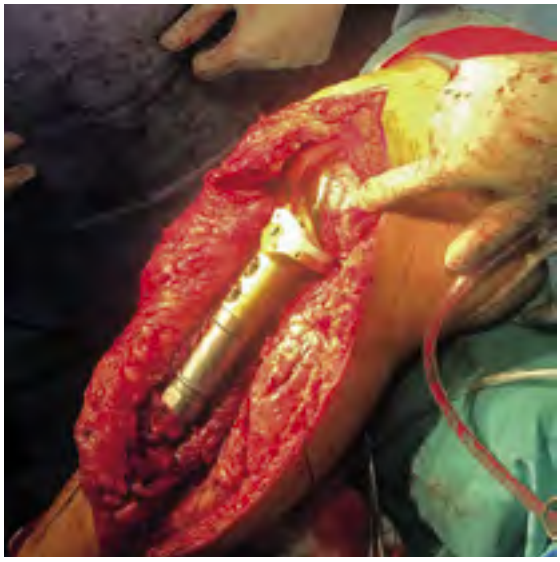
CONCLUSION:

Clinical features of primary bone LMS and relevant prognostic factors are not well defined due to the few available data obtained mostly from retrospective analy-

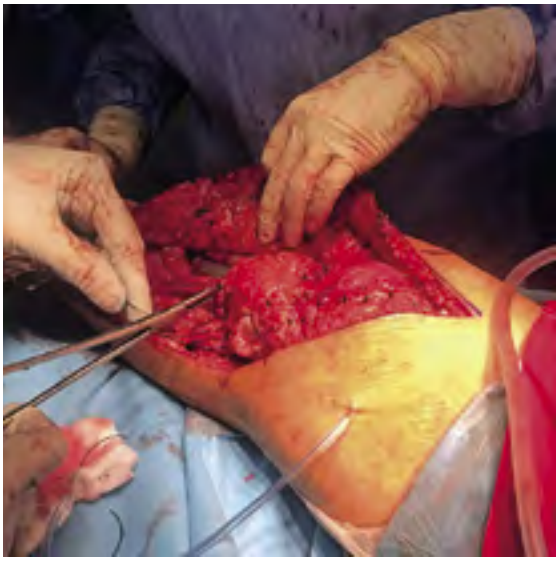


Picture 8. Stages of prosthesis implantation

Picture 10. Completion of surgery



Picture 9. Stages of prosthesis implantation



Picture 11. Completion of surgery

ses, case reports, and small case series. Surgical excision with wide margins remains the gold standard for curative management, amputation usually being reserved for tumors surrounding the neurovascular bundle, or with extensive soft tissue involvement. The role of adjuvant and neo-adjuvant treatments in localized primary bone LMS and their effect on a long-term prognosis is still unclear.

Due to the rarity of this disease, treatment of bone LMS remains highly personalized. [3] Brewer at all have shown that prognosis is based on the stage of diagnosis; with Enneking stages 1b or 2a cases achieving 100% survival, stage 2b tumours having a 60% survival at 5 years and 43% survival at 10 and 15 years.

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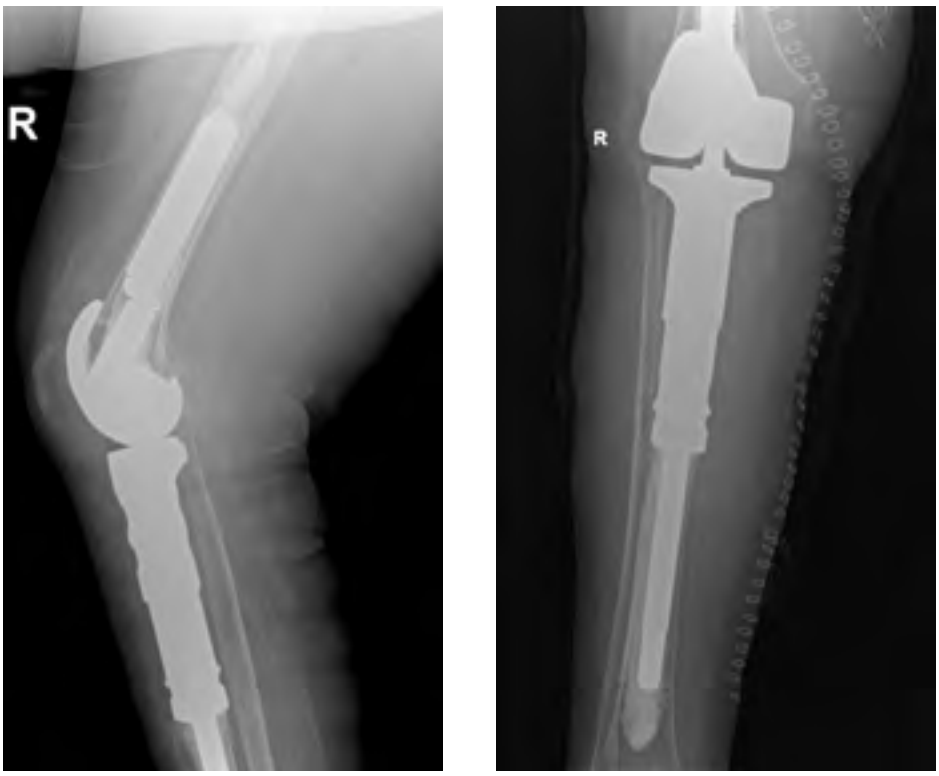

Picture 12. Control X-ray

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ძვლის პირველადი ლეიომიოსარკომა (შემთხვევის აღწერა)

მიხეილ ზიმლიცკი, გიორგი ზიმლიცკი, გრიგოლ ბრეგაძე, გიორგი ლორია

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რეზიუმე ავტორთა ჯგუფის მიერ წარმოდგენილია ლეიომიოსარკომის (ძვალ სახსროვანი სისტემის ავთვისებიანი სიმსივნე) ძალზედ იშვიათი ოპერაციული მკურნალობის შემთხვევა. სტატიაში აღწერილია პათოლოგიის ოპერაციული მკურნალობის თანამედროვე მეთოდი - წვივის პროქსიმალური სეგმენტის ონკოპროთეზირება, თანამდევი სირთულეებითა და გამოსავლით. აღწერილია ამ დაავადების სწორი მენეჯმენტი და მულტიდისციპლინური მიდგომის აუცილებლობები.

საკვანძო სიტყვები: ლეიომიოსარკომა, ძვლის სიმსივნე, ქიმიოთერაპია