

Limb Salvage Surgery in Enneking's Stage III a – A Case of Leiomyosarcoma

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ABSTRACT

Limb saving surgery is today's tendency in treating the musculoskeletal tumours. Preserving the limb should not only pursue a good oncological and functional result but also aim at good psychological outcome. Our aim was to treat a very rare case of proximal femoral leiomyosarcoma with present solitary lung metastasis by conservative operative procedures.

Key words: limb saving surgery, tumour, leiomyosarcoma, lung metastasis

Introduction

Soft tissue sarcomas (STS) are a heterogeneous group of malignancies that arise most commonly in the extremities, with a frequency at this site of about 17 per million¹. Primary leiomyosarcoma of bone is a very rare STS firstly reported by Carmody et al. in 1944². Clinical follow-up suggests that primary osseous leiomyosarcoma has an aggressive biologic behaviour with early metastasis^{3–8}. Radiological features of the tumors are rather nonspecific, and most have an osteolytic lesion^{5,9,10}. Early surgical intervention by radical or wide excision offers the best chance of cure^{11,12}. Adjuvant therapy is still a question^{4–6,8,12–15}. Today, tendency in treating the musculoskeletal tumours is the preserving of the limb¹³. The limb-saving surgery should not only pursue a good oncological and functional result but also aim at good psychological outcome¹³. Our aim was to treat the very rare case of proximal femoral leiomyosarcoma with present solitary lung metastasis by conservative operative procedures.

Case Report

A sixty years old woman came to our department with gradually growing pain in the left hip. The symptoms started four months ago and she could not recall any relevant trauma to the area. Despite rest and pain-killers the patient developed antalgic gait and night pain.

Physical examination revealed antalgic gait and with pain limited passive or active motion in the left hip. Vas-

cular and neurological parameters were normal, with no apparent peripheral, sensory, or motor loss except muscular hypotrophy of the left leg.

The abnormal laboratory findings were a sedimentation rate of 76 mm in the first hour and the high levels of liver enzymes. The erythrocytes, leukocytes, electrolytes, and serum enzymes levels were within normal limits.

Radiograph of the left hip revealed a poorly defined osteolytic mass, with suspected destruction of bone in the upper cortex of femoral neck. The tumour had a mainly metaphyseal location extending from linea intertrochanterica into the femoral head (Figure 1). Magnetic resonance revealed large intraosseous component with heterogeneous signal intensity throughout the lesion. The tumour occupied the femoral neck and extended into the femoral metaphysis to the level of lesser trochanter and femoral head without extension to surrounding soft tissue (Figure 2). The scintigraphy (Tc-99m) revealed accumulation only in the left proximal femur. The CT scan of chest, abdomen and pelvis was performed to exclude primary or secondary malignancy. The uterus and GI tract were evaluated and no malignancy was found¹⁴ but in the chest a soft tissue, nodose formation was seen in the left lung. Metastatic disease was suspected.

The patient was admitted to hospital and had an open biopsy of the lesion. Histological examination, including immunohistochemical studies, determines the precise diagnosis of primary leiomyosarcoma. Wide resection of the tumour was planned. A proximal femoral resection



Fig. 1. Radiograph of the left hip with a poorly defined osteolytic mass and suspected bone destruction in the upper cortex of femoral neck.



Fig. 3. Proximal femoral resection with wide surgical margins reconstructed with composite allograft endoprosthesis reconstruction.

with wide surgical margins was reconstructed with composite allograft prosthesis reconstruction (Figure 3).

Two weeks after the orthopaedic procedure the patient was suitable for the thoracic surgery treatment for the lung tumour. The right lung was treated operatively by lobectomy of the affected lobus and confirmed to be a leiomyosarcoma metastasis by pathohistological analysis.

Five cycles of adjuvant chemotherapy was performed according to ifosfamide + epirubicin chemotherapy regimen¹⁵. The patient has been checked up every three months.

One year after the surgery the patient was very satisfied: She was self-sufficient and the hip was painless.



Fig. 2. Magnetic resonance reveals large intraosseous component with the tumour occupying the femoral neck and extending into the femoral metaphysis to the level of lesser trochanter and femoral head without extension to surrounding soft tissue.

After two years the patient went to our hospital with signs of gradually growing pain in the left hip and night pain. After ultrasound and radiographic examination the local recurrence was suspected and biopsy proven. The patient died after two months for pulmonal metastases.

Discussion

In the 19th century, amputation was the primary choice for surgical treatment in musculoskeletal oncology¹³. After introduction of endoprosthesis¹⁶, allografts^{17,18} and vascularised autogenous bone grafts¹⁹ in reconstruction after surgery for musculoskeletal tumours, limb saving surgery was born.

The aim of limb-saving surgery should not only pursue a good oncological and functional result but also a good psychological outcome¹³. Limb-saving surgery was firstly attempted on the thigh, upper part of the tibia, and distal part of the femur¹⁶. There are general indications for the limb salvage: tumours of the extremities, axial skeleton, or both, in which optimum surgical margins are attainable, soft-tissue extension is moderate, the neurovascular bundles are not compromised, and metastases are absent or responsive to curative treatment¹³. In other cases the amputation is the procedure of choice. Catton et al.²⁰ agrees that despite the presence of metastasis the limb salvage procedures are indicated.

All diagnostic procedures on our patient demonstrated primary femoral and solitary metastatic lung intracompartmental tumours (Enneking's stage III a), without other malignancy localizations. Despite the presence of the solitary lung metastasis, we decided to treat the patient with limb salvage procedure and lung metastasectomy, because of the localization of the tumour and very demanding rehabilitation procedure after hip disarticulation. Indeed, pulmonary metastasectomy in soft

tissue sarcoma (STS) can lead to long-term survival and good prognosis even in cases of repeated lung metastasis²¹.

It is known that optimum surgical margins are 3 cm of normal bone and 1 cm of normal soft tissues around a malignant bone¹³. Furthermore, there are several ways to reconstruct the operated limb. Reconstructions can be achieved with allograft bone, mega-endoprosthetic replacements or composites, or with vascularised bone grafts. Taking these facts into consideration and after accurate preoperative planning, we performed a proximal femoral resection three centimetres under the lesser trochanter to obtain wide margin, and we reconstructed the defect with composite allograft prosthesis. Naturally, the quality of life in patients with bone tumours is affected both by limb-saving methods and by amputation^{22,23}.

However, 75% of amputees report difficulties in sexual relationships, whereas hardly any limb-salvage patients do²⁴. Also, the same percentage of amputated patients feel embarrassed to show their prostheses, and they refrain from certain social activities²⁴.

To assess the functional outcome we used the scoring system of the Musculoskeletal Tumor Society (MTS)²⁵. The MTS is based on six categories: pain, function, emotional acceptance, use of supports, ability to walk, and gait. Each category was assigned a numerical value from 0 to 5 points, with 5 points indicating the best function. The values for each category were added, and the functional score was presented as a percentage of the maximum possible score (30 points). The Musculoskeletal Tumor Society functional score at the time of the follow-up,

one year after the surgery was 4 for pain, 2 for function, 4 for emotional acceptance, 1 for use of supports, 3 for the ability to walk and 2 for gait – MTS was 53%. Herein, it is important to point out that the patient was personally satisfied with operative procedure.

Local relapse of extremity STS after seemingly adequate combined conservative therapy is uncommon, since it is potentially devastating for the patient and is extremely frustrating for the physician, who has only limited data on which to determine the most appropriate course of action. When local relapse occurs the tendency has been to recommend limb amputation for local relapse, partly because in this setting local recurrence may not be amenable to a conservative resection, but also because of uncertainty about the efficacy of conservative surgery when used alone^{20,26}. The available data for the most appropriate salvage therapy in cases of tumour relapses is limited. In our case the patient was unsuitable for reoperation and he died soon after the tumour relapse.

Our case suggests that primary leiomyosarcoma has to be considered as differential diagnostic possibility in case of bone tumours seen on routine initial plain radiographs as lytic lesions. Despite moderate result in MTS and short survivorship of our patient, we suggest that limb salvage procedure is indicated in case of primary bone leiomyosarcoma even in case of present metastasis due to very good psychological outcome.

The results of limb salvage should be judged in terms of oncology, function, and especially quality of life for so aggressive and rare tumours like primary bone leiomyosarcoma.

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KIRURGIJA SPAŠAVANJA EKSTREMITETA KOD LEIOMIOSARKOMA STUPNJA III A PREMA ENNEKING-U

S A Ž E T A K

Današnja je tendencija u liječenju muskuloskeletnih tumora kirurgija spašavanja ekstremiteta. Čuvanjem ekstremiteta postićemo ne samo dobar onkološki i funkcionalni rezultat već i dobar psihološki rezultat. Cilj našeg rada bio je liječenje vrlo rijetkog slučaja uznapredovalog leiomiosarkoma sa prisutnom plućnom metastazom, pomoću konzervativne kirurške procedure.