*Case report*

# OSSEOUS INVASION IN ALVEOLAR SARCOMA OF SOFT TISSUES: A HEALING APPROACH AND LIMB-SALVAGING SURGERY

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## Summary

The alveolar soft part sarcoma (ASPS) is a rare tumor, affecting mainly young age. It appears more often between 15 and 35 years of age, and rarely before 5 or after 50 years of age. It is found most often in the limbs (in the thigh). We present a case of a 47-year-old woman with a primary diagnosis of ASST made after initial open biopsy along the front surface of the right thigh. Pre-operative (PO) examinations included conventional radiography (X-ray), computer axial tomography (CT), magnetic resonance imaging (MRI), bone scan (BS) and biplane angiography (BA) to determine the extent, location, invasion towards the bone and the proximal joint, as well as the extent of femoral osteostomy. Without involving superficial femoral artery (a FS) and preserving patency, a wide en block bone-soft-tissue resection was made with removal of the entire front muscular group and knee joint and subtotal resection of the femoral diaphysis 13 cm below the major trochanter. Subsequently, reconstruction of the defect was carried out, and recovery of limb function achieved.

**Key words:** alveolar soft part sarcoma (ASPS), limb-salvaging operation, osseous invasion, tumor, endoprosthesis.

## Introduction

The alveolar soft part sarcoma (ASPS) is a rare tumor of soft tissues (incidence of 0.5 % - 0.9 %), most often affecting the deep tissues of the thigh [1, 2, 3]. It is characterized by slow growth and local recurrences after total removal, and is highly metastatic [4, 5]. Metastases can be detected at an early stage, during the very process of diagnosing, even before detecting the primary tumor, or very late after its removal, despite the lack of recurrence [1, 5].

Survival rates in patients without metastases are as follows: 5 years (60 %), 10 years (38 %) and 20 years (15 %). Basic prognostic factors are age, degree of malignancy, size of the tumor, presence of metastases at diagnosing [1, 4, 6]. Most commonly, metastases occur in the lung, bones and brain. Metastases in lymph nodes are rare [5, 7].

The method of treatment of soft tumors of the limbs is amputation or wide resection – removal of

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the tumor together with the surrounding reactive zone and part of the normal tissue, preserving the limb and its function (limb-salvaging operation) [8, 9]. Surgery is often combined with pre- and postoperative chemo- and radiotherapy [3, 6]. Tumor recurrences rates after this combined approach are comparable to those, following amputation [1, 2, 6]. Effective treatment of such tumors with osseous invasion necessitates bone resection en- block with the tumor, followed by reconstruction [5, 8, 9, 10]. The precise assessment of the extent, location and depth of invasion into the adjacent structures is carried out by means of X-ray, CT, and MRI. MRI, bone scan and BA are of particularly importance in planning the intervention in terms of approach, resection level, size and type of prosthesis, and chances for vascular prosthesis application and reconstruction of defects.

**Fig. 1.** Outline of skin flaps surrounding the tumor (12 – 18 cm) involving the front surface of right thigh. It is visible that the outline contains the preceding biopsy. The approach allows for access to the pelvic structures, as well as structures under knee joint.

## Case Description

We present a 47-year-old woman presenting with a swelling in the middle third of the right thigh. She was referred for examination in September 2005 because the swelling, first seen two years before, had enlarged and become painful. Examination revealed a palpable tumor mass measuring 10 by 15 cm (Fig. 1). The mass was located deep along the front thigh surface, firm and immobile, with highly widened veins subcutaneously. MRI investigation obviated an intramurally located tumor formation in the region of the lateral vastus muscle and medial vastus muscle measuring 5 by 7 cm at coronary view, which had caused a lytic lesion in the femoral bone cortex. MRI did not show involvement of the medular canal. CT scan revealed hyperaemic tumor formations in the region of the middle and distal third of the thigh. In view of the presenting complaints and image diagnostic findings, open biopsy was done which revealed ASPS. Immunochemistry revealed Desmin (+), Myo D1 (+) cell groups. After 45 days, a CT scan of lungs and arteriography of the small pelvis and right lower limb was performed. She was referred to the national Oncology Center in Sofia for pre-operative chemotherapy. MRI and CT investigations in February 2006 revealed osseous invasion of the tumor in the medullary canal as well (fig. 2, fig. 3 and fig. 4) and increase in tumor size to 12 by 18 cm. Bone scan obviated a new focus located in the patella. The tumor was classified as G-2 T-2 N-0 [6, 11]. On March 16, 2006 a wide resection was performed with removal of the entire front muscle group with the appertaining skin and under-skin after an intraoperative gefrir examination of the tumor borders. The material was collected from the fascia of deep head of the femoral rectus muscle, sartorius muscle and a lymph node (Fig. 5). After a proximal osteostomy and extra-articular resection of the knee joint, the tumor was removed en block (Fig. 6). Reconstruction of the osseous defect was performed by means of a prosthesis and transposition of the lateral head of the femoral biceps to the distal part of the sartorius muscle (Fig. 7). Five months after surgery, physical rehabilitation was started, during which a peri-prostethic infection occurred and the operative wound reopened. Revision of the prosthesis was performed, followed by lavage and drainage. The infection was managed, and treatment continued with aseptic bandages to enhance secondary granulation. Removal of the prosthesis and external fixation were considered, as well as a second re-operation for placement of an intramedulary nail and soft-tissue flap from the gastrochemius muscle, or a flap from another part of the body with accomplishment of microvascular anastomoses. In view of the risk of a vascular incident (the sole source of being the superficial femoral artery), the wound was left open for secondary granulation.

After 42 months, the patient was free from metastases and tumor recurrence (fig. 8, fig. 9).

**Fig. 2.** CT finding – presence of tumor formations, located intramurally in vastus lateralis muscle and vastus intermedius, and a lytic lesion in the zone of the femoral cortex.

**Fig. 3**. Obvious osseous invasion with involvement of the medullary canal and two lytic foci, including the femoral shaft.

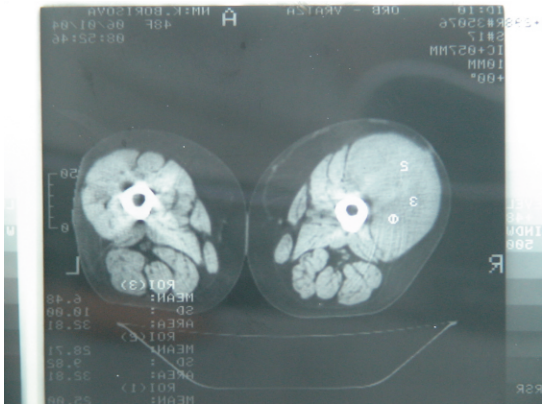

**Fig. 4.** CT finding: the tumor involves the entire front muscular group (quadriceps fem.). Lytic lesion of the bone.

**Fig. 5** Intraoperative removal of the entire front muscular group with preservation of patency of superficial femoral artery.

**Fig. 6.** Macroscopic material after total excision of quadriceps femoral muscle, with the involved femoral shaft and knee joint.

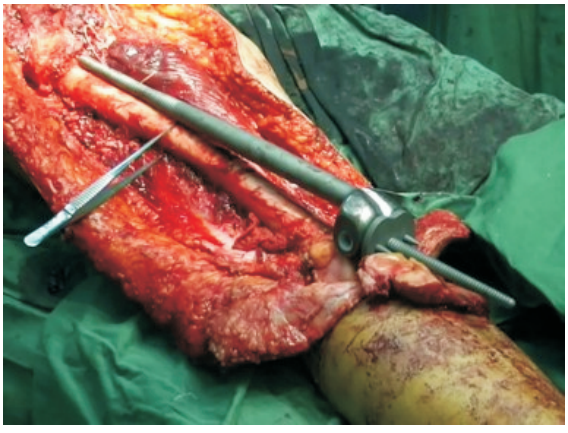

**Fig. 7.** Intraoperative verification of predominant level of femoral osteotomy and incorporation of the tailored tumor endoprosthesis.

**Fig. 8.** Forty-two months after the operation. Lack of data for loosening of endoprosthesis.

**Fig. 9.** Forty-two months after the operation, the patient is free from metastases and tumor recurrences.

## Discussion

Osseous invasion, without metastases in lungs, is a rare defect of the soft-tissue limb sarcomas. ASPS is a very rare tumor in the deep tissues of the thigh. The adequate behaviour in case of bone invasion is removal of the tumor with the involved adjacent bone or joint and recovery of the defect, while preserving the limb function (a limb-salvaging operation).

The role of MRI, CT and BA in the pre-operative period is particularly important in regard to tumor size, location and invasion, as well as planning the surgical approach by an interdisciplinary team.

The estimation of the superficial femoral artery and the level of osteostomy is also crucial. In case no data of invasion towards a rear muscular group, it is necessary to cover the front part of the prosthesis and recover the extensor mechanism of the limb [9, 10].

Good soft-tissue coverage and recovery of limb function are of major importance in the prevention of decubital ulcers in the of skin-underskin flaps, because ulcers may lead to subsequent surfacing of the prosthesis and a chronic infection.

The development of microsurgery and high-technology orthopedics allowed for the performance of limb-salvaging operations. Hence, the survival rates are enhanced, as well as, the possibility for immediate post-operative chemotherapy. We consider that in such cases, the limb-salvaging operation is a method of choice.

## Reference


1. Elias DA, White LM, Simpson DJ, Kandel RA, Tomlinson G, Robert S. Bell RS, Wunder JJ. MR Assessment of Osseous Invasion by Sarcoma. Radiology. 2003;229(1):145-51.
2. Panice K, Go SD, Healey JH, Leung DH, Lewis JJ. Soft-tissue sarcomas involving bone or neurovascular structures: MR imaging prognostic factors. Radiology. 1997;205:871-5.
3. Eiber FR, Morton D. Advance in the treatment of sarcomas of the extremity. Current status of limb salvage. Cancer 1984;54:2695-701.
4. Peabody TD, Simon MA. Principles of staging of soft-tissue sarcomas.Clinical Orthop. 1993;289;19-31.
5. Bell RS, O'Sullivan B, Liu F.The surgical margin in soft-tissue sarcoma. JBJS Am. 1989;71:370-5.
6. Davidson T, Cooke J, Parsons C, Westbury G. Pre-operative assessment of soft-tissue sarcomas by computed tomography. Br J Surgery 1987;74:474-8.
7. Tomori Nakamura, Katsuyuki Kusuraki, Masasiti Seto, Akihiko Matsumine and Atsumasa Uchida. Case Report: Recurrence of soft-tissue MFH in bone due to minute intravenous tumor emboli detected by MRI. Oncology Reports. 2003;10:1957-60.
8. Malawer MM. Distal Femoral Resection with Endoprosthetic Reconstruction. Musculoskeletal Cancer Surgery. Chapter 30: 457-480.

9. Malawer MM, Jacob Bickels, Isaac Meller, Robert Henshaw. Proximal Total Femur Resection with Endoprosthetic Reconstruction. Chapter 29: 437-455.
10. Malawer MM and Paul Sugarbaker: Quadriceps Muscle Group Excision. Musculoskeletal Cancer Surgery. Chapter 14: 254-64.
11. Enneking WF, Spanier SS, Goodman MA. A system for the surgical staging of musculoskeletal sarcoma. Clinical Orthop. 1980;153:106-120.