

# Challenging Limb Salvage in a Crural Malignant Peripheral Nerve Sheath Tumor with Pre-existing Long-Stem Implants: A Case Report

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## Learning Point of the Article:

This case underscores the importance of including rare musculoskeletal tumors in the differential diagnosis of atypical orthopaedic lesions and demonstrates that accurate imaging, histopathological confirmation, and appropriate surgical management are essential for achieving optimal oncologic and functional outcomes.

## Abstract

**Introduction:** Malignant peripheral nerve sheath tumors (MPNST) are uncommon soft-tissue sarcomas that typically arise in proximal segments of the extremities or pelvis. Their management requires precise biopsy planning, careful surgical assessment, and coordinated multidisciplinary care. Tumors involving multiple longitudinal levels of the leg are rare and pose significant challenges for limb salvage.

**Case Report:** We report the case of a 79-year-old male presenting with a rapidly progressive mass extending along the posterior cruris, in the setting of a prior hip hemiarthroplasty and a revision total knee arthroplasty with long femoral and tibial stems. Magnetic resonance imaging demonstrated two heterogeneous solid masses in the posterolateral proximal right cruris and biopsy revealed a high-grade MPNST (Fédération Nationale des Centres de Lutte Contre le Cancer grade 3). The tumor board concluded that limb-sparing resection was unsafe due to tumor extent, limited soft-tissue envelope, and anticipated difficulty achieving durable wound closure. The presence of existing long-stem implants further complicated determination of a safe osteotomy level. Above-knee amputation was therefore selected as the most reliable method for achieving oncologic control while minimizing post-operative complications.

**Discussion/Results:** The amputation was performed with fluoroscopy-guided planning of the osteotomy level to avoid stem interference, followed by myoplasty and myodesis for stable stump reconstruction. Histopathology confirmed a high-grade MPNST with diffuse S100 and SOX10 positivity, extensive necrosis, high mitotic activity, and a Ki-67 index of 50%.

**Conclusion:** This case illustrates the critical importance of methodical biopsy planning and multidisciplinary coordination in managing sarcomas of the lower extremity. The unusual longitudinal distribution of the tumor, combined with the constraints imposed by pre-existing long-stem implants, made limb salvage infeasible. Individualized surgical planning, incorporating both oncologic and reconstructive considerations, is essential when managing complex MPNSTs in reconstructively compromised limbs.

**Keywords:** Malignant peripheral nerve sheath tumor, limb salvage, above-knee amputation, lower extremity sarcoma, revision arthroplasty implants.

## Introduction

Malignant peripheral nerve sheath tumor (MPNST) represents the sixth most common subtype of soft-tissue sarcomas,

accounting for roughly 5–10% of all cases [1, 2]. Although its precise histogenetic origin is still uncertain, most tumors arise in association with peripheral nerves and are believed to derive

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## Author's Photo Gallery



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from neural crest-related cells [3]. Approximately half of all MPNSTs occur sporadically, while the remaining 50% develop in individuals with neurofibromatosis type 1 [4].

Clinically, MPNST typically occurs in a manner similar to other soft-tissue sarcomas, often presenting as a progressively enlarging mass over months or years. These tumors most frequently originate near major nerve bundles of the extremities and pelvis—such as the sciatic nerve, brachial plexus, and sacral plexus—resulting most commonly in the proximal upper and lower limbs [5]. Common symptoms include pain, paresthesia, and may include various neurologic deficits.

For localized high-grade MPNST, the current standard of care involves surgical resection followed by adjuvant radiotherapy. Despite aggressive treatment, local recurrence occurs in approximately 40–65% of patients, and metastatic spread develops in 30–60%, most commonly to the lungs [6]. Overall, MPNST carries a poor prognosis, with reported 5-year survival rates ranging from 20% to 50% and mortality reaching up to 75% in high-grade cases [7].

### Case Report

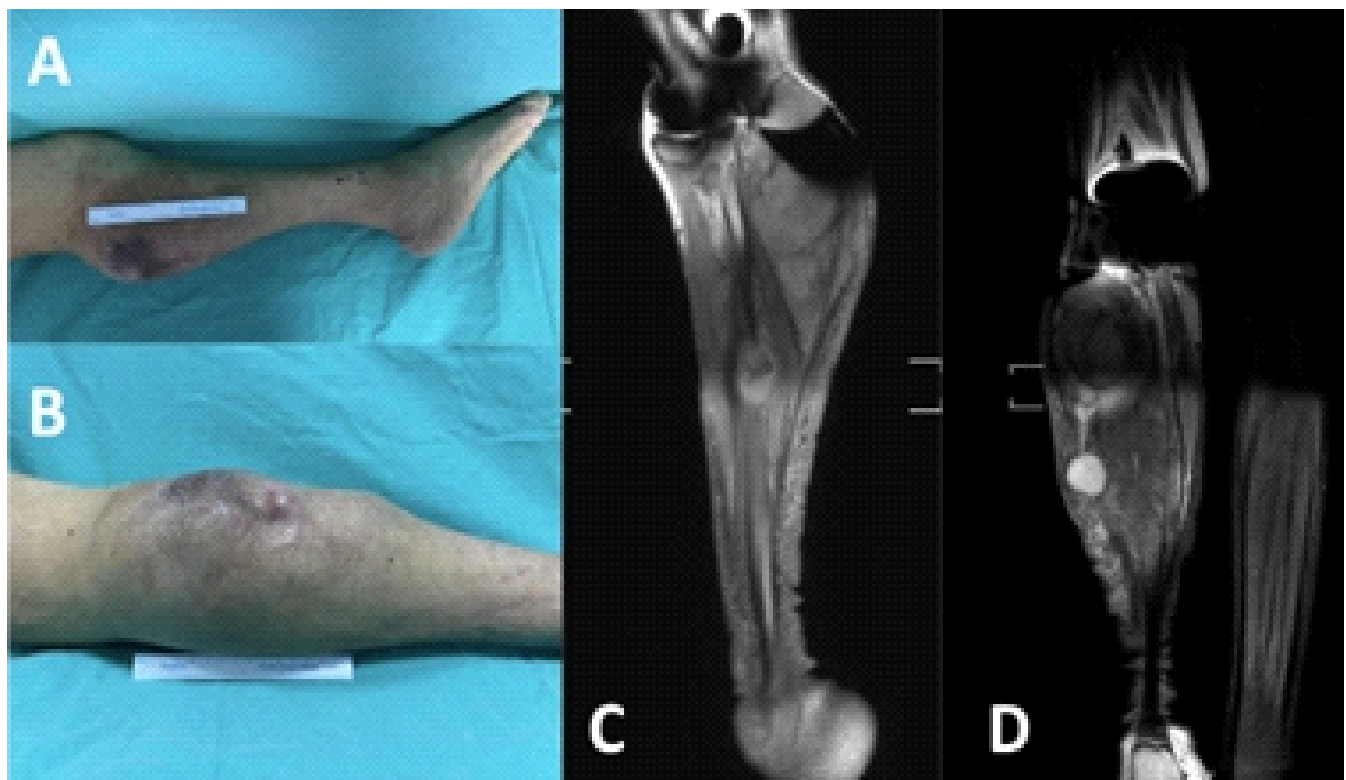
A 79-year-old male with no known comorbidities presented with a 2-year history of a progressive soft-tissue mass located along the posterior aspect of the right leg, extending from the

distal popliteal region to the mid-crusis. A previous incision scar was noted over the lesion, reportedly from an earlier biopsy, although neither the patient nor his relatives could provide reliable information regarding the prior procedure. The initial biopsy performed at an outside center had been reported as incomplete.

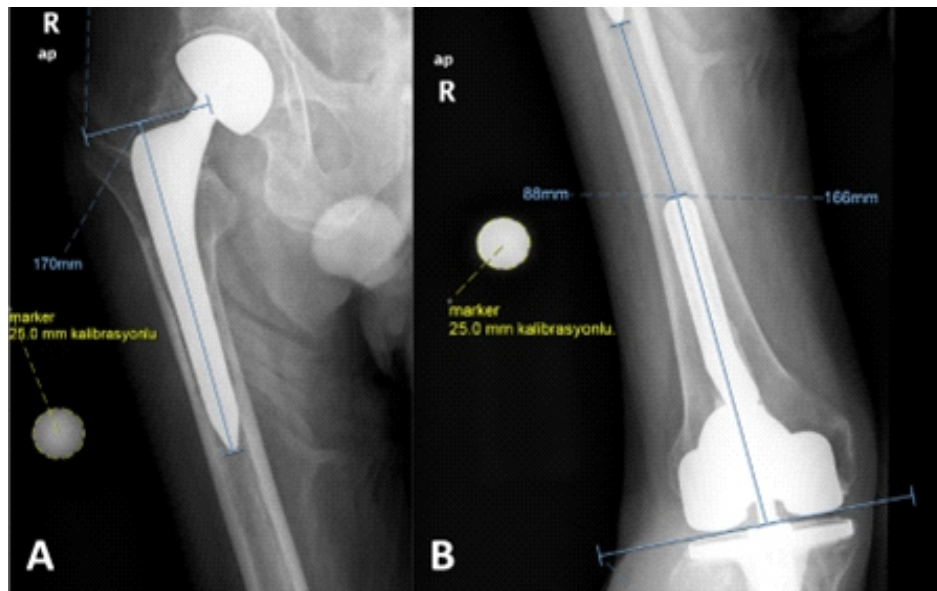
The patient had undergone a revision total knee arthroplasty with long femoral and tibial stems at another institution approximately 2 years earlier. He reported that the mass had increased significantly in size over the past year.

Initial radiographs demonstrated soft-tissue swelling without clearly identifiable calcifications. The patient was referred for anesthesia evaluation for a repeat biopsy. Pre-operative magnetic resonance imaging demonstrated two heterogenous solid masses in the posterolateral proximal right crusis: A larger lesion measuring approximately  $90 \times 77 \times 105$  mm with central cystic/necrotic, non-enhancing areas, and a smaller adjacent caudal lesion measuring approximately  $40 \times 38 \times 42$  mm (Fig. 1). Core biopsy results were consistent with a high-grade MPNST. Clinically, he reported progressive pain, decreased appetite, and weight loss. positron emission tomography-computed tomography (PET-CT) was recommended for staging.

The case was discussed in a multidisciplinary tumor board, where palliative radiotherapy was initially considered.



**Figure 1:** Pre-operative evaluation of the right crusis showing lateral [a] and posterior [b] clinical photographs, coronal magnetic resonance imaging (MRI) demonstrating both lesions [c], and sagittal MRI showing their longitudinal extent [d].



**Figure 2:** Pre-operative radiographs showing the hip anteroposterior (AP) view with the hemiarthroplasty stem [A] and the knee AP view with the revision type long femoral and tibial stems [B], including measured distances from the greater trochanter and knee joint line to the respective stem tips.

and closure. Following standard preparation, an anterior incision was created and extended circumferentially to form anterior and posterior flaps. Dissection proceeded through the subcutaneous tissues to the medial aspect of the thigh, where the adductor canal was identified. The femoral artery and vein were isolated and ligated proximally and distally. The saphenous nerve was divided sharply at a proximal level.

Dissection continued posteriorly to expose the sciatic nerve, which was transected in a single sharp cut at a proximal location. The deep femoral vessels were identified and ligated in a similar fashion. After completing circumferential mobilization, the femur was osteotomized at the pre-determined mid-femoral level using a

Gigli saw, safely distal to both pre-existing femoral stems. The bone edges were smoothed.

The stump was then reconstructed with adductor myoplasty through drill holes in the femur, followed by myodesis of the anterior and posterior muscle compartments to provide stable muscular coverage. The anterior and posterior flaps were approximated with fascial sutures, and the wound was closed in a layered fashion without tension. A single closed-suction drain was placed, and the stump was dressed and compressed with an elastic bandage. The amputated specimen was submitted for pathological examination.

Following PET-CT evaluation, the tumor board concluded that limb-sparing surgery would not be feasible due to the large tumor size, aggressive pathology, and expected soft-tissue deficiency in a limb previously reconstructed with long-stem implants. An above-knee amputation was recommended.

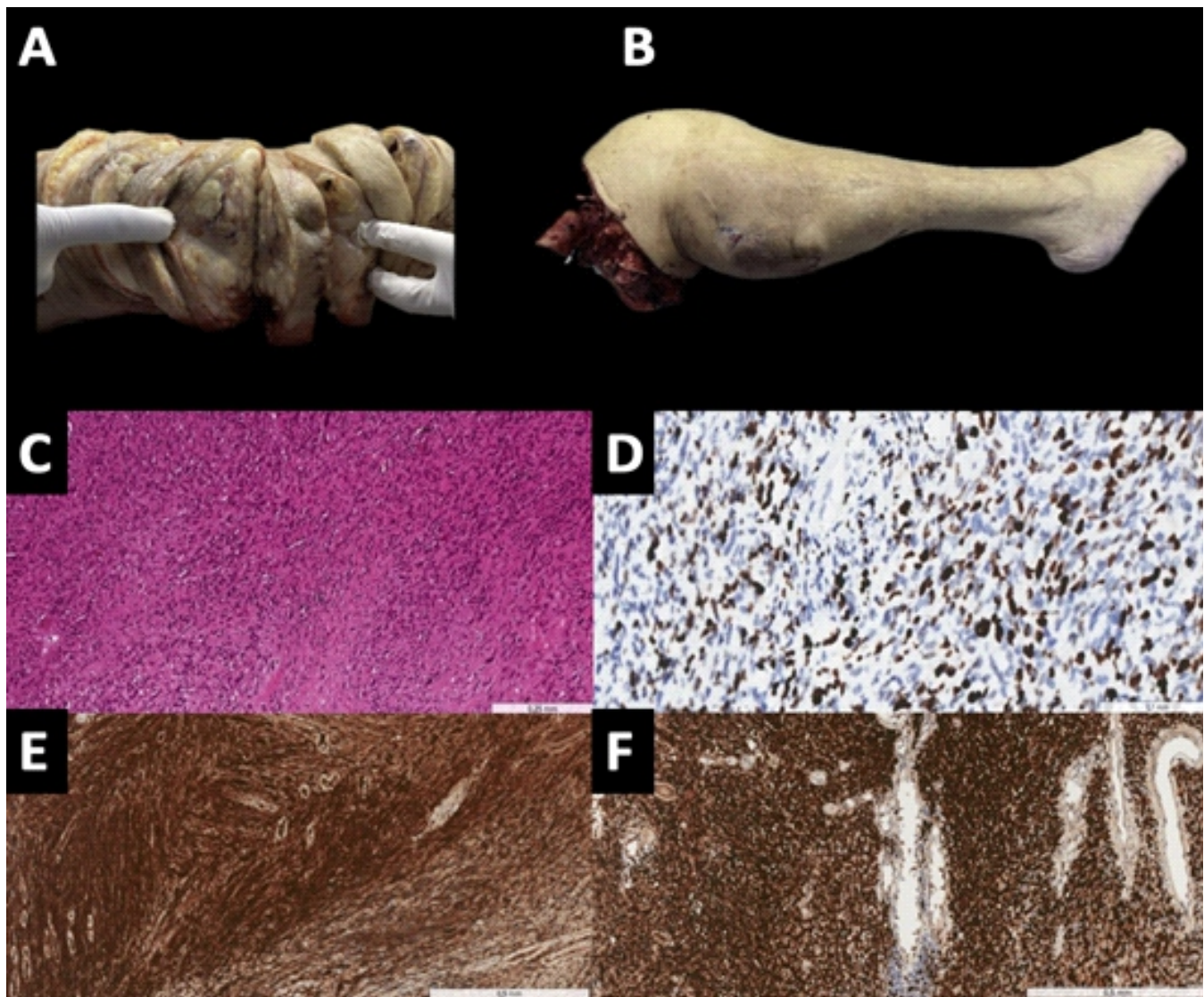
The patient and his family were informed in detail about the planned procedure, including the possibility of a high-risk post-operative course and the need for intensive care monitoring. He was subsequently referred to anesthesia for pre-operative assessment.

### Surgical procedure

An above-knee amputation was planned after multidisciplinary evaluation due to the size and extent of the high-grade MPNST and the anticipated inability to achieve durable soft-tissue coverage with limb-sparing options. Pre-operative imaging confirmed the presence of a hip hemiarthroplasty stem and a long revision femoral stem from the patient's prior total knee arthroplasty. Pre-operative X-ray demonstrated that the tibial revision stem extended approximately 170 mm proximal to the knee joint line, while the tip of the femoral stem was located roughly 170 mm distal to the greater trochanter. These measurements confirmed that both implants occupied substantial portions of the femur and tibia. The planned transfemoral amputation level was positioned approximately 1 cm proximal to the proximal end of the tibial stem, thereby avoiding stem interference while preserving an adequate residual femoral length for prosthetic fitting (Fig. 2). A fish-mouth configuration was selected to optimize stump contour

### Pathology and histology

Final pathology from multiple incisional and Tru-cut biopsies of the proximal and distal crural masses demonstrated a high-grade MPNST. All specimens showed a densely cellular spindle-cell neoplasm composed of intersecting fascicles of atypical, elongated, and occasionally serpentine cells with coarse chromatin and scant cytoplasm. The tumor exhibited a mixed diffuse and nodular growth pattern with alternating hypercellular and hypocellular areas. Geographic tumor necrosis was prominent, involving approximately 50% of the sampled tissue. Frequent mitotic figures (10–19 mitoses/10 high-power fields) were identified. Multinucleated tumor cell groups were also present. According to the Fédération Nationale des Centres de Lutte Contre le Cancer system, the tumor demonstrated a differentiation score of 3, mitotic score of 2, and necrosis score of 2, corresponding to an overall grade 3



**Figure 3:** Macroscopic appearance of the tumor, measuring  $19 \times 11 \times 7$  cm [A], macroscopic view of the above-knee amputation specimen, measuring  $66 \times 17 \times 13$  cm [B], malignant mesenchymal tumor composed of spindle cells arranged in a fascicular pattern, with irregular bundles and short storiform areas (Hematoxylin and Eosin,  $\times 15$ ) [C], Ki-67 immunostaining demonstrating strong nuclear positivity in tumor cells, with a proliferation index of 50% (Ki-67,  $\times 40$ ) [D], diffuse nuclear and cytoplasmic positivity in tumor cells (S-100,  $\times 10$ ) [E], diffuse nuclear positivity in tumor cells with SOX10 staining (SOX10,  $\times 10$ ) [F].

sarcoma (a total score of 7).

Immunohistochemically, the tumor showed strong diffuse positivity for S100 and SOX10, supporting neural crest-related differentiation. Additional positive markers included CD34, CD56, CD99, collagen IV, FXIIIa, and focal epithelial membrane antigen. The Ki-67 proliferation index was markedly elevated at approximately 50%, and p53 expression was detected in approximately 70% of tumor nuclei (Fig. 3). Markers for skeletal muscle differentiation (actin, caldesmon), mesothelial differentiation (calretinin), cytokeratin (pan-CK), and others were negative.

Taken together, the morphologic features and immunohistochemical profile confirmed the diagnosis of high-

grade MPNST according to the 2020 World Health Organization classification.

### Discussion

MPNSTs typically arise in the proximal segments of the extremities and pelvis, often in association with major nerve trunks, such as the sciatic or brachial plexus [8]. In contrast, the present case demonstrated an uncommon pattern of multifocal involvement along the cruris, with lesions identified both proximally and distally. This longitudinal distribution significantly complicated attempts at limb-sparing resection. Standard treatment for high-grade MPNST includes wide excision with negative margins followed by adjuvant

radiotherapy, with chemotherapy considered in selected cases [9]. However, in this patient, radical resection posed substantial risks: The expected inability to achieve durable wound closure, the limited soft-tissue envelope, and the consensus opinion of the orthopedic, plastic surgery, and oncology teams led to above-knee amputation as the safer option. The presence of a hip hemiarthroplasty stem and a long-stem revision femoral component introduced additional challenges, as the osteotomy level had to be carefully selected to avoid violating existing implants while still achieving oncologic control. This constraint also influenced the planning of soft-tissue coverage and stump stability. Furthermore, even if limb-sparing excision had been attempted, the tibial revision stem and prior surgical field would have created a high-risk environment for post-operative infection, potentially compromising both oncologic and functional outcomes [10]. Taken together, these factors supported amputation as the most appropriate management strategy to ensure reliable disease control and minimize complications in this anatomically complex and surgically constrained limb [11].

### Conclusion

This case highlights the importance of a methodical and

**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Conflict of interest:** Nil    **Source of support:** None

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### Clinical Message

Complex MPNSTs in limbs containing long-stem implants require meticulous planning to determine safe biopsy routes, resection margins, and amputation levels. In select patients, amputation may offer a safer and more reliable outcome than limb salvage.

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