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*SNI: Spine*

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*Case Report*

# Spinal cord compression from cutaneous malignant peripheral nerve sheath tumor metastasis in a patient with neurofibromatosis Type 1

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## ABSTRACT

**Background:** Neurofibromatosis Type 1 (NF-1) and previous irradiation are two common risk factors that can result in malignant peripheral nerve sheath tumors (MPNSTs), extremely rare soft-tissue sarcomas. Here, a 63-year-old male with NF-1 presented with diffuse spinal metastases from a subcutaneous MPNST.

**Case Description:** A 63-year-old male with NF-1 presented acutely with paraplegia and urinary incontinence. Both CT and MR studies of the thoracic-lumbosacral spine showed multiple metastases from a subcutaneous MPNST. In addition, the patient had a T12 vertebral body pathological fracture.

**Conclusion:** Despite its aggressive behavior, some cases of MPNST can be managed with gross total resection and adjuvant radiotherapy. In addition, in the presence of multiple metastases, chemotherapy may play an additional, although questionable role.

**Keywords:** Gross total removal, Metastasis, Neurofibromatosis, Paraplegia, Spine

## INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are rare malignant mesenchymal tumors that are found in approximately 1/100.000/year in the general population. For patients with neurofibromatosis Type 1 (NF-1), about one of ten patients will develop this tumor during their lifetime.[1] MPNSTs are commonly localized in extremities, where the optimal treatment is radical tumor resection.[8] When and where feasible, the preference is for a gross total tumor removal gross total resection (GTR) with microscopically negative margins, followed by adjuvant radiotherapy (RT); the role of chemotherapy is still being debated.[4,7,8] Here, the authors present a rare and complex case of diffuse spinal metastasis from a subcutaneous MPNST in a 63-year-old patient with NF-1.

## CASE DESCRIPTION

A 63-year-old male patient with NF-1 presented with a 2-week history of low back pain and progressive lower extremity weakness. On admission, he was paraplegic with acute urinary retention (i.e., 0/5 lower extremity function with diffuse hyperreflexia, clonus, and bilateral Babinski signs).

### Diagnostic studies: MR and CT

He underwent a lumbosacral MRI that showed a T12 burst pathological fracture with severe spinal cord compression. Findings included: a thick epidural mass, a large subcutaneous ovalar lesion near the L3 and L4 spinous processes (54 × 60 × 39 mm) that was inhomogeneous in pattern on T1/T2-weighted images and STIR sequences [Figure 1]. The patient underwent an urgent T12 decompressive laminectomy with T10-T11-L1 fixation using transpedicle titanium screws. Intraoperatively, the dura mater was markedly compressed by a circumferential, vascularized, and reddish lesion that was posteriorly removed and sent for histological evaluation. The cutaneous/subcutaneous lesion was also excised *en bloc* [Figure 2]. The postoperative total body CT scan showed no evidence of other primitive lesions.

### Histology

The histological samples documented a MPNST with spinal metastasis. The high mitotic index was confirmed by the presence of multiple necrotic areas and a 1 cm sarcomatous focus. A small region of atypical neurofibroma was also shown, proving the evolution of the malignant lesion from an original benign neurofibroma.

### Postoperative recovery

Postoperatively, the patient regained partial motor function (BMRC 3/5) and urinary continence.

## DISCUSSION

### Frequency and etiology of MPNST

MPNSTs are rare tumors, affecting about 10% of patients with NF-1.[8] The previous RT is a well-known risk factor for developing these lesions.[7] They occur with different frequency that is different body locations. The most common locations include the proximal extremities and pelvis.[1,8]

### Treatment strategies

The optimal treatment for MPNST is a GTR with microscopically negative surgical margins followed by adjuvant RT. RT is useful both in reducing the risk of recurrence and in improving local control if GTR is not feasible.[1,6] The 5-year overall survival rate for patients undergoing GTR with clear margins is 67%, instead 22% for surgery without such clear margins. The median survival in patients who have undergone GTR is around 4 years after surgery.[7,8]

### Role of RT for MPNST

The role of RT in preventing early recurrence is well established. In the literature, there are two cases of patients

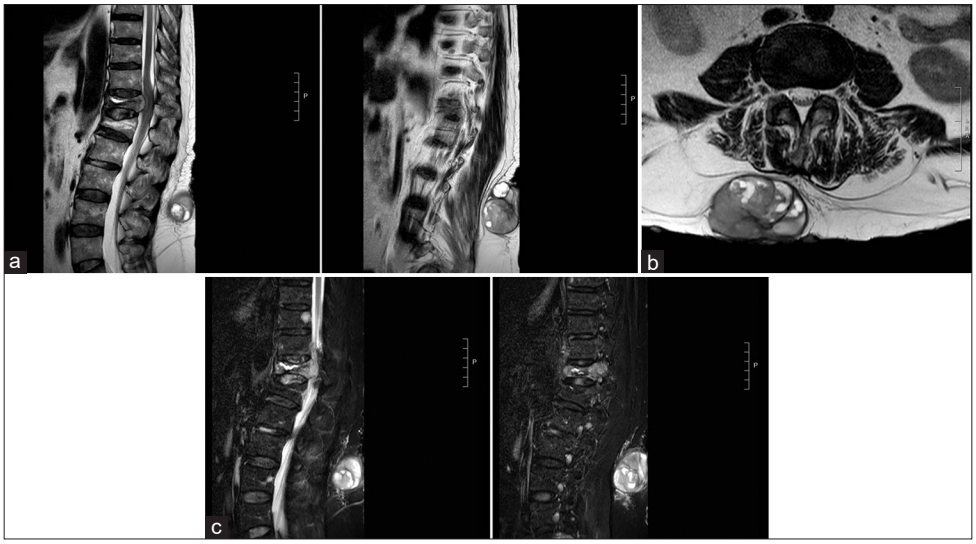


**Figure 1:** Preoperative thoracolumbosacral MRI T2-weighted sagittal (a) and axial (b) images documented a T12 burst pathological fracture with severe spinal cord compression by a thick epidural tissue and a large subcutaneous paraspinal ovalar lesion (54 × 60 × 39 mm). These lesions appear inhomogeneously hyperintense on STIR sequences (c).

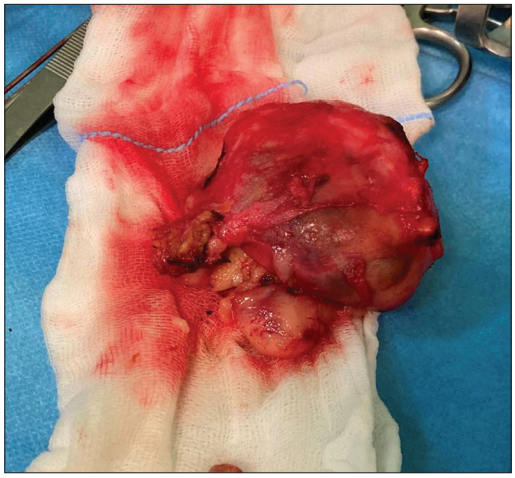

**Figure 2:** Intraoperative image showed *en bloc* removal of the cutaneous/subcutaneous Malignant Peripheral Nerve Sheath Tumors.

affected by MPNST treated with GTR without adjuvant RT. These patients both presented with early recurrences, 2 months and 6 months, respectively, after surgery.[2,3] According to recent studies, the recurrence rate varies from 40% in patients treated with adjuvant RT up to 68% in those managed without adjuvant RT.[2]

### Prognostic factors

Several prognostic factors have been analyzed in the literature. These include tumor size, tumor grade and histology, patients with NF-1, previous radiation therapy, and surgical clean margins. A recent paper by Yan *et al*. in selected patients diagnosed with MPNST from the surveillance, epidemiology, and end results database showed that age, histology, grading, and chemotherapy were independent prognostic factors for OS, while the location of primitive lesion, GTR, grading, and chemotherapy are prognostic factors for cause-specific survival.[5,9]

### Locally aggressive tumors and surgical resection considerations

MPNSTs are aggressive tumors, with tendency of local recurrence and hematogenous spread to lymph nodes, lung, liver, and rarely bones. Intradural presentation is rare. There is an increased risk of the central nervous system metastasis from either paraspinal or subcutaneous lesions.[1,8]

With spinal lesions, GTR is challenging. In these cases, one must strike a balance between GTR and the potential risk of creating neurological deficits. As shown in a recent case, a patient underwent a total sacrectomy with severe postoperative deficits compromising his life.[4,9]

Moreover, spinal MPNSTs have a doubled risk of metastasis if when compared to other locations.[4]

Considering the risk-benefit balance, in paraspinal lesion surgeons should remove the maximal percentage of the tumor to minimize the risk of neurological deficits.[8]

### Marginal utility of chemotherapy

Chemotherapy plays a marginal role in the treatment of MPNST. It is typically only used as a salvage therapy in patients with multiple metastases where neither surgery nor RT can be performed.[4]

## CONCLUSION

MPNST are rare that mostly affecting NF-1 patients. They can present as aggressive tumors that require GTR along with adjuvant RT in case of spinal metastases.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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