

# Case Report on Malignant Peripheral Nerve Sheath Tumor

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## Abstract

Malignant peripheral nerve sheath tumours (MPNST) are an uncommon type of cancer that spreads from the brain stem into the body and affects the covering of the nerves. Anywhere in the body can develop MPNST, but the deep tissues of the arms, legs, and trunk are where they most frequently occur. They result in discomfort and lethargy, mostly in the affected region, and it may develop a bulge or tumour. A 22-year-old woman presented in cancer ward with the complaints of right lower limb pain with the pain score of 4/10, the intensity of pain aggravates while sitting or performing activity and relieves while at rest, also felt heaviness since 7 months. A vast blood investigation was carried out. MRI and Biopsy were done to make her diagnosis. After following investigations, she was diagnosed as a "malignant peripheral nerve sheath tumours". Initially she was started on I.V. fluids, analgesics and injectables on the day of admission. Retroperitoneal mass excision was performed under general anaesthesia. Patient's prognosis remains good.

**Keywords:** malignant schwannoma, malignant neurilemmoma, MPNST, malignant mesenchymal lesion, neurofibrosarcomas.

## INTRODUCTION

An uncommon type of soft-tissue sarcoma with ectomesenchymal genesis is known as a malignant peripheral nerve sheath tumour (MPNST). Schwann cells or pluripotent cells generated from the neural crest are the source of MPNSTs, which develop from either significant or minor peripheral nerve segments or sheaths of peripheral nerve fibres. (1)

The most relevant terminology for tumours growing from peripheral nerves or cells linked to the nerve sheath, such as Schwann cells and perineural cells, are "malignant schwannoma", "malignant neurilemmoma", and "neurofibrosarcoma" (MPNST). (2) These accounted for about 10% of all soft tissue sarcomas. MPNST often increases between the ages of twenty and fifty in adult clients. They come from a peripheral nerve's sheath or a significant or minor branch. (3)

The sciatic nerve, brachial plexus, and sacral plexus are often the sources of basis in the limbs and thorax. We discussed a case of MPNST that only affected the right thigh, and it was identified cytologically and histopathologically. (4) Total excision represents the only recognised curative treatment. Adjuvant radiotherapy is recommended for larger lesions with more invasive histology. (5) The prognosis is still poor despite the availability of multimodal therapy, which includes surgical removal and adjuvant radiation, comprising brachytherapy.

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To deal with the disease, contemporary clinical investigations and the creation of efficient chemotherapy are required. (6)

### Case Presentation:

A 22-year-old woman presented in cancer ward with the chief complaints of right lower limb pain with the pain score of 4/10, the intensity of pain aggravates while sitting or performing activity and relieves while at rest, also felt heaviness since seven months. Primary management was done in the initial basis. A vast examination and investigation was carried out.

On Initial physical examination, The Patient appeared weak and had a dull look. During the lower extremities examination, in the right lower limb, there is a mass formation with intensive pain with redness. Her laboratory test indicated microcytic anemia with Hb- 9.8gm%, MCV- 62.1fl and thrombocytosis with PLT-5.11lack/cumm. In imaging findings, the MRI reveals measures the lesion at approximately 10.3 x 5.5 x 17.6cm (Trans x AP x cc). She appeared with mixed density heterogeneously enhancing solid cystic lesion appearing to arise from nerve roots of S1 and S2 on the right side diagnosed as malignant peripheral nerve sheath tumour. After following examinations and investigations, she was clinically diagnosed with malignant peripheral nerve sheath tumours. Initially she was started on I.V. fluids, analgesics and injectables on the day of admission.

She was treated with Tab Ultracet (OD) , Tab. Pregaba NT 75mg/10 (HS for 20 days), Tab. Contramal 100mg (BD for 20 days), Tab. Emset 4mg (BD for 15 days), Inj. Ceftriaxone 1g BD, inj. Pan 40mg OD. She underwent retroperitoneal mass excision which was performed under epidural anaesthesia. After surgery, the Patient is on symptomatic treatment. The patient and her family suffered psychological stress, resolute to a level by being an active listener and providing suitable counselling. Patient's prognosis remains good.

### Discussion:

The World Health Organization (WHO) describes MPNSTs as tumours that develop from a peripheral nerve or exhibit differentiation of the nerve sheath. Clinically, MPNST presents as a single, deeply seated, palpable mass that suddenly grows in size, is aggressive, locally invasive, and has a high rate of recurrence.(7)The neck, upper and lower extremities are the most frequently affected areas. Additionally, a right thigh tumour that was rapidly expanding was seen in our patient. In our case biopsy was done in the current case, and MRI was done and finally diagnosed MPNST. Surgery is the mainstay of treatment. (8-15)

Histologically and radiographically, MPNSTs might mimic

benign tumours. When malignancy is suspected, an MRI assessment and biopsy should be done very early to make an early diagnosis of MPNST.(9) Recent research indicates that it is particularly beneficial to distinguish MPNST from neurofibroma and/or plexiform neurofibroma using positron emission tomography-computed tomography (CT) scanning.(10)Although hemorrhagic or necrotic heterogeneity on an MRI or CT scan may indicate cancer, benign peripheral nerve sheath tumours may also exhibit this result.(16-21)

Although MPNSTs are frequently linked with poor prognosis and have significant local recurrence rates, nothing is known about the prognostic variables or efficient clinical therapy for this tumour type.(22) Surgery's ability to remove the tumour, local invasiveness, tumour size (greater than 5 cm), the scope of the procedure, the Intergroup Rhabdomyosarcoma Study grade, tumour location (central vs. peripheral), the presence of NF1, and adjuvant therapy are the most important factors influencing the prognosis of MPNST in the paediatric age group. In 75% of cases, recurrence is observed, and the 5-year survival rate ranged from 16% to 53%. A tumour size greater than 5 cm, partial tumour excision, and pre-existing NF1 are all linked to a poor prognosis.(22)

### Conclusion:

Although they can develop in any part of the body, MPNST typically develop in the deep tissues of the limbs, legs, and thorax. They frequently result in discomfort, a loss of strength, and a developing lump or tumour in the affected region. Malignant MPNST tumours have aggressive behaviour in spite of complete radical excision. Additionally, this example demonstrates the initial tumor's significant osseous and cartilaginous divergent differentiation, which was curiously lacking in the metastatic lesions. Surgical intervention is frequently used to treat MPNST. Chemotherapy & radiotherapy might also be suggested in some circumstances. In this specific instance, the client was managed first according to standard procedures, which included I.V. fluids, analgesics and injectables medications and also retroperitoneal mass excision surgery was also performed. After surgery, the Patient is on symptomatic treatment, and also providing suitable counselling. The patient's prognosis remains good.

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