JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

RECURRENT MALIGNANT PERIPHERAL NERVE SHEATH TUMOR OF THE LEFT FEMUR POST MULTIMODAL THERAPY IN 61 YEARS OLD MALE: A CASE REPORT

Diaz Alifarizki Zuvarcan[1]*, Christopher Bryan[1], Sri Marwanta[2]
[1]Clinical Clerkship, Faculty of Medicine, Sebelas Maret University, Surakarta, Indonesia
[2]Division of Hematology and Oncology, Department of Internal Medicine, Dr. Moewardi Regional Hospital, Central Java, Indonesia

Keywords:
Case report,
Malignant peripheral nerve sheath tumor,
Multimodal therapy,
Recurrent tumor,
Soft tissue sarcoma

Received: 24 August 2025
Revised: 03 October 2025
Accepted: 06 October 2025
Available online: 01 May 2026

Corresponding Author:
E-mail: alifarizki1@gmail.com

ABSTRACT

Background: Malignant peripheral nerve sheath tumors (MPNST) are rare, aggressive soft tissue sarcomas with a high rate of recurrence and metastasis. Sporadic MPNSTs, especially in older adults without neurofibromatosis type 1, present additional diagnostic and therapeutic challenges. **Case Presentation:** We report a 61-year-old male with a recurrent sporadic MPNST of the left femur. Over six years, he underwent three surgical excisions, radiotherapy, and two lines of chemotherapy. Despite initial partial response, the disease progressed with pulmonary metastases. Histopathology revealed spindle cell sarcoma with lymphovascular and skeletal muscle invasion. **Conclusion:** This case highlights the difficulty of achieving durable control in recurrent MPNST despite multimodal therapy. It underscores the importance of vigilant follow-up, individualized treatment plans, and supportive care in advanced disease settings.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) represent a rare subset of soft tissue sarcomas, arising from peripheral nerves or showing nerve sheath differentiation.[1] These tumors account for only about 5–10% of all soft tissue sarcomas, with an estimated incidence of 0.001% in the general population.[2] Despite their rarity, MPNSTs are highly aggressive, with a pronounced tendency toward local recurrence and distant metastasis. This malignant behavior, combined with the difficulty of early diagnosis, contributes to a generally poor prognosis, especially in recurrent or metastatic cases.[3]

The majority of MPNSTs are associated with neurofibromatosis type 1 (NF1), a genetic disorder that predisposes individuals to various tumors. However, approximately half of MPNST cases occur sporadically, in patients without NF1.[4] Sporadic MPNSTs are often diagnosed later and tend to exhibit more aggressive biological behavior compared to their NF1-associated counterparts.[5] In either scenario, timely diagnosis and effective intervention are critical, but the vague and insidious nature of early symptoms, such as a painless mass or mild discomfort, often leads to delayed recognition.[6]

Standard treatment protocols for MPNSTs typically involve wide surgical resection aimed at achieving negative margins, supported by radiotherapy and systemic chemotherapy.[1] Nonetheless, response to chemotherapy is frequently limited, particularly in recurrent tumors. Radiotherapy plays a crucial role in improving local control, but its impact on overall survival remains uncertain.[7] Due to the rarity of this neoplasm, most available evidence is derived from small case series and retrospective reviews, making individualized and multidisciplinary approaches central to optimizing patient outcomes.[8]

This case report presents a 61-year-old male with a recurrent MPNST of the left femur, who underwent multiple surgical resections, radiotherapy, and systemic chemotherapy. The patient's case is notable for its sporadic nature, resistance to initial therapy, and eventual pulmonary metastasis. Through this report, we aim to highlight the challenges in managing recurrent MPNST, the importance of long-term

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

follow-up, and the critical role of coordinated multimodal therapy in such rare oncologic scenarios.

CASE PRESENTATION

Patient Profile

A 61-year-old male with no prior history of neurofibromatosis presented with a progressively enlarging and painful mass in his left thigh. The initial lump was first noticed in 2019 and began as a small, firm, and painless swelling, roughly the size of a marble. Over a period of six months, the lesion expanded rapidly to the size of an adult fist and became increasingly painful, prompting the patient to seek medical attention.

Chronology of Illness and Diagnosis

In 2019, the patient underwent his first tumor excision at Hospital A, where the mass was surgically removed. Histopathological examination at that time was not clearly documented in the referral data. However, the patient remained asymptomatic for over a year.

By 2021, a recurrence of the mass was noted at the same anatomical location, his left femur. The new lesion grew to approximately the size of a chicken egg. He underwent radiotherapy (33 sessions) at Hospital B, followed by a second surgical excision due to persistent and enlarging residual mass. Post-operative wound closure was aided by a flap procedure. Despite these interventions, the tumor recurred again in 2022.

The patient was referred to Hospital C, where he underwent another excision in May 2023. Histopathological evaluation of the excised tissue revealed a spindle cell sarcoma, with strong suspicion for malignant peripheral nerve sheath tumor (MPNST). Notably, the pathology report indicated positive lymphovascular invasion and skeletal muscle infiltration, features associated with aggressive tumor behavior, while perineural invasion was absent.

Following this, the patient received adjuvant chemotherapy, including Ifosfamide (Haloxan), Mesna, and Epirubicin, as part of a first-line regimen. This therapy was administered between June and July 2022. However, the patient developed further local progression of disease, with worsening symptoms including severe pain and restricted range of motion in the affected leg. In mid-2024, he was referred to hospital C for further evaluation and treatment.

Upon his referral to hospital C in mid-2024, the patient's disease was closely monitored with serial imaging. A Multi-Slice Computed Tomography (MSCT) scan of his left thigh, performed in July 2024, was particularly informative. The scan, which included detailed axial images taken at the level of the femoral diaphysis, revealed a substantial soft tissue mass located deep within the adductor brevis muscle, measuring 7.8 x 17.4 x 8.2 cm (Figure 1A, 1C). After the patient resumed chemotherapy, a follow-up MSCT in January 2025 confirmed a significant partial response to the treatment. At the same anatomical level, the tumor's dimensions had decreased to 3.6 x 18.3 x 5.5 cm (Figure 1B, 1D), which corresponded to an estimated 67% reduction in overall tumor volume.

Treatment

Over the course of six years, the patient underwent three major surgical excisions, each prompted by local recurrence of the tumor. His radiotherapy was completed in 2022, consisting of 33 fractions delivered at Hospital B. Two courses of systemic chemotherapy were administered. The first-line chemotherapy in mid-2022 comprised Haloxan 3000 mg, Mesna 1800 mg, and Epirubicin 50 mg. In July 2024, the patient resumed this same regimen at a higher dosage: Haloxan 6000 mg, Mesna 3600 mg, and Epirubicin 50 mg, in response to disease progression.

The patient’s treatment involved a multidisciplinary approach, coordinated among surgical oncology, medical oncology, radiology, internal medicine, and pain management teams. This collaborative effort was essential to managing both the oncologic progression and the patient’s quality of life, especially in the face of recurrent and now metastatic disease.

Outcome and Follow-Up

Radiologic evidence showed a partial tumor response following the second-line chemotherapy, with MSCT scans indicating a 67% reduction in tumor volume. However, the disease trajectory remained unfavorable. Follow-up thoracic imaging revealed multiple pulmonary lesions, raising concern for metastatic spread to the lungs. Clinically, the patient reported some relief in pain intensity, with a decrease

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

in VAS (Visual Analog Scale) score from 8 to 4. Nonetheless, physical activity was limited due to persistent stiffness and pain in the left thigh.

As of the latest follow-up in May 2025, the patient remains under palliative and supportive care at Hospital C. Management now focuses on optimizing pain control, maintaining functional status, and monitoring for systemic disease progression. Given the aggressive nature of sporadic MPNST and evidence of metastasis, curative options are limited, and treatment goals have shifted toward quality-of-life preservation and symptom palliation.

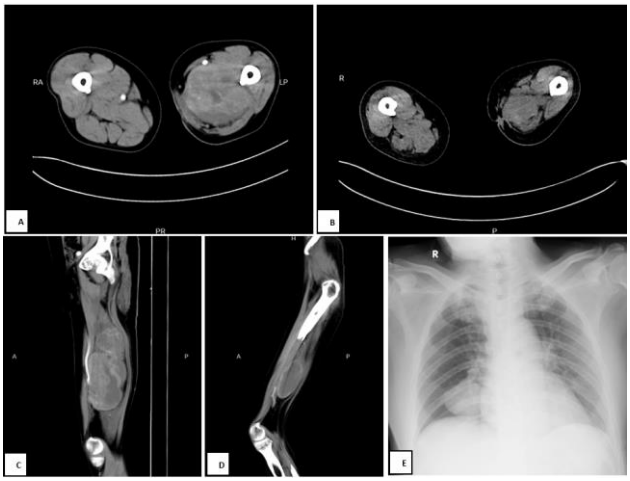

Figure 1. (A) Pre-chemotherapy MSCT (Axial): Scan at the level of the femoral diaphysis showing a large, 7.8 x 17.4 x 8.2 cm soft tissue mass in the left thigh.
(B) Post-chemotherapy MSCT (Axial): Scan at the same level showing significant tumor reduction to 3.6 x 18.3 x 5.5 cm.
(C) Pre-chemotherapy MSCT (Sagittal): showing the longitudinal extent of the tumor along the femoral shaft.
(D) Post-chemotherapy MSCT (Sagittal): showing the reduction in tumor size after treatment.
(E) Chest Radiograph: Posteroanterior view showing multiple nodules consistent with pulmonary metastases.

Table 1. Timeline of Clinical Events and Management

Date	Event	Details / Outcome
2019	Initial Presentation & First Surgery	The patient first noticed a small, firm, painless lump on his left thigh. He underwent his first tumor excision at Hospital A
2020	*Asymptomatic Period*	The patient remained asymptomatic for over a year following the initial surgery
2021	First Recurrence & Second Surgery	A recurrence of the mass was noted at the same location The patient underwent a second surgical excision at Hospital B, which included a flap procedure for wound closure
2021 - 2022	Adjuvant Radiotherapy	Following the second surgery, the patient underwent a course of radiotherapy, consisting of 33 sessions, at Hospital B.
2022	Second Recurrence & First-Line Chemotherapy	Despite radiotherapy, the tumor recurred again. In mid-2022, the patient received first-line chemotherapy (Ifosfamide/Haloxan, Mesna, Epirubicin)
May 2023	Third Surgery & Definitive Diagnosis	The patient was referred to Hospital C and underwent a third excision. Histopathology confirmed a spindle cell sarcoma, consistent with MPNST, with lymphovascular and skeletal muscle invasion
July 2024	Disease Progression & Second-Line Chemotherapy	After further local progression, the patient was referred back to Hospital C. An MSCT scan revealed a large mass (7.8 x 17.4 x 8.2 cm) He was started on a higher-dose, second-line chemotherapy regimen (same drugs)
Januar y 2025	Treatment Response	A follow-up MSCT scan showed a significant partial response, with the tumor volume reduced by an estimated 67%
May 2025	Metastatic Progression & Shift to Palliative Care	The patient developed shortness of breath. Thoracic imaging confirmed multiple pulmonary metastases. Given the advanced disease, treatment goals shifted to palliative and supportive care

DISCUSSION

Introduction

Malignant Peripheral Nerve Sheath Tumors (MPNSTs) are aggressive sarcomas with a well-documented high rate of recurrence and a generally poor prognosis.[1,2,7] While often linked to Neurofibromatosis type 1 (NF1), a significant portion of cases, such as the one presented, arise sporadically.[5,6] This case report is significant because it details the clinical course of a sporadic, high-grade MPNST in a 61-year-old male, illustrating the profound challenges of managing the disease through a multi-year, multimodal treatment regimen. The patient's journey provides a valuable clinical lesson on the limitations of current therapeutic strategies and the aggressive nature of this tumor.[26]

The particular novelty of this case lies in its detailed longitudinal documentation of sequential therapeutic failure over a six-year period. Unlike

JURNAL KEDOKTERAN DIPONEGORO
(DIPONEGORO MEDICAL JOURNAL)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

reports that may focus on a single treatment modality, this presentation offers a comprehensive view of the entire treatment arc, from the inadequacy of complete surgical resection, to the resistance to adjuvant radiotherapy, and finally to the transient response of multiple lines of systemic chemotherapy. It thereby serves as a stark, real-world illustration of the current therapeutic ceiling for this disease and underscores the aggressive biology of sporadic MPNST in the non-NF1 population.

Pathophysiology and Histopathology

The pathophysiology of sporadic MPNST is complex and involves various genetic mutations that lead to uncontrolled cell growth.[11,12] While a deep dive into molecular pathways is beyond the scope of a case report, the histopathological findings here are directly illustrative of the tumor's aggressive behavior. The diagnosis of a high-grade spindle cell sarcoma, confirmed via immunohistochemistry[13,14], is the key prognostic determinant.[10] Critically, the findings of significant mitotic activity along with perineural, lymphovascular, and skeletal muscle invasion are not just diagnostic markers; they are the pathological signature of a tumor with high metastatic potential and inherent resistance to therapy, which foreshadowed the difficult clinical course that followed.[15]

Clinical Presentation

MPNSTs typically present as a rapidly enlarging and often painful palpable mass, which was consistent with the initial presentation of this patient.[1] The symptoms are often related to the tumor's location and its compression or infiltration of adjacent nerves and tissues. In the extremities, this can lead to pain, paresthesia, or motor weakness along a specific nerve distribution.[5] In this case, the significant size of the tumor in the adductor compartment of the femur without profound neurological deficit at the outset is not unusual, but the rapid growth and recurrence are hallmark features of the tumor's aggressive nature.[7]

Diagnosis

The diagnostic process for a suspected MPNST is multi-faceted. Imaging, such as the Multi-Slice Computed Tomography (MSCT) used in this case, is essential for defining the tumor's size, location, and relationship to surrounding structures, including its vascularity and evidence of invasion.[10] However, imaging findings are not specific, and a definitive diagnosis can only be made through histopathological analysis of a tissue biopsy.[11] The use of immunohistochemical staining is a critical step to differentiate MPNST from other spindle cell sarcomas that can appear morphologically similar, such as synovial sarcoma or fibrosarcoma.[13,14] The combination of imaging and definitive tissue analysis, as performed here, represents the standard diagnostic pathway.

Multimodal Therapy

Surgical Resection

The primary and most critical intervention for localized MPNST is wide surgical excision aiming for negative margins (R0), as this provides the only real opportunity for a cure.[3,4,8,9] The initial R0 resection in this patient represented the best possible first step. However, the subsequent local recurrence is a stark illustration of a key challenge in MPNST management: even with histologically clear margins, microscopic residual disease often leads to local failure.[26] Each subsequent surgery becomes more complex and carries higher morbidity, and as seen in this case, may fail to achieve lasting local control.[16]

Adjuvant Radiotherapy

To mitigate the high risk of local recurrence, adjuvant radiotherapy is a frequently used modality, especially for large (>5 cm) and high-grade tumors like this one.[17,18] The rationale is to eradicate any microscopic cancer cells left behind after surgery.[19] The use of radiotherapy after the first recurrence was appropriate and aligned with standard guidelines. However, the eventual emergence of a second local recurrence within the treated field demonstrates that MPNST can be radio-resistant, and this modality, while beneficial, is not a panacea for preventing relapse in tumors with aggressive biology.[20]

Systemic Chemotherapy

Once the tumor recurred again and pulmonary metastases were detected, the disease was considered systemic, necessitating chemotherapy.[21] The patient's treatment with two different regimens highlights the limited efficacy of cytotoxic agents for MPNST. The initial doxorubicin-based therapy is a standard first-

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

line approach for soft tissue sarcomas.[22,24] Its failure prompted a switch to a second-line combination of ifosfamide and dacarbazine. The partial but ultimately transient response seen here is characteristic of MPNST's notorious chemo-resistance.[23] This resistance is a major obstacle; currently, there is no established, highly effective chemotherapy for recurrent or metastatic disease.[27,28] This case powerfully illustrates the current therapeutic ceiling, where after exhausting standard options, the focus invariably shifts to palliative care aimed at maintaining quality of life and managing symptoms.[25,29]

Palliative Care

When curative-intent therapies are exhausted and disease progression is inevitable, the focus of care rightly shifts to a palliative approach. For this patient, the development of pulmonary metastases and the failure of second-line chemotherapy marked this transition. The primary goals become managing symptoms (such as pain from the tumor bulk or dyspnea from lung metastases), providing psychosocial support, and optimizing the patient's quality of life.[25] This is a critical and active phase of management, not a cessation of care, and it underscores the need for early and integrated palliative services in the treatment trajectory of patients with high-grade sarcomas.[29]

Prognosis

The prognosis for patients with MPNST is generally poor and is influenced by several well-established factors.[2] Key negative prognostic indicators include large tumor size (typically >5 cm), high histological grade, deep location (subfascial), association with NF1, and, most importantly, the presence of local recurrence or distant metastases.[6,26] This patient presented with multiple negative prognostic factors: a large, high-grade, deep-seated tumor. His subsequent local recurrences and eventual development of pulmonary metastases are the strongest predictors of poor survival and are consistent with the natural history of this aggressive disease.[7] The inability to achieve lasting disease control, despite R0 resections and adjuvant therapies, underscores the grim prognosis once an MPNST has recurred.[20]

CONCLUSION

This case report details the challenging clinical course of a recurrent, sporadic malignant peripheral nerve sheath tumor in a 61-year-old male. Despite an aggressive multimodal treatment strategy that included multiple surgeries, adjuvant radiotherapy, and two lines of systemic chemotherapy, durable disease control could not be achieved, ultimately leading to distant metastasis and the need for palliative care.

The patient's journey underscores the profound limitations of current standard therapies in the face of recurrent high-grade MPNST. It highlights the aggressive nature of this malignancy and reinforces the critical need for vigilant long-term surveillance after initial treatment. This case serves as a clear example of the therapeutic ceiling in modern oncology for this rare sarcoma and emphasizes the urgent need for novel systemic agents to improve outcomes for these patients.

ETHICAL APPROVAL

This case report was conducted in accordance with the ethical principles of the Declaration of Helsinki. Informed consent was obtained from the patient for the publication of his clinical details and any associated images. The patient was assured that all personal data would be kept confidential and anonymized.

CONFLICTS OF INTEREST

The authors declare no conflict of interest.

FUNDING

No specific funding was provided for this article

AUTHOR CONTRIBUTIONS

DAZ and CB as conceptualization, analysis, data curation, writing original draft preparation, and resources. SM as investigation, review editing, and supervision.

ACKOWLEDGEMENTS

This work was supported by Faculty of Medicine, Sebelas Maret University and Dr. Moewardi Regional Hospital.

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

REFERENCES

1. Knight SWE, Knight TE, Santiago T, Murphy AJ, Abdelhafeez AH. Malignant Peripheral Nerve Sheath Tumors-A Comprehensive Review of Pathophysiology, Diagnosis, and Multidisciplinary Management. Children 2022;9. https://doi.org/10.3390/children9010038 .
2. Huang J, Shi H, Ge L, Zhu B, Liu J, Wang X, et al. Epidemiologic and survival analysis of malignant peripheral nerve sheath tumors: a retrospective cohort study. Int J Surg 2024;110:8210–4. https://doi.org/10.1097/JS9.0000000000001756.
3. Cai Z, Tang X, Liang H, Yang R, Yan T, Guo W. Prognosis and risk factors for malignant peripheral nerve sheath tumor: A systematic review and meta-analysis. World J Surg Oncol 2020;18. https://doi.org/10.1186/s12957-020-02036-x .
4. Yuan Z, Xu L, Zhao Z, Xu S, Zhang X, Liu T, et al. Clinicopathological features and prognosis of malignant peripheral nerve sheath tumor: a retrospective study of 159 cases from 1999 to 2016. Oncotarget 2017;8:104785–95.
5. Gutmann DH, Ferner RE, Listernick RH, Korf BR, Wolters PL, Johnson KJ. Neurofibromatosis type 1. Nat Rev Dis Primers 2017;3. https://doi.org/10.1038/nrdp.2017.4 .
6. Watson KL, Al Sannaa GA, Kivlin CM, Ingram DR, Landers SM, Roland CL, et al. Patterns of recurrence and survival in sporadic, neurofibromatosis Type 1-associated, and radiation-associated malignant peripheral nerve sheath tumors. J Neurosurg 2017;126:319–29. https://doi.org/10.3171/2015.12.JNS152443 .
7. Al-Mistarehi A-H, Zaitoun KJ, Khalifeh J, Saint-Germain MA, Horowitz MA, Ghaith AK, et al. An Assessment of Surgical Outcomes in Malignant Peripheral Nerve Sheath Tumors: A Systematic Review and Meta-Analysis of Surgical Interventions. Cancers (Basel) 2025;17:1997. https://doi.org/10.3390/cancers17121997 .
8. Pellerino A, Verdijk RM, Nichelli L, Andratschke NH, Idbaih A, Goldbrunner R. Diagnosis and Treatment of Peripheral and Cranial Nerve Tumors with Expert Recommendations: An EUropean Network for RAre CANcers (EURACAN) Initiative. Cancers (Basel) 2023;15. https://doi.org/10.3390/cancers15071930 .
9. Sbaraglia M, Bellan E, Dei Tos AP. The 2020 WHO Classification of Soft Tissue Tumours: News and perspectives. Pathologica 2021;113:70–84. https://doi.org/10.32074/1591-951X-213 .
10. Shurell-Linehan E, Dipardo BJ, Elliott IA, Graham DS, Eckardt MA, Dry SM, et al. Pathologic response to neoadjuvant therapy is associated with improved long-Term survival in high-risk primary localized malignant peripheral nerve sheath tumors. American Journal of Clinical Oncology: Cancer Clinical Trials 2019;42:426–31. https://doi.org/10.1097/COC.0000000000000536
11. Zhang L, Lemberg KM, Calizo A, Varadhan R, Siegel AH, Meyer CF, et al. Analysis of treatment sequence and outcomes in patients with relapsed malignant peripheral nerve sheath tumors. Neurooncol Adv 2023;5. https://doi.org/10.1093/noajnl/vdad156 .
12. Reilly KM, Kim AR, Blakely J, Ferner RE, Gutmann DH, Legius E, et al. Neurofibromatosis Type 1–Associated MPNST State of the Science: Outlining a Research Agenda for the Future. J Natl Cancer Inst 2017;109. https://doi.org/10.1093/jnci/djx124 .
13. Hirbe AC, Dehner CA, Dombi E, Eulo V, Gross AM, Sundby T, et al. Contemporary Approach to Neurofibromatosis Type 1–Associated Malignant Peripheral Nerve Sheath Tumors. American Society of Clinical Oncology Educational Book 2024;44. https://doi.org/10.1200/edbk_432242 .
14. Miao R, Wang H, Jacobson A, Lietz AP, Choy E, Raskin KA, et al. Radiation-induced and neurofibromatosis-associated malignant peripheral nerve sheath tumors (MPNST)have worse outcomes than sporadic MPNST. Radiotherapy and Oncology 2019;137:61–70. https://doi.org/10.1016/j.radonc.2019.03.015 .
15. Yao C, Zhou H, Dong Y, Alhaskawi A, Hasan Abdullah Ezzi S, Wang Z, et al. Malignant Peripheral Nerve Sheath Tumors: Latest Concepts in Disease Pathogenesis and Clinical Management. Cancers (Basel) 2023;15. https://doi.org/10.3390/cancers15041077 .
16. Zhang X, Murray B, Mo G, Shern JF. The role of polycomb repressive complex in malignant

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v15i3.53468
JKD (DMJ), Volume 15, Number 3, May 2026 : 287-293

peripheral nerve sheath tumor. Genes (Basel) 2020;11. https://doi.org/10.3390/genes11030287.
17. Cortes-Ciriano I, Steele CD, Piculell K, Al-Ibraheemi A, Eulo V, Bui MM, et al. Genomic Patterns of Malignant Peripheral Nerve Sheath Tumor (MPNST) Evolution Correlate with Clinical Outcome and Are Detectable in Cell-Free DNA. Cancer Discov 2023;13:654–71. https://doi.org/10.1158/2159-8290.CD-22-0786 .
18. Hornick JL, Nielsen GP. Beyond "Triton" Malignant Peripheral Nerve Sheath Tumors With Complete Heterologous Rhabdomyoblastic Differentiation Mimicking Spindle Cell Rhabdomyosarcoma. Am J Surg Pathol 2019.
19. Ito Y, Kohashi K, Endo M, Yoshimoto M, Ishihara S, Toda Y, et al. Clinicopathological and prognostic significance of H3K27 methylation status in malignant peripheral nerve sheath tumor: correlation with skeletal muscle differentiation. Virchows Archiv 2021;479:1233–44. https://doi.org/10.1007/s00428-021-03189-0.
20. Seres R, Hameed H, McCabe MG, Russell D, Lee ATJ. The Multimodality Management of Malignant Peripheral Nerve Sheath Tumours. Cancers (Basel) 2024;16. https://doi.org/10.3390/cancers16193266 .
21. Magallón-Lorenz M, Fernández-Rodríguez J, Terribas E, Creus-Batchiller E, Romagosa C, Estival A, et al. Chromosomal translocations inactivating CDKN2A support a single path for malignant peripheral nerve sheath tumor initiation. Hum Genet 2021;140:1241–52. https://doi.org/10.1007/s00439-021-02296-x .
22. Du P, Zhu J, Zhang ZD, He C, Ye MY, Liu YX, et al. Recurrent epithelioid malignant peripheral nerve sheath tumor with neurofibromatosis type 1: A case report and literature review. Oncol Lett 2019;18:3072–80. https://doi.org/10.3892/ol.2019.10676 .
23. Khan N, Hashmi I, Atallah L, Shaaban H, Guron G, Fedida A. A rare case report of malignant peripheral nerve sheath tumor involving both the small bowel and large bowel. Ann Afr Med 2021;20:228–31. https://doi.org/10.4103/aam.aam_10_20 .
24. Farzin M, Jafari F, Yousefi A. Two-Year Local and Distant Control of Malignant Peripheral Nerve Sheath Tumor (MPNST) in NF1 Patients after Multiple Recurrences: A Case Report. Case Reports in Clinical Practice 2024;9:57–9. https://doi.org/10.1002/cncr.22098 .
25. Moldovan H, Ciomaga I, Nechifor E, Tigănașu R, Badea A, Dobra I, et al. A Rare Case of Left Ventricular Malignant Peripheral Nerve Sheath Tumour-Case Report and Review of the Literature. Medicina (Lithuania) 2022;58. https://doi.org/10.3390/medicina58101404 .
26. Mencia MM, Moonsie R. Successful Outcome Following Limb Conservation Surgery for a Recurrent Malignant Peripheral Nerve Sheath Tumour: A Case Report. J Orthop Case Rep 2023;13:53–7. https://doi.org/10.13107/jocr.2023.v13.i12.4076 .
27. Rodon J, Rodriguez E, Maitland ML, Tsai FYC, Socinski MA, Berlin JD, et al. A phase I study to evaluate the safety, pharmacokinetics, and pharmacodynamics of PF-06939999 (PRMT5 inhibitor) in patients with selected advanced or metastatic tumors with high incidence of splicing factor gene mutations. ESMO Open 2024;9. https://doi.org/10.1016/j.esmoop.2024.102961 .
28. Chen Q, Cui H, Zheng K, Xu M, Yu X. Denosumab combined with chemotherapy followed by anlotinib in the treatment of multiple metastases of malignant peripheral nerve sheath tumor: a case report and literature review. Front Oncol 2024;14. https://doi.org/10.3389/fonc.2024.1399021 .
29. Widemann BC, Italiano A. Biology and Management of Undifferentiated Pleomorphic Sarcoma, Myxofibrosarcoma, and Malignant Peripheral Nerve Sheath Tumors: State of the Art and Perspectives. Journal of Clinical Oncology 2017;36:160–7. https://doi.org/10.1200/JCO.2017.