Cancer Association of South Africa (CANSA)

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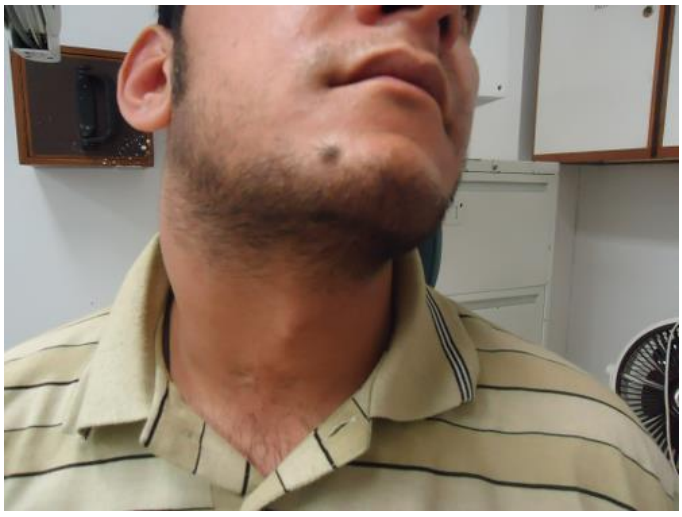
Fact Sheet
on
Malignant Peripheral Nerve Sheath Tumour (MPNST)

Introduction

Malignant peripheral nerve sheath tumours are aggressive tumours. The chance of surviving a diagnosis of MPNST depends on the size and location of the tumour. Individuals who have a small tumour tend to survive longer than those with a large tumour, and people with a tumour in the arms or legs tend to survive longer those with a tumour in the head and neck regions. Also, MPNSTs that are treated when they first occur have a better prognosis than when the tumour has regrown after initial treatments (recurred) or spread to distant parts of the body (metastasised).



[Picture Credit: MPNST]

Malignant Peripheral Nerve Sheath Tumour (MPNST)

A malignant peripheral nerve sheath tumour (MPNST) is a tumour that develops in the protective lining that covers nerves. MPNST is considered an aggressive tumour because there is up to a 65% chance of the tumour re-growing after surgery (a recurrence), and approximately 40% chance of spreading to distant parts of the body (a metastasis), most commonly to the lung. Treatment of MPNST begins with surgery to remove as much of the tumour as possible. Approximately 25-50% of MPNSTs are associated with a genetic condition known as neurofibromatosis type 1.

Malignant peripheral nerve sheath tumours can occur anywhere in the body, but most often occur in the deep tissue of the arms, legs and trunk. They tend to cause pain and weakness in the affected area and may also cause a growing lump or mass.

There are 2 major types of Malignant Peripheral Nerve Sheath Tumour:

- Spindled Malignant Peripheral Nerve Sheath Tumour (95% of the cases are of this type)
- Epithelioid Malignant Peripheral Nerve Sheath Tumour (constitute about 5% of the cases)

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Tomizawa, K., Miyazaki, T., Yamaguchi, A., Honda, R., Hoshino, M., Yanai, M., Miyamae, Y., Kurosaki, R., Shimizu, H., Arakawa, K. & Ide, M. 2020.

Background: Malignant peripheral nerve sheath tumour (MPNST) is a very rare disease, and its pathogenesis is unknown. There are few reports of MPNST of the oesophagus. We report a case of an MPNST that was diagnosed and resected.

Case presentation: A 30-year-old female presented with dysphagia. She had been aware of the dysphagia approximately 6 months before presentation. The chest X-ray showed shadows in the right mediastinum. Barium fluoroscopy revealed a semicircular raised lesion in the lower oesophagus. Upper gastrointestinal endoscopy revealed a type 1 oesophageal tumour centred on the posterior wall 26-35 cm from the incisors. The surface was ulcerated, and the tumour was exposed. The affected area showed no iodine uptake. The EUS showed an isoechoic mass. The CT scan showed a mass of 71 × 61 × 55 mm in the beginning of the lower oesophagus with low density mass and swelling of the right recurrent nerve lymph node to 12 mm. On FDG-PET, the tumour showed an SUVmax of 11.05, and no abnormal accumulation was found in lymph nodes or other organs. The MRI showed a hyperintense mass on the T2WI, which had prolonged contrast enhancement, and no findings of invasion into surrounding tissue were found. The patient underwent right thoracotomy and open thoracic oesophagectomy. The affected lymph node was tumour negative by rapid pathological diagnosis during the operation. Histologically, spindle cells with different-sized nuclei were mixed throughout the tissue. Some regions showed nuclear polymorphism or a storiform pattern, and locally, there were approximately 7 mitoses/10 HPFs. The margin was relatively clear, but spindle-shaped tumour cells infiltrated the surrounding interstitium and basal myoepithelium, and the patient was diagnosed with MPNST. In this case, the postoperative course was good, and 16 months after the operation, the patient is currently under observation at the outpatient stage without recurrence.

Conclusions: MPNST in the oesophagus is a relatively rare disease. Diagnosis before treatment is sometimes difficult, but the prognosis is good if radical resection is possible.

Farid, M., Demicco, E.G., Garcia, R., Ahn, L., Merola, P.R., Cioffi, A. & Maki, R.G. 2014.

"Malignant peripheral nerve sheath tumors (MPNST) are uncommon, biologically aggressive soft tissue sarcomas of neural origin that pose tremendous challenges to effective therapy. In 50% of cases, they occur in the context of neurofibromatosis type I, characterized by loss of function mutations to the tumor suppressor neurofibromin; the remainder arise sporadically or following radiation therapy. Prognosis is generally poor, with high rates of relapse following multimodality therapy in early disease, low response rates to cytotoxic chemotherapy in advanced disease, and propensity for rapid disease progression and high mortality. The last few years have seen an explosion in data surrounding the potential molecular drivers and targets for therapy above and beyond neurofibromin loss. These data span multiple nodes at various levels of cellular control, including major signal transduction pathways, angiogenesis, apoptosis, mitosis, and epigenetics. These include classical cancer-driving genetic aberrations such as *TP53* and phosphatase and tensin homolog (*PTEN*) loss of function, and upregulation of mitogen-activated protein kinase (MAPK) and (mechanistic) target of rapamycin (TOR) pathways, as well as less ubiquitous molecular abnormalities involving inhibitors of apoptosis proteins, aurora kinases, and the Wingless/int (Wnt) signaling pathway. We review the current understanding of MPNST biology, current best practices of management, and recent research developments in this disease, with a view to informing future advancements in patient care."

Incidence of Malignant Peripheral Nerve Sheath Tumour (MPNST)

The National Cancer Registry (2017) does not provide any information regarding the incidence of Malignant Peripheral Nerve Sheath Tumour (MPNST).

MPNSTs generally occur in adulthood, typically between the ages of 20 and 50 years of age. Approximately 10-20% of cases have been reported to occur in the first 2 decade of life, with occasional cases involving infants as young as 11 months of age.

Signs and Symptoms of Malignant Peripheral Nerve Sheath Tumour (MPNST)

Signs and symptoms of malignant peripheral nerve sheath tumours may include:

- Pain in the affected area
- Weakness when trying to move the affected body part
- A growing lump of tissue under the skin. The main clinical symptoms of MPNST are increasing size of tumours, local or radicular pain, paraparesis, and paraesthesia and/or weakness of extremities.

Higham, C.S., Dombi, E., Rogiers, A., Bhaumik, S., Pans, S., Connor, S.E.J., Miettinen, M., Sciot ,R,. Tirabosco, R., Brems, H., Baldwin, A., Legius, E., Widemann, B.C. & Ferner, R.E. 2018.

BACKGROUND: Neurofibromatosis 1 (NF1) leads to the development of benign and malignant peripheral nerve sheath tumors (MPNST). MPNST have been described to develop in preexisting benign plexiform neurofibromas (PN) and have a poor prognosis. Atypical neurofibromas (ANF) were recently described as precursor lesions for MPNST, making early detection and management of ANF a possible strategy to prevent MPNST. We aimed to clinically characterize ANF and identify management approaches.

METHODS: We analyzed clinical, imaging, and pathology findings of all patients with NF1 and ANF at 3 institutions.

RESULTS: Sixty-three patients had 76 ANF (32M/31F; median age 27.1 y). On MRI, most ANF appeared as distinct nodular lesions and were 18F-fluorodeoxyglucose (FDG) avid. Forty-six ANF were associated with pain, 19 with motor weakness, 45 were palpable or visible, and 13 had no clinical signs. Completely resected ANF (N = 57) have not recurred (median follow-up, 4.1 y; range, 0-14 y). Four ANF transformed into MPNST and 17 patients had a history of MPNST in a different location than was their ANF.

CONCLUSIONS: Growth of distinct nodular lesions, pain, and FDG-PET avidity should raise concern for ANF in NF1. Patients with ANF are at greater risk for development of MPNST. Complete resection of ANF may prevent development of MPNST.

Risk Factors for Malignant Peripheral Nerve Sheath Tumour (MPNST)

Factors that increase the risk of malignant peripheral nerve sheath tumours include:

- Previous radiation therapy for cancer. A malignant peripheral nerve sheath tumour may develop in the area treated with radiation 10 to 20 years after treatment.
- Environmental radiation exposure.
- Noncancerous nerve tumours. Malignant peripheral nerve sheath tumours can develop from noncancerous (benign) nerve tumours, such as neurofibroma.
- An inherited condition that increases risk of nerve tumours. Malignant peripheral nerve sheath tumours occur more frequently in people with neurofibromatosis 1 (Von Recklinghausen Disease).

Malbari, F., Spira, M., B Knight, P., Zhu, C., Roth, M., Gill, J., Abbott, R. & Levy, A.S. 2018.

OBJECTIVE: The main objective of this study was to determine if family history of malignant peripheral nerve sheath tumor (MPNST) increases risk of developing an MPNST in patients with neurofibromatosis-1 (NF-1).

MATERIALS AND METHODS: Individuals with NF-1 registered with the Children's Tumor Foundation's Neurofibromatosis Registry were emailed an anonymous 15-minute survey with regard to personal and family

history of NF-1, MPNST, ages of onset, and symptomatology. Participation was voluntary and information was self-reported.
RESULTS: The survey was sent to 4801 registrants, 878 responded. Presence of a family history of MPNST was found to be a risk factor for the development of MPNST; 19.4% of respondents confirming a family history of MPNST developed MPNST compared with 7.5% of respondents with no family history (odds ratio, 2.975; 95% confidence interval, 1.232-7.187; P=0.021). NF-1 patients with a positive family history developed MPNST at a younger age than those with no family history (8.3% vs. 0.5% P=0.003 and 13.9% vs. 2.4% P=0.003, for onset before 10 and 20, respectively). In the MPNST population with a known family history, onset prior to age 10 was significantly more prevalent (42.9% vs. 7% P=0.029).
CONCLUSIONS: These results suggest a positive family history of MPNST represents a risk factor for the development and early onset of MPNST in individuals with NF-1.

Diagnosis of Malignant Peripheral Nerve Sheath Tumour (MPNST)
A diagnosis of Malignant Peripheral Nerve Sheath Tumour may involve the following steps:

- Complete physical examination and comprehensive evaluation of individual's medical history (including history of NF1)
- CT or MRI scan of the affected region to aid in obtaining a clear image of the tumour, prior to surgery. To some extent, MPNSTs share basic imaging characteristics with their benign counterparts such as neurofibromas and schwannomas. These include a fusiform shape and a longitudinal orientation in the direction of the nerve. However, some distinctions are noteworthy. Large tumours (> 5 cm), invasion of fat planes, heterogeneity, ill-defined margins, and oedema surrounding the lesion are more suggestive of MPNSTs.
- Tissue biopsy: A tissue biopsy is performed and sent to a laboratory for a pathological examination, who examines the biopsy under a microscope. After putting together clinical findings, special studies on tissues (if needed) and with microscope findings, the pathologist may arrive at a diagnosis
- Molecular analysis to exclude other tumours
- If tumour has metastasised (into the lungs or other regions) then PET scan, bone scan, and chest X-rays may be taken. FDG PET is a dynamic imaging modality which evaluates metabolic activity by quantitatively assessing intracellular glucose use. It has been shown to reliably identify areas of increased metabolic activity such as those seen in malignancies
- A tissue biopsy cannot help definitively diagnose a benign MPNST from a malignant MPNST. A tissue biopsy may show overlapping features between a malignant peripheral nerve sheath tumour and a benign peripheral nerve sheath tumour, when examined under a microscope by a pathologist. Clinical correlation as regards the behaviour of the tumour is often necessary for a definitive diagnosis of malignancy.

Mustapar, N, Zawawi, M.S.F. & Tuan Sharif, S.E. 2020.
Background: Diagnosis of malignant peripheral nerve sheath tumor (MPNST) is rather challenging due to its divergent morphologic heterogeneity and lack of specific ancillary test. The emergence of H3K27 trimethylation (H3K27me3) as a new immunohistochemistry (IHC) marker for MPNST have recently available to assist pathologists in differentiating MPNST from other histologic mimics. We aim to study the expression pattern of H3K27me3 in MPNST and its histologic mimickers and their association with the clinicopathological data.
Methodology: A total of 59 benign and malignant spindle cell tumours (18 MPNST and 41 of its histologic mimickers which included 10 schwannoma, 13 neurofibroma, 4 synovial sarcoma, 3 fibrosarcoma, 2 gastrointestinal stromal tumour (GIST), 4 leiomyosarcoma, 1 spindle cell liposarcoma, 1 solitary fibrous tumour, 2 low grade fibromyxoid sarcoma and 1 unclassified spindle cell sarcoma), diagnosed from January

1998 to April 2018 in Hospital Universiti Sains Malaysia (HUSM) were tested for H3K27me3 by IHC. The MPNST histological grade was assessed based on the French Fe'de' ration Nationale des Centres de LutteContre le Cancer (FNCLCC) for 3 tiers system (low grade, intermediate grade and high grade). The clinicopathological data were retrieved from the patients' record.
Results: A total of 61.1% (11/18 MPNST) showed loss of H3K27me3 expression which is statistically significant as compared to its histologic mimics ($p<0.001$). Similar findings ($p=0.026$) were also observed in high grade MPNST (81.8%), intermediate grade MPNST (100%) and 0% in low grade MPNST.
Conclusion: H3K27me3, combined with other panel of markers, is useful in MPNST diagnosis to differentiate it from the histological mimickers.

Sangiorgio, V., Zanagnolo, V., Aletti, G., Bocciolone, L., Bruni, S., Landoni, F., Colombo, N., Maggioni, A. & Ricciardi, E. 2018.
"Cervical sarcomas are rare neoplasms, accounting for <1% of all cervical malignancies and characterized by an aggressive course despite radical excision. We report the clinical and microscopic features of a spindle cell sarcoma arising as a polypoid endocervical mass in a 45-yr-old woman. The neoplasm was characterized by a monotonous, mildly atypical proliferation of spindle cells, displaying a fibrosarcoma-like parallel pattern of highly dense fascicles, growing under the cervical epithelium. Mitotic activity was conspicuous, with up to 40 mitoses per 10 HPF. On immunohistochemistry, tumor cells were patchy S-100 protein positive. Additional immunohistochemical markers performed to rule out smooth muscle, melanocytic, epithelial, and sarcomatous differentiation were negative. A possible monophasic synovial sarcoma was also excluded by negative fluorescence in situ hybridization t(X;18) analysis. Interestingly, the neoplasm showed a focal CD34 positivity, as reported in normal fibrocytic cells of the endocervical stroma. Giving the morphologic and immunohistochemical features, the neoplasm was eventually defined as malignant peripheral nerve sheath tumor. Histologic examination following radical surgery revealed the neoplasm was confined to the uterine cervix (FIGO stage IB1) and at 12 mo of follow-up, the patient is still free of disease. Malignant peripheral nerve sheathtumors are highly aggressive sarcomas that can rarely involve the uterine cervix. They have to be differentiated from melanoma, leiomyosarcoma, endometrial stromal sarcoma, synovial sarcoma, and other spindle cell neoplasms."

Treatment of Malignant Peripheral Nerve Sheath Tumour (MPNST)
The treatment of Malignant Peripheral Nerve Sheath Tumour may be undertaken as:

- Any combination of chemotherapy, radiation therapy, and invasive procedures may be used to treat MPNST
- Embolization is used to provide temporary relief from the symptoms, and reduce blood loss during a surgical procedure
- Wide surgical excision and removal of the entire tumor, followed by radiation therapy (or chemotherapy) is the standard treatment mode
- Post-operative care is important: One must maintain minimum activity levels, until the surgical wound heals
- Follow-up care with regular screening and check-ups are important and encouraged

Martin, E., Coert, J.H., Flucke, U.E., Slooff, W.M., Ho, V.K.Y., van der Graaf, W.T., van Dalen, T., van de Sande, M.A.J., van Houdt, W.J., Grünhagen, D.J. & Verhoef, C. 2020.
Background: Despite curative intents of treatment in localized malignant peripheral nerve sheath tumours (MPNSTs), prognosis remains poor. This study investigated survival and prognostic factors for overall survival in non-retroperitoneal and retroperitoneal MPNSTs in the Netherlands.

Methods: Data were obtained from the Netherlands Cancer Registry and the Dutch Pathology Database. All primary MPNSTs were collected. Paediatric cases (age ≤18 years) and synchronous metastases were excluded from analyses. Separate Cox proportional hazard models were made for retroperitoneal and non-retroperitoneal MPNSTs.
Results: A total of 629 localized adult MPNSTs (35 retroperitoneal cases, 5.5%) were included for analysis. In surgically resected patients (88.1%), radiotherapy and chemotherapy were administered in 44.2% and 6.7%, respectively. In retroperitoneal cases, significantly less radiotherapy and more chemotherapy were applied. In non-retroperitoneal MPNSTs, older age (60+), presence of NF1, size >5 cm, and deep-seated tumours were independently associated with worse survival. In retroperitoneal MPNSTs, male sex and age of 60+ years were independently associated with worse survival. Survival of R1 and that of R0 resections were similar for any location, whereas R2 resections were associated with worse outcomes. Radiotherapy and chemotherapy administrations were not associated with survival.
Conclusion: In localized MPNSTs, risk stratification for survival can be done using several patient- and tumour-specific characteristics. Resectability is the most important predictor for survival in MPNSTs. No difference is present between R1 and R0 resections in both retroperitoneal and non-retroperitoneal MPNSTs. The added value of radiotherapy and chemotherapy is unclear.

Fenlon, J.B., Khattab, M.H., Ferguson, D.C., Luo, G., Keedy, V.L., Chambless, L.B. & Kirschner, A.N. 2019.
BACKGROUND: Malignant peripheral nerve sheath tumors (MPNSTs) are rare, aggressive soft tissue sarcomas. MPNST intracranial metastasis is exceedingly rare with only 22 documented cases in the literature and, to our knowledge, only 1 case with intraparenchymal brain metastasis. Most have been managed surgically; however, 2 documented cases were treated with Gamma Knife radiosurgery. Excluding this case report, there are no other documented cases of linear accelerator-based stereotactic radiosurgery (SRS) to treat MPNST brain metastasis.
CASE DESCRIPTION: A 41-year-old man with MPNST of the lung initially underwent tumor resection. He developed multiple systemic metastases that were managed with directed radiation therapy. A parietal brain metastasis was treated with linear accelerator-based SRS. Following SRS therapy, the patient was treated with a tropomyosin receptor kinase inhibitor. Complete resolution of brain metastasis was seen on brain magnetic resonance imaging 5 months after treatment with SRS. At 11 months after SRS, there was no evidence of recurrence or progression of the intraparenchymal disease. The patient continued to have stable extracranial disease on his ninth cycle of systemic treatment.
CONCLUSIONS: This report provides important insights into efficacy of linear accelerator-based SRS to treat MPNST brain metastases.

About Clinical Trials
Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The **South African National Clinical Trials Register** provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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Malignant Peripheral Nerve Sheath Tumour (MPNST)

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