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Neurolymphomatosis

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This report is submitted to discuss the Neurolymphomatosis

**Abstract**: The term neurolymphomatosis (NL) encompasses nerve infiltration by neurotropic neoplastic cells in the setting of an unknown or a known hematologic malignancy. It is a rare neurologic manifestation of non-Hodgkin lymphoma (NHL) and leukemia with a poorly defined incidence.(1).

**Introduction**

A rare lymphoma with a predilection for peripheral and cranial nerves, lumbar and brachial plexi and nerve roots, meninges, and vessels in the brain;1⁄2 of cases are accompanied by systemic lymphoma, which may be clinically silent; NLM may present as a painful chronic sensorimotor neuropathy with asymmetric distribution and prominent bulbar involvement; it is associated with HIV and HTLV-I infections. (1)

**Discussion**

 A 56-year-old woman was diagnosed with Stage IV diffuse large B-cell lymphoma (DLBCL) after presenting with hypercalcemia, widespread lymphadenopathy, and bone marrow involvement. She was treated with 8 cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) and achieved a complete remission. Three months later, she presented with a right wrist drop and a right Bell's palsy. MRI of the brain and spine were non-contributory. However, PET scan showed evidence of recurrent lymphoma in the brachial plexus of the right arm She was commenced on high-dose methotrexate (8 gm/m2 every 2 weeks) for NL. Her disease continued to progress despite treatment, with persistence of her initial neurological problems and development of new left cranial nerve IX/X palsies. After consultation with the patient and family, active treatment was withdrawn because of poor performance status and toxicity. The patient was palliated until her death 4 months after the diagnosis of NL.(2)

A fit 75-year-old woman presented with dyspnea and a painful radiculopathy of the right leg was consistent with pulmonary emboli and she was anticoagulated. She was also found to have Stage IV DLBCL with involvement of the liver, spleen, bone marrow, and multiple lymph node fields. MRI scans of her spine showed long-standing degenerative spinal disease but no abnormality of the central spinal cord or the lumbosacral nerve roots. CSF examination was unremarkable. PET scanning revealed fluorodeoxyglucose (FDG)-avid lesions at the sites of lymphoma listed above but no lesion of the spinal cord or nerve roots. The patient was treated with R-CHOP and prophylactic intrathecal methotrexate. Interval re-staging showed a complete response at all sites. However, after the fourth cycle of R-CHOP, the patient developed a new and painful radiculopathy of her right arm distribution. MRI of her neck and spine showed evidence of cervical spondylosis and a small, non-specific lesion in her brachial plexus A repeat PET scan showed intense FDG uptake in the brachial plexus abnormality seen on MRI and also a second lesion in the upper arm No other FDG-avid lesions were seen, consistent with persisting disease control elsewhere. A diagnosis of NL was made and the patient's treatment changed to high-dose methotrexate (8 g/m2) every 2 weeks. The patient's neurological condition stabilized for 1 month but she then developed further weakness of her legs and right arm which rendered her quadraparetic. The patient continued to deteriorate with bulbar dysfunction despite treatment with high-dose methylprednisolone (500 mg IV daily for 3 days) and DICE-R (dexamethasone, ifosfamide, cisplatin, etoposide, rituximab). The patient died 4 months after her initial diagnosis of NL..

Tobacco smoking and pregnancy is related to many effects on health and reproduction, in addition to the general health effects of tobacco. A number of studies have shown that tobacco use is a significant factor in miscarriages among pregnant smokers, and that it contributes to a number of other threats to the health of the fetus

Ideally, women should not smoke before, during or after pregnancy. If this is not the case, however, the daily number of cigarettes can be reduced to minimize the risks for both the mother and child. This is particularly important for women in developing countries where breastfeeding is essential for the child's overall nutritional status.(3)

 A 64-year-old man presented with 1 month of thoracic back pain and 1 week of left leg weakness. MRI spine showed a soft-tissue mass causing pathological fracture of the T2 vertebra and cord compression. Open biopsy at the time of a C7-T4 posterior fusion was consistent with DLBCL. Staging showed he had Stage 1AE disease. He achieved a complete remission with 6 cycles of R-CHOP, intrathecal methotrexate, and consolidation radiotherapy (30 Gy) to the T2 region. He relapsed 7 months later with an isolated cutaneous lesion over the right scapula and left footdrop consistent with peroneal neuropathy. CT and MRI scans showed no overt disease. A provisional diagnosis of NL was made and he commenced treatment with R-ICE (rituximab, ifosfamide, carboplatin, etoposide). However, he continued to deteriorate despite treatment, with the advent of bilateral leg weakness and multiple cranial nerve palsies. His treatment was changed to ESHAP (etoposide, methylprednisolone, cytosine arabinoside, and cisplatin). However, the patient continued to deteriorate and died of pneumonia 3 months after his diagnosis of NL.(4)

**Conclusion:**

Neurolymphomatosis (NL) is an uncommon syndrome of peripheral or cranial nerve root dysfunction secondary to infiltration by B-cell non-Hodgkin's lymphoma (NHL). A high index of suspicion is required as presenting symptoms are varied, conventional radiology has only modest sensitivity, and pathological diagnosis is often difficult. Treatment with chemotherapy alone has an objective response rate of 82%, although long-term outcomes are highly variable. This case series describes outcomes in THREE patients whose management incorporated PET scanning and the use of rituximab in combination with chemotherapy. PET scanning could often diagnose NL where other diagnostic modalities were non-diagnostic. Although combination therapy with rituximab and chemotherapy has been shown to be superior to chemotherapy alone in other forms of NHL, this does not appear to be the case in patients with NL. This may reflect the inability of rituximab to adequately penetrate into the central and peripheral nervous system. This is supported by the common finding that patients will relapse solely with NL despite on-going complete remission at sites outside the nervous system. The prognosis of these patients is poor, with the disease often following a progressive course despite treatment.

**Reference:**

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