Case Report

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Primary Spinal Lymphoma: A Case Report and Review of the Literature

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Introduction

Primary spinal intramedullary lymphoma (PIML) is a very rare lesion. It has not specific laboratory or radiologic findings. Therefore false and delayed diagnosis is frequent. Malignant lymphoma of the CNS usually occurs in the brain tissue. Patients were classified as "primary" if there was no evidence of systemic lymphoma by the screening in use at the time of diagnosis [1].

Case Report

A 75 year old man with no past medical history was referred to our department of neurosurgery of the Military hospital for lower limbs weakness and urinary incontinence worsening over the 3 previous months. Neurological examination revealed paraplegia and lower legs hypoesthesia. Routine blood biochemical parameters, infection markers and peripheral blood cell counts were at normal levels and serologic tests including human immunodeficiency virus (HIV) and hepatitis viruses were negative. Abdominal and thoracic computerized tomography and others investigations performed for primary tumor screening were also normal.

Spinal magnetic resonance imaging (MRI) did not reveal any intramedullar mass, but after intravenous contrast injection there was a homogenous enhancement in dura matter, the lumbar segments of the spinal cord and conus region **Figures 1 and 2**. Brain, cervical and thoracic MRIs did not reveal other abnormalities. He underwent surgery in our department, and biopsy revealed malignant lymphoma. He was addressed to radiotherapy (RT) but unfortunately he was died after 4 months.

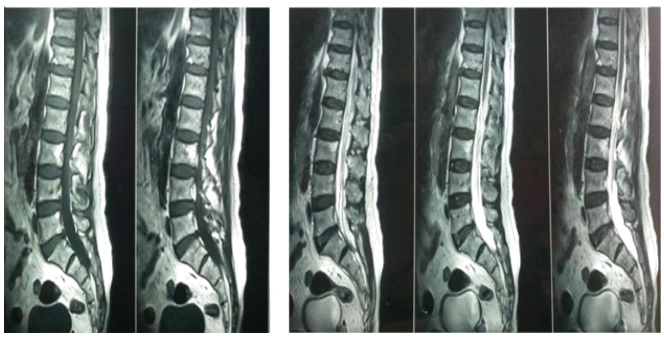

Figure 1 Normal Spinal MRI T1 weighted SAG T1 and Normal Spinal MRI: T2 weighted SAG T2.

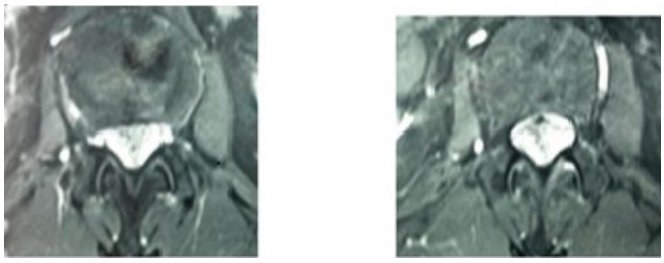

Figure 2 Spinal MRI T1 weighted axiale with gadoliniumT2 homogenous enhancement in dura matter, the lumbar segments of the spinal cord and conus region homogenous enhancement in dura matter and in the filum terminal after intravenous contrast.

Discussion

Malignant lymphoma of the CNS usually occurs in the brain tissue. Primary central nervous system (CNS) lymphoma arising from brain, leptomeninges, eyes or spinal cord is an uncommon type of extranodal non-Hodgkin's lymphoma which constitutes only 1% of all lymphomas.

Patients were classified as "primary" if there was no evidence of systemic lymphoma by screening in use at the time of diagnosis [2]. The most frequent sites are cerebral hemispheres followed by cerebellum and brain stem [3]. Dissemination of the disease to the spinal cord is not a rare event during follow-up of these cases however intramedullary lymphoma on admission is quite rare. Because of its rarity, diagnosis is difficult in this disease and there may be an important delay in both diagnosis and treatment even in the MRI era.

Clinical picture was related to localization of the tumor in the spinal cord, and there were not any specific symptoms or signs. However, these tumors were quickly progressive, and the time between symptoms onset and the admission was quite short with a median of 1.5 months [1].

Among patients presenting with lymphoma, the incidence of VZV infection is higher than in the general population [4].

Varicella zoster virus infection refers to an impaired immunity and can be used as a predictor of acquired immunodeficiency syndrome (AIDS), and in AIDS-risk groups, VZV should be regarded as a poor prognostic sign [5].

Marsh et al. reported a patient with primary leptomeningeal T-cell lymphoma where herpes zoster infection developed prior to the onset of malignant lymphoma [6].

The most important diagnostic technique for this disease is MRI. There is a typical appearance on MRI: Iso-hypointensity on T1-weighted sections, hyperintensity on T2-weighted sections, dense and usually homogeneous contrast enhancement with spinal cord enlargement and edema. Masses usually had indistinct borders. However, these characteristics are not specific for PIML. Various pathologies such as astrocytomas, metastatic tumors or neurosarcoidosis may cause similar appearances [1].

Histologically, it was composed of lymphocytes and plasmacytoid histiocytes. This tumor is indistinguishable from lymphomas arising elsewhere, as pointed out by Schaumburg [7].

The most important steps in the approach to spinal cord lymphoma are biopsy and histological diagnosis, followed by treatment with chemotherapy (CT), RT or combination of these. Neurological good response to steroids is a very important sign that must suggest lymphoma. Even unexpected remissions may be observed on MRI after steroid treatment [1].

Herrlinger et al. recommended CT including high-dose methotrexate alone or in combination with local RT according to the general guideline for treatment of other primary CNS lymphomas [8].

Unfortunately, prognosis of PIML is poor despite aggressive treatment.

Primary CNS lymphomas are radiosensitive tumors, and the use of high-dose methotrexate combined with whole-brain radiation increases median survival to 30 to 60 months.

It would seem important to reaffirm that myelopathy of undetermined etiology may be due to primary intramedullary malignant lymphoma of the spinal cord.

The effect of chemotherapy has not been definitively evaluated. Conventional chemotherapy has been generally applied subsequent to radiotherapy, or has been reserved for the treatment of recurrences and to prevent systemic relapses.

Conclusion

Primary spinal intramedullary lymphoma is a rare lesion. It must be kept in mind especially in middle aged patients. Prognosis is poor despite aggressive treatment with RT, CT or combination of them. Our work aims to highlight the fact that confronted to rapidly progressive myelopathy, practitioner should think about PIML.

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