Lymphomatosis Cerebri: MRI findings of a rare form of primary central nervous system lymphoma.

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Learning objectives

To describe the radiologic characteristics of this rare variant of primary central nervous system lymphoma.

To provide the radiologist the main diagnostic clues for the diagnosis of lymphomatosis cerebri with MRI in order to include this entity in their differential diagnostic list of diffuse white matter disorders.

Background

Primary central nervous system lymphoma (PCNSL) is an unusual form of non-Hodgkin type neoplasm, which represents 6.2%. Besides brain, this tumor can also involve meninges, eyes and spinal cord. Its appearance in immunocompetent patients in MRI is characterized by the presence of a unique enhancing mass lesion, whereas, in immunocompromised patients the typical MRI finding is the presence of enhancing lesions.

Lymphomatisis cerebri (LC) is a rare PCNSL presenting as diffuse non-mass like lesion of white matter, similar seen in gliomatosis cerebri, a rare glial tumor, and may account for its name. LC is usually described in immunocompetent patients.

Clinically, it presents with a rapidly progressive dementia and unsteadiness of gait. Its appearance in histology, the vast majority of PCNSL are diffuse large B-cell lymphoma, the remaining cases are low-grade B-cell (8%), Burkitt lymphoma (5%) or T-cell lymphoma (2-3%) (Fig.1).

According to the treatment, resection is usually not considered due to the infiltrative nature of these tumors. The main treatment in PCNSL is high dose methotrexate-based chemotherapy. However, the prognosis of lymphomatous cerebri is worse than that of PCNSL.

Findings and procedure details
We performed a retrospective review of 7 cases in order to depict the typical radiologic presentation of this disease.

At MRI, LC shows hyperintense T2 and FLAIR signal lesions involving the cerebral subcortex and occasionally extend to involve basal ganglia, thalamus and brainstem (Fig.2,3,4), reflecting widespread infiltration of the cerebral white matter by lymphomatous cells without contrast enhancement (Fig.5).

However, subtle or patchy enhancements may be seen in some cases, in our series 3 of the 7 patients showed contrast enhancement (Fig.6,7).

On diffusion sequences, restriction was seen in 66.7% (Fig. 8). In our 7 patients, 95% showed bilateral hemispheric involvement and 54% infratentorial spread.

Spinal cord MRI performed in 6 patients, showed medullar lesions in four, based on a signal abnormality in the spinal cord (Fig.9).

MRI - spectroscopy was performed in five patients and showed a peak of choline in 4, lactate in 2 and lipids in 1 patient (Fig. 10).

Due to the symmetrical distribution of white matter changes, to the prevalent infratentorial involvement and the absence of contrast enhancement, the differential diagnosis included hypertensive encephalopathy, acute disseminated encephalomyelitis (ADEM), toxic-metabolic diseases, neoplastic (gliomatosis cerebri) (Fig.11), and other infectious and autoimmune encephalitis.

**Conclusion**

LC can be a diagnostic challenge. Vague clinical and radiological findings and a lack of awareness about lymphomatosis cerebri may delay the early diagnosis. Typical MRI findings are a diffuse bilateral leukoencephalopathy, without contrast enhancement. Spinal cord and infratentorial lesions can be seen in more than half of cases. Although the presentation might not be specific, the diagnosis should be considered in patients with rapidly progressive subcortical dementia.

**Personal information**
References


