Primary CNS Lymphoma and Mimicking Disease

김 태 민
서울대학교병원 신경과

Tae Min Kim, MD, PhD
Seoul National University Hospital, Seoul, Korea

Primary CNS lymphoma (PCNSL) is a rare B-cell subtype of non-Hodgkin lymphoma arising in the CNS in the absence of lymphoma outside the nervous system[1] and diffuse large B-cell lymphoma of the CNS is the most common.[2] Personality change, cognitive impairment, and weakness were observed in cases of lymphoma infiltrating white matter tracts of the corpus callosum and internal capsule. Seizures (10%), cranial nerve palsies, and symptoms of increased intracranial pressure were less frequent.[2,3] Vitreous cellular infiltration and subretinal cellular infiltration were seen in patients with intraocular involvement (15-20%) who presented with blurred vision and eye floaters.[3] Typical radiologic findings include intense and homogeneously enhancing lesions that are either single (60-70%) or multiple (30-40%) without necrosis and with a relatively small edema usually in the periventricular space in immunocompetent patients.[2] Corticosteroids may reduce or eliminate abnormal contrast enhancement that compromise the interpretation.[4] In immunocompromised patients, brain imaging shows multifocal abnormalities and a peripheral rather than homogeneous enhancement pattern.[4] Although definite diagnosis of leptomeningeal lymhpomatosis is made by a positive CSF cytology, the sensitivity of CSF cytology varies widely (2-32%). Sensitivity improves when a large volume (≥ 10.5 mL) is analyzed and serial CSF samples are evaluated.[4] Differential diagnoses include tumors (glioblastomas and metastases), infectious diseases (tuberculosis and toxoplasmosis), cerebral vasculitis or multiple sclerosis plaques. Rare differential diagnoses include chronic lymphocytic inflammation with pontine perivascular enhancement response to steroids (CLIPPERS) and Langerhans cell histiocytosis.

Key Words: Primary CNS lymphoma; Diffuse large B-cell lymphoma

References

1. Deckert M, Paulus W: Malignant lymphomas, in Louis DN, Ohgaki H, Wiestler OD, et al (eds): WHO Classification of Tumours of the Central Nervous System. Lyon, France, IARC, 2007, pp 188-92
2. Korfel A, Schlegel U: Diagnosis and treatment of primary CNS lymphoma. Nat Rev Neurol 9:317-27, 2013
3. Hochberg FH, Baehring JM, Hochberg EP: Primary CNS lymphoma. Nat Clin Pract Neurol 3:24-35, 2007
4. Scott BJ, Douglas VC, Tihan T, et al: A systematic approach to the diagnosis of suspected central nervous system lymphoma. JAMA Neurol 70:311-9, 2013

Tae Min Kim, MD, PhD
Department of Internal Medicine, Seoul National University Hospital, 101 Daehak-ro, Jongno-gu, Seoul 110-744, Korea
TEL: +82-2-2072-3559 FAX: +82-2-764-2199
E-mail: gabriel9@snu.ac.kr