A case of intravascular malignant lymphoma in the brain that occurred with subacute monoplegia of the right leg

Takao Mitsui[#1], Arisa Shimizu[#1]

#1 Department of Clinical Research, Tokushima National Hospital, National Hospital Organization 1354 Shikiji, Kamojima, Yoshinogawa, Tokushima 776-8585, Japan.

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Introduction

Intravascular lymphoma (IVL), also known as intravascular lymphomatosis, is a rare neoplastic disorder in which tumor cells are initially confined to the vascular lumen without parenchymal infiltration. This condition preferentially affects the central nervous system (CNS) and skin. One-third of IVL patients present with heterogeneous neurological symptoms at diagnosis [1]. In cases of CNS involvement, most of the common symptoms observed are related to ischemia and infarction due to small vessel occlusion by the tumor cells. While cases of systemic intravascular lymphoma are more frequently encountered, cases of intravascular lymphoma with restricted central nervous system involvement are uncommon [2-5]. CNS and cutaneous involvement are less common in the Asian variant than in the Western variant [1,6]. Under these conditions, isolated IVL of the CNS tends to be overlooked or misdiagnosed as cerebral infarctions, especially in Asian countries. Here, we report the case of an IVL patient whose initial symptom was monoplegia of the right leg.

Case Report

The patient (39-year-old woman) suffered from lassitude of the right leg and visited the Tokushima National Hospital. She worked in a factory and conducted manufacturing duties of the calculation drill. In 2004, she underwent surgery of the right knee for arthrosis. We were working in the middle of June, 2016, and the sense of incongruity of the right leg appeared. She was received by a nearby orthopedics department on June 27. At that time, there was not the objective exercise, or sensory disturbance. Her right leg lassitude appeared at the end of July and gradually increased. She was received by the orthopedics department again at the beginning of August. There was muscle weakness of the right leg. A continuous light hyperintensity area was detected in a dorsal by MRI on August 15. She came to Tokushima National Hospital on suspicion of myelitis on August 16. In a neurological examination, the cranial nervous system was normal. The right leg had moderate or severe muscle weakness. Other extremities did not have muscle weakness. Algesthesia of the right anterior surface of the leg decreased. Deep tendon reflexes were normal in the upper limbs but aggravated in the lower limbs. Babinski reflexes were bilaterally positive. Brain MR imaging (**Figure 1** and **Figure 2**) showed a solid mass approximately 3cm in left parietal lobe lower white matter. Furthermore, the invasive image which opened around a mass was detected. A thoracic MRI showed a similar invasive image from the 5th to 8th thoracic segments (not shown). She transferred to a brain surgery hospital and underwent a brain

biopsy, conducted using the stereotaxic technique. Pathological examination demonstrated intravascular B cell lymphoma.

Discussion

In the present case, the initial manifestation of a neurological sign was monoplegia of the right leg. This symptom does not strongly indicate brain lesion. It has been reported in 79 cases of IVL in 1989 and it was found that 32% of the patients had symptoms attributable to CNS lesions alone, whereas 12% had CNS and systemic symptoms [7]. IVL is a systemic disease and owing to advances in imaging technology in the past two decades, asymptomatic lesions of other organs have been found in most cases. CNS lesions were detected in 40% of European IVL patients as multiorgan infiltration but lesions limited to the CNS were found only in 5% [8]. An Asian retrospective study of 96 patients with IVL reported that CNS involvement accounted for 27%, and two of 81 (2.5%) IVL patients underwent brain biopsy, providing more evidence of this rare condition with restricted CNS involvement [9]. In our case, FDG-PET did not detect any systemic lesions except for the right eyeball. IVL with restricted CNS involvement is not only a rare condition but also refractory for treatment. In any case, early diagnosis is extremely important.

References

1. Ponzoni M, Ferreri AJ. Intravascular lymphoma: a neoplasm of 'homeless' lymphocytes? Hematol Oncol, 2006; 24: 105–112
2. Baehring JM, Longtine J, Hochberg FH: A new approach to the diagnosis and treatment of intravascular lymphoma. J Neurooncol 2003, 61: 237–48.
3. Debiais S, Bonnaud I, Cottier JP, et al.: A spinal cord intravascular lymphomatosis with exceptionally good outcome. Neurology 2004, 63: 1329–30.
4. DiGiuseppe JA, Nelson WG, Seifter EJ, Boitnott JK, Mann RB: Intravascular lymphomatosis: a clinicopathologic study of 10 cases and assessment of response to chemotherapy. J Clin Oncol 1994, 12: 2573–9.
5. Pless ML, Chen YB, Copen WA, Frosch MP: Case records of the Massachusetts General Hospital. Case 9–2010. A 37-year-old woman with paresthesias and ataxia. N Engl J Med 2010, 362: 1129–38.
6. Ferreri AJ, Dognini GP, Campo E, Willemze R, Seymour JF, Bairey O, Martelli M, De Renz AO, Doglioni C, Montalbán C, Tedeschi A, Pavlovsky A, Morgan S, Uziel L, Ferracci M, Ascani S, Gianelli U, Patriarca C, Facchetti F, Dalla Libera A, Pertoldi B, Horváth B, Szomor A, Zucca E, Cavalli F, Ponzoni M; International Extranodal Lymphoma Study Group (IELSG). Variations in clinical presentation, frequency of hemophagocytosis and clinical behavior of intravascular lymphoma diagnosed in different geographical regions. Haematologica, 2007; 92: 486–492.
7. Domizio P, Hall PA, Cotter F, Amiel S, Tucker J, Besser GM, Levison DA. Angiotropic large cell lymphoma (ALCL): morphological, immunohistochemical and genotypic studies with analysis of previous reports. Hematol Oncol, 1989; 7: 195–206.
8. Ferreri AJ, Campo E, Seymour JF, Willemze R, Ilariucci F, Ambrosetti A, Zucca E, Rossi G, López-Guillermo A, Pavlovsky MA, Geerts ML, Candoni A, Lestani M, Asioli S, Milani M, Piris MA, Pileri S, Facchetti F, Cavalli F, Ponzoni M; International Extranodal Lymphoma Study Group (IELSG). Intravascular lymphoma: clinical presentation, natural history, management and prognostic factors in a series of 38 cases, with special emphasis on the 'cutaneous variant'. Br J Haematol, 2004; 127: 173–183.
9. Murase T, Yamaguchi M, Suzuki R, Okamoto M, Sato Y, Tamaru J, Kojima M, Miura I, Mori N, Yoshino T, Nakamura S. Intravascular large B-cell lymphoma (IVLBCL): a clinicopathologic study of 96 cases with special reference to the immunophenotypic heterogeneity of CD5. Blood, 2007; 109: 478–485.

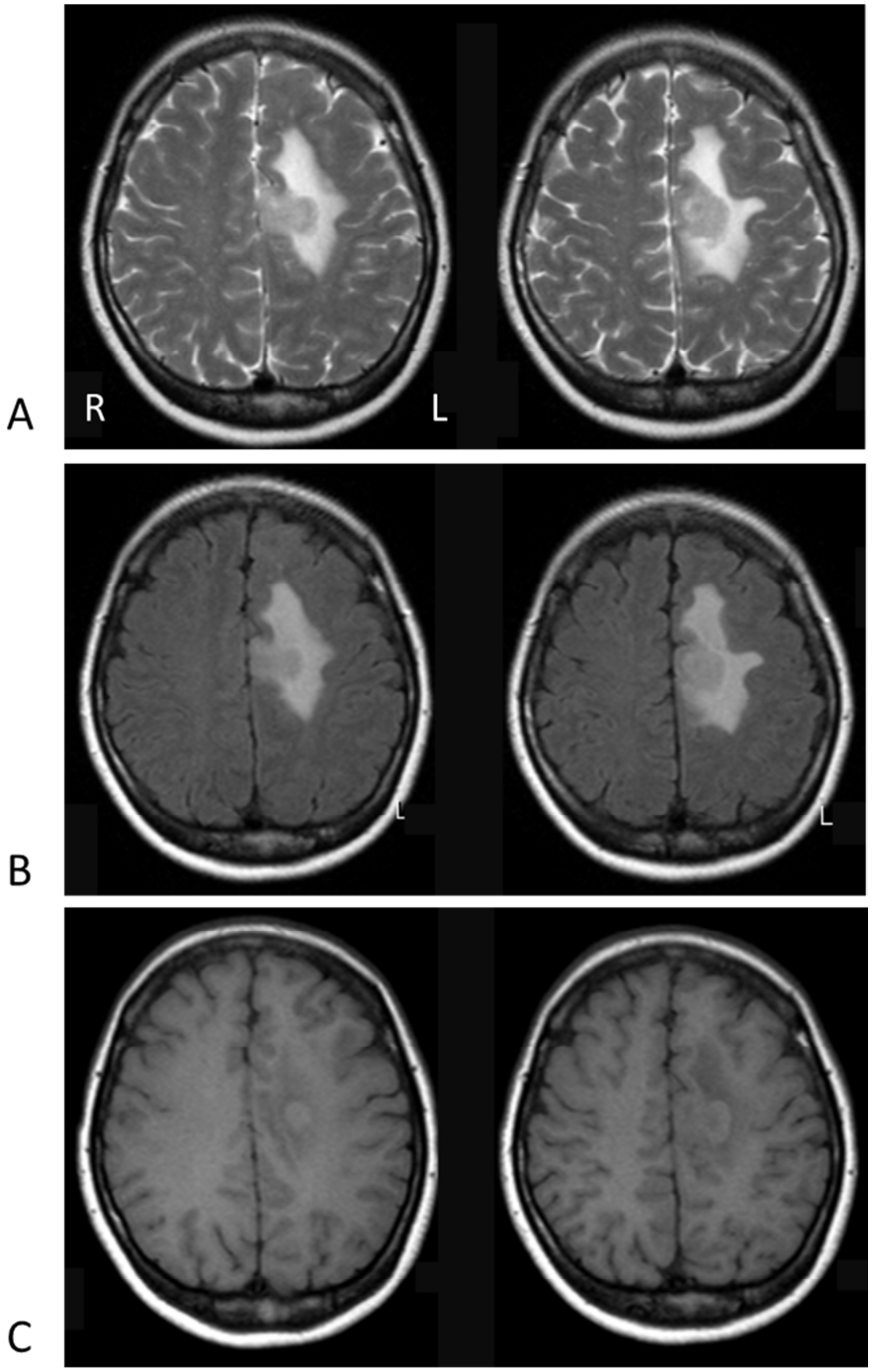

Figure 1. Brain MRI of the present patient. Axial sections. A, T2-weighted image; B, proton density-weighted image; T1-weighted image.

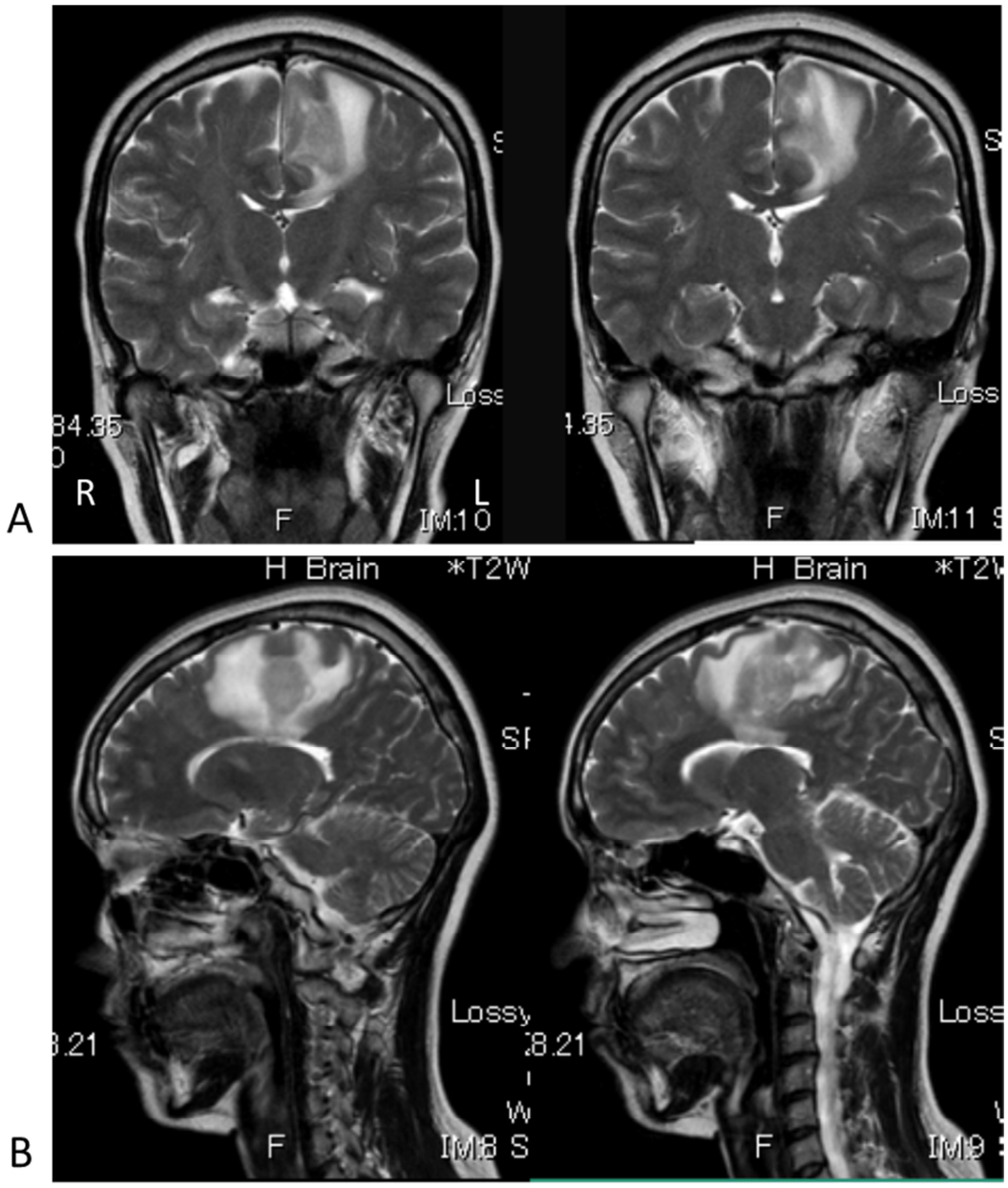

Figure 2. Brain MRI of the present patient. A, coronal section; B, sagittal section.