BMJ Neurology Open

Clinical manifestations and outcomes of patients with intravascular large B-cell lymphoma with neurological involvement: highlighting longitudinally extensive myelopathy as a distinct feature

Ekdanai Uawithya ,[1,2] Palakorn Lertsakworakul,[3] Weerapat Owatthanapanich,[4] Jiraporn Jitprapaikulsan[2,3]

To cite: Uawithya E, Lertsakworakul P, Owatthanapanich W, *et al*. Clinical manifestations and outcomes of patients with intravascular large B-cell lymphoma with neurological involvement: highlighting longitudinally extensive myelopathy as a distinct feature. *BMJ Neurology Open* 2025;**7**:e000915. doi:10.1136/bmjno-2024-000915

Received 19 September 2024
Accepted 14 December 2024

For numbered affiliations see end of article.

Correspondence to
Jiraporn Jitprapaikulsan; jiraporn.jit@mahidol.ac.th

ABSTRACT

Objective This study aimed to elucidate the clinical manifestations, laboratory findings and outcomes of patients with intravascular large B cell lymphoma (IVLBCL) with neurological involvement and to differentiate IVLBCL with and without neurological involvement.

Methods A cohort study was conducted at Siriraj Hospital, Mahidol University, Thailand, between January 2005 and September 2024. Clinical data, laboratory values and central nervous system imaging results were analysed. Categorical variables were compared using the χ^2 or Fisher's exact test, while continuous variables were analysed with the Mann-Whitney U test, as appropriate.

Results Of the 30 patients with IVLBCL, 10 had neurological involvement and 20 without neurological symptoms, including myelopathy (5 patients, 50%); cognitive impairment (3 patients, 30%); seizures (2 patients, 20%); optic neuropathy, hemiparesis, homonymous hemianopia, vertigo and global aphasia (each affecting 1 patient, 10%). 60% of IVLBCL with neurological involvement had systemic symptoms, including prolonged fever, anaemia, anorexia and weight loss. MRI showed hyperintense lesions in the supratentorial, infratentorial and spinal cord with the prominent findings being longitudinally extensive cord lesions (four patients, 40.0%). The median survival time of the IVLBCL with neurological involvement was 4.1 months (95% CI: 0.0 to 17.1 months), with a 1-year survival rate of 37.5% and a 2-year survival rate of 25.0%.

Interpretation This study highlights the distinct clinical, laboratory features and imaging of IVLBCL with neurological involvement and compares it to IVLBCL without neurological involvement. Early recognition of these findings is crucial for accurate diagnosis and improved patient outcomes despite the aggressive nature of IVLBCL.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Intravascular large B-cell lymphoma (IVLBCL) is a rare and aggressive lymphoma that often presents with non-specific symptoms. Neurological symptoms are observed in some patients, especially those with the haemophagocytic syndrome-associated variant.

WHAT THIS STUDY ADDS

⇒ IVLBCL in the Thai population can be presented as myelopathy (50.0%), cognitive decline (30.0%), seizures (20.0%), optic neuropathy, hemiparesis, homonymous hemianopia, vertigo and global aphasia (each affecting one patient, 10%). This study emphasises interesting findings of longitudinally extensive cord lesions and stroke-like lesions on the MRI.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ The findings can help distinguish IVLBCL from other conditions using clinical manifestations, laboratory findings and imaging.

INTRODUCTION

Intravascular lymphoma large B-cell lymphoma (IVLBCL) is a rare hematological malignancy characterised by abnormal lymphomatous cells within vessel lumina. It affects approximately less than 1 out of 1 000 000 individuals.[1 2] The mean survival postdiagnosis is a mere 340 days, indicating that IVLBCL is one of the most aggressive malignancies.[3] IVLBCL is classified into haemophagocytic syndrome-associated and classical variants (based on geographical and clinical characteristics, with the haemophagocytic syndrome-associated variant showing greater incidence in the East Asian population), greater liver and spleen involvement but fewer neurological and skin manifestations than its counterpart. Neurological symptoms are observed in 27% of cases of the haemophagocytic syndrome-associated variant, while they occur in 39–76% of the classical variant.[4 5]

IVLBCL can be presented as a variety of systemic symptoms, such as fever, headache and weight loss.[4 6] A 2016 meta-analysis revealed that neurological manifestations can occur in up to 42% of patients with IVBCL.[7] Common neurological symptoms include cognitive impairment, paralysis and seizures,[8] whereas peripheral nervous system involvement is less prominent. These neurological symptoms may mimic those of neuroinflammatory diseases such as multiple sclerosis or encephalomyelitis and stroke.[9] Despite the critical nature of this issue, limited data exist on the neurological manifestations of IVLBCL, particularly in the Thai population. Understanding the differences in clinical presentation and laboratory findings of IVLBCL is crucial for early diagnosis and treatment.

This study aimed to elucidate the clinical manifestations, laboratory findings, pathological imaging and overall survival rate of patients with IVLBCL with neurological involvement and to differentiate from IVLBCL without neurological involvement.

METHODS

Patient population and data collection

This cohort study was conducted at Siriraj Hospital, Mahidol University, a tertiary university-based centre in Thailand. Data was collected from Siriraj's electronic database for cases documented between January 2005 and September 2024, including patient characteristics, clinical presentations, laboratory values, central nervous system (CNS) imaging, pathological findings and treatments. The inclusion criteria required patients aged 18 years or older with pathologically confirmed IVLBCL, either premortem or postmortem. The included patients were then divided into two groups: those with IVLBCL who had neurological involvement and those with IVLBCL who did not.

Statistical analysis

Statistical analyses were performed using PASW Statistics, V.18 (SPSS, Chicago, Illinois, USA). The χ^2 or Fisher's exact test was used for categorical variables, while the Mann-Whitney U test was employed for continuous variables. Continuous data were presented as medians and IQRs, whereas categorical variables were reported as frequencies and percentages. Survival data were estimated using the Kaplan-Meier method. The median survival time was reported with 95% CIs. A p value less than 0.05 was considered to indicate statistical significance.

Outcome parameters

The primary outcomes were to elucidate (1) the neurological manifestations, laboratory findings, pathological findings and imaging characteristics of patients with IVLBCL and neurological involvement and (2) the comparison of clinical characteristics and laboratory findings between IVLBCL with and without neurological patients. The secondary outcomes were the survival rates of patients with IVLBCL with neurological manifestations and patients with IVLBCL without neurological manifestations.

RESULTS

Baseline characteristics and demographics

This study included 30 patients with IVLBCL, 10 of whom exhibited neurological involvement, while 20 did not. The median age at disease onset was 69.0 years (IQR 61.0–72.3 years) and the cohort comprised 21 females (70.0%). All patients had initial modified Rankin Scale scores between 0 and 3. The entire study population consisted of Asian individuals. Two-thirds (20/30 cases) of this cohort had positive bone marrow. Approximately 80% (21/26 cases) of the patients had lymphoma involvement in the skin from a random skin biopsy. All patients had stage IV IVLBCL according to the Ann Arbor staging classification.[10] 11 patients (36.7%) had liver involvement and 12 patients (40.0%) had spleen involvement. The International Prognostic Index scores were distributed as follows: 2 in 2 patients (6.7%), 3 in 9 patients (30.0%), 4 in 13 patients (43.3%) and 5 in 6 patients (20.0%) (table 1).

Neurological symptoms of IVLBCL

All 10 patients with IVLBCL with neurological involvement exhibited CNS involvement as their initial symptom. Neurological symptoms consisted of myelopathy (five patients, 50%); cognitive impairment (three patients, 30%); seizures (two patients, 20%); optic neuropathy, hemiparesis, homonymous hemianopia, vertigo and global aphasia (each affecting one patient, 10%) (figure 1 and table 2).

Systemic symptoms differences between patients with IVLBCL with and without neurological involvement

Among the IVLBCL with neurological involvement patients, five (50.0%) had prolonged fever, three (30.0%) with significant weight loss and one (10.0%) patient with anorexia. For IVLBCL without neurological involvement, 19 patients (95.0%) had prolonged fever, 14 (70.0%) with significant weight loss and 8 (40.0%) with anorexia (table 1). There was a statistically significant higher proportion of prolonged fever in the IVLBCL without neurological involvement patients compared with the neurological involvement patients (p=0.009).

Differences in laboratory parameters between patients with IVLBCL with and without neurological involvement

90% of IVLBCL with neurological involvement had anaemia; median haemoglobin was 89 (68–105) g/L. Patients with IVLBCL with neurological manifestations had a statistically significantly lower serum lactate dehydrogenase (LDH) than IVLBCL without neurological manifestations (475.5 (368.0–960.5) vs 1393.5 (896.0–2428.5) U/L, p=0.001). Conversely, the haemoglobin count was higher in the neurological involvement group (89 (68–105) g/L vs 80 (68–98) g/L, p=0.301)

Table 1 Baseline characteristics and laboratory findings of patients with intravascular large B cell lymphoma

	All patients (n=30)	IVLBCL without neurological involvement (n=20)	IVLBCL with neurological involvement (n=10)	P values
Characteristics				
Female, n (%)	21 (70.0)	14 (70.0)	7 (70.0)	1.000
Age at onset, years, median (IQR)	69.0 (61.0–72.3)	67.5 (61.0–71.0)	70.0 (61.5–75.8)	0.320
Time from onset to diagnosis, days, median (IQR)	97.0 (66.3–146.5)	96.0 (63.0–129.0)	130.0 (83.5–173.5)	0.210
Systemic involvement				
Prolonged fever	24 (80.0%)	19 (95.0%)	5 (50.0%)	0.009
Significant weight loss	17 (56.7%)	14 (70.0%)	3 (30.0%)	0.056
Anorexia	9 (30.0%)	8 (40.0%)	1 (10.0%)	0.204
Baseline laboratory				
Haemoglobin, median (IQR)	8.1 (6.9–9.8)	8.0 (6.8–9.8)	8.9 (6.8–10.5)	0.301
Haematocrit, median (IQR)	24.8 (21.4–29.1)	24.7 (21.5–29.1)	26.7 (21.1–33.0)	0.391
White blood cell, median (IQR)	6270.0 (4185.0–9397.5)	5560.0 (3905.0–9050.0)	7020.0 (4575.0–11 127.0)	0.403
Platelet (×1000), median (IQR)	129.0 (76.3–204.3)	128.0 (73.5–188.5)	134.0 (97.5–211.1)	0.725
LDH level, median (IQR)	1067.5 (677.5–1723.8)	1393.5 (896.0–2428.5)	475.5 (368.0–960.5)	0.001

IVLBCL, intravascular large B cell lymphoma; LDH, lactate dehydrogenase.

but without statistical significance. The same trend goes for haematocrit (0.27 [(0.21–0.33]) L/L vs 0.25 [(0.22–0.29]) L/L, p=0.391), white blood cell count (7.0 [(4.6–11.1)]× 109/L vs 5.6 [(3.9–9.0]) ×109/L, p=0.403) and platelet count (134 [(97.5—211.1])×109/L vs 128[(73.5–188.5])×109/L, p=0.725) with no statistical significance (table 1).

Imaging findings in patients with IVLBCL with neurological involvement

All patients with IVLBCL and neurological involvement underwent CNS imaging. Nine patients (90.0%) had abnormal findings detected by MRI. Lesions were observed in the subcortical area, cortex and cerebellar region. The characteristics of brain haemorrhage and

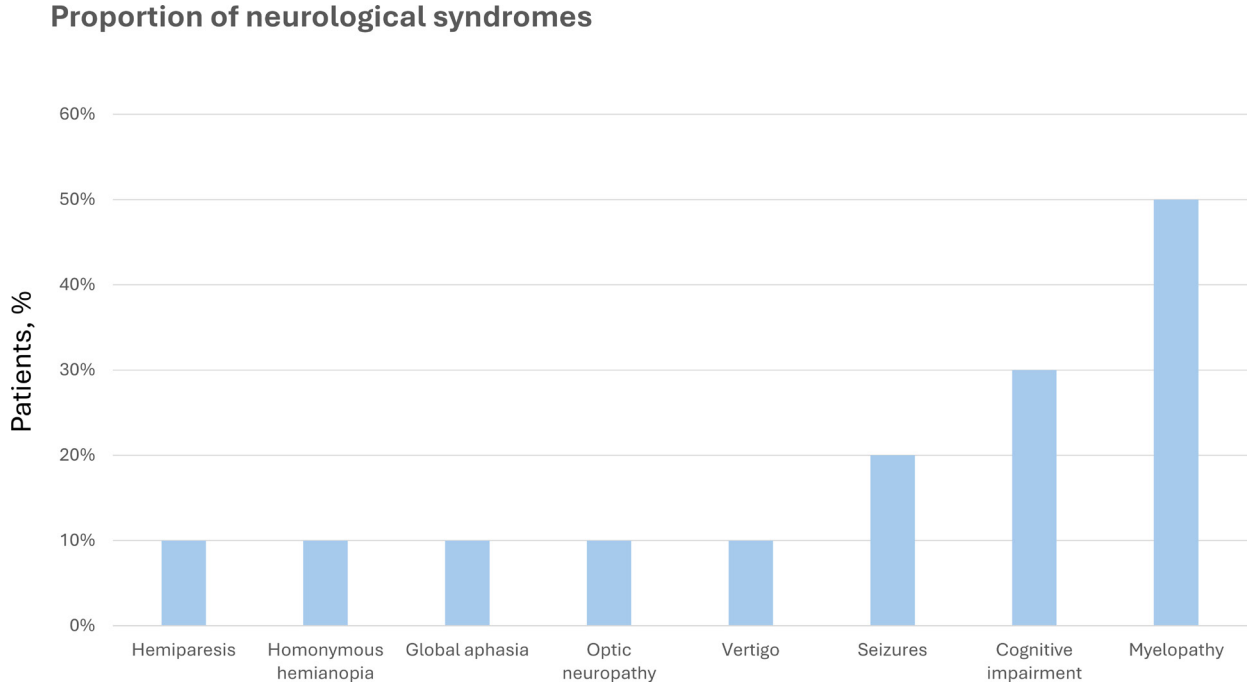

Figure 1 Neurological symptoms in patients with intravascular large B cell lymphoma.

Table 2 Clinical manifestations and laboratory findings of patients with intravascular large B cell lymphoma with neurological involvement

Patients number	Sex/age at onset	Neurological syndrome	Systemic symptoms	White blood cell count, / mm^3 CSF protein, mg/dL; CSF glucose/serum glucose	Diagnostic methods	Treatment
1	F/71	Non-convulsive status epilepticus, left homonymous hemianopia, left-sided hemiparesis	NA	7 (L 100%); 99; NA	Skin/bone marrow biopsy	Supportive
2	M/45	Cognitive impairment, multistage cerebral infarction and haemorrhage	NA	48 (N 78%, L 14%); 177; 77/135	Skin biopsy	Supportive
3	M/78	Longitudinally extensive thoracic myelopathy	Prolonged fever	0; 40; 34/123	Skin biopsy	Supportive
4	M/70	Longitudinally extensive thoracic myelopathy, Cauda equina syndrome, optic neuropathy	NA	8 (N 9%, L 84%); 45; 42/94	Skin biopsy	Supportive
5	F/70	Cognitive impairment, recurrent stroke, behavioural changes	NA	10 (N 37%, L 55%); 106; 65/161	Brain biopsy	Supportive
6	F/76	Longitudinally extensive thoracic myelopathy	Prolonged fever	NA	Skin biopsy	Supportive
7	F/75	Conus medullaris syndrome	Prolonged fever, significant weight loss	8 (L 100%); 32; 110/217	Skin biopsy	R-CHOP
8	M/65	Longitudinally extensive thoracic myelopathy, vertigo	Anaemic symptoms	8 (N 2%, L 98%); 161; 106/117	Skin/bone marrow biopsy	Supportive
9	F/72	Cognitive impairment, aphasia	Prolonged fever, significant weight loss	12 (N 8%, MN 91%); 64.5; 151/239	Skin/bone marrow biopsy	Supportive
10	F/51	Generalised tonic-clonic seizures	Prolonged fever, significant weight loss, anorexia, anaemic symptoms	0; 30; 73/149	Bone marrow biopsy	R-CHOP

CSF, cerebrospinal fluid; F, female; L, lymphocyte; M, male; MN, mononuclear cell; N, neutrophil; NA, not available; R-CHOP, rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisone.

infarction were also presented in some patients. The affected regions were the cerebral cortex, cerebellar hemisphere, basal ganglia and periventricular area.

Among five patients presenting with myelopathy, the mean onset was 97 days (range 12–168 days). Four of the five patients had extensive long spinal cord lesions (>3 vertebral segments) in T2-weighted imaging, whereas one patient with conus medullaris syndrome had a normal spinal cord MRI. Of the four patients with abnormal long cord lesions, two had faint enhancement and two had no gadolinium enhancement (figures 2–4).

Outcomes of patients with IVLBCL

12 of 30 patients (40.0%) received chemotherapy, all of which were treated with the R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisone) as induction therapy. The overall median survival time for all patients was 44.1 months (95% CI: 0.0 to 232.8 months), with a 1-year survival rate of 56.5% and a 2-year survival rate of 52.7%.

Among patients with IVLBCL and neurological involvement, only two patients (20.0%) received chemotherapy. Eight patients were not fit for chemotherapy and received symptomatic treatments. The median survival time in this group was 4.1 months (95% CI: 0.0 to 17.1 months), with a 1-year survival rate of 37.5% and a 2-year survival rate of 25.0%. Conversely, patients with IVLBCL without neurological involvement had a notably longer median survival time of 321 months (95% CI: 0.0 to 663.3 months) and achieved a 1-year and 2-year survival rate of 65.0%.

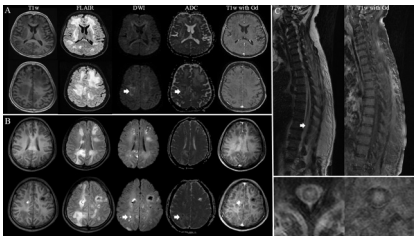

Figure 2 MRI features of patients 1–3 with IVLBCL. (A) Patient 1: This patient presented with non-convulsive status epilepticus, left homonymous hemianopia and left hemiparesis. MRI revealed bilateral asymmetrical hypointense signals on T1-weighted (T1W) images and hyperintense signals on T2/FLAIR images in the bilateral subcortical regions and periventricular white matter without restricted diffusion. T1-weighted imaging with gadolinium (Gd) enhancement revealed punctate and curvilinear enhancement with multiple foci of blooming artefacts within these lesions (SWI not shown). (B) Patient 2: This patient presented with rapid cognitive decline. MRI revealed multifocal hypointense and hyperintense signals on T1W and T2/FLAIR images, with restricted diffusion suggesting multistage intraparenchymal infarction (white arrow) and haemorrhage. Two Gd-enhanced lesions in the occipital region were compatible with subacute infarction. (C) Patient 3: This patient presented with chronic progressive thoracic myelopathy and prolonged fever. MRI revealed hyperintense signals on T2/FLAIR images with faint enhancement from T7 to the conus medullaris (white arrow). IVLBCL, intravascular large B cell lymphoma, ADC, Apparent diffusion coefficient; DWI, Diffusion-weighted imaging; FLAIR, Fluid-attenuated inversion recovery; SWI, Susceptibility-weighted imaging.

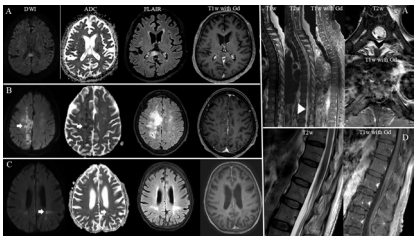

Figure 3 MRI features of patients 4–7 with IVLBCL. (A) Patient 4: This patient presented with subacute thoracic myelopathy and optic neuropathy. Spinal MRI revealed isointense T1-weighted (T1W) signals and hyperintense T2-weighted (T2W) signals from T2 to T7 without gadolinium (Gd) enhancement. Brain MRI revealed a hyperintense FLAIR signal in the splenium without restricted diffusion or Gd enhancement. (B) Patient 5: This patient presented with rapid cognitive decline, recurrent stroke and behavioural changes. MRI showed multifocal cortical/subcortical white matter hyperintensities on T2/FLAIR images with restrictive diffusion (white arrow) and punctate Gd enhancement. (C) Patient 6: This patient presented with longitudinally extensive transverse myelopathy and prolonged fever. MRI revealed multifocal hyperintense signals on T2/FLAIR images with restricted diffusion in bilateral periventricular lesions and Gd enhancement. Extensive long-segment spinal enlargement with high signal intensity was observed on T2-weighted images and enhancement from T1 to the conus medullaris (head arrow). (D) Patient 7: This patient presented with conus medullaris syndrome, prolonged fever and significant weight loss. MRI showed no abnormal signal change on T2/FLAIR or T1W images with Gd enhancement. IVLBCL, intravascular large B cell lymphoma, ADC, Apparent diffusion coefficient; DWI, Diffusion-weighted imaging; FLAIR, Fluid-attenuated inversion recovery.

DISCUSSION

One-third of patients with IVLBCL in this cohort had neurological involvement. In most cases, neurological symptoms were the initial symptoms. All neurological involvements were confined to the CNS, including myelopathy, seizures, cognitive impairment, multistage stroke, encephalopathy and optic neuropathy. 90% of IVLBCL with neurological involvement had systemic symptoms, including prolonged fever, anaemia, anorexia and weight loss. Longitudinally extensive spinal cord lesions without or with faint enhancement were the majority in IVLBCL with spinal cord involvement.

The significance of neurological symptoms in intravascular lymphoma cannot be overstated. Previous meta-analyses have reported cognitive impairment, paralysis and seizures in 60.9%, 22.2% and 13.4% of patients, respectively, with the majority showing CNS rather than peripheral nervous system involvement.[7] Comparatively, the neurological symptoms of the haemophagocytic syndrome-associated variant of IVLBCL can range from facial palsy to diplopia and altered mental status.[11]

Clinical and laboratory findings were consistent with the results of previous research.[12–14] Earlier studies reported that nearly half of patients with IVLBCL had systemic symptoms, with fever being the most common. Laboratory abnormalities frequently reported in IVLBCL include elevated high LDH and

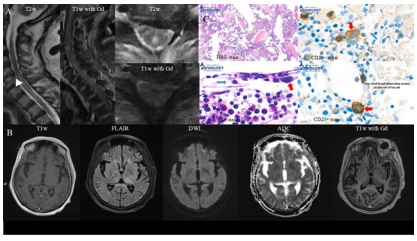

Figure 4 MRI features of patients 8–9 with IVLBCL and pathology of patient 8. (A) Patient 8: This patient presented with longitudinally extensive transverse myelopathy and vertigo. Spinal MRI revealed long segmental intramedullary cord lesions at C2 to T2 levels. (white arrow). (B) Patient 9: presented with cognitive impairment and aphasia. MRI demonstrated a diffuse patchy hyperintense signal in T2 at the bilateral midbrain, pons, bilateral thalamus, bilateral posterior limb of the internal capsule, left basal ganglion and a few small enhancements at the left splenium. (C) Pathological findings of bone marrow patient 8 revealed that H&E-stained showed no cluster or aggregate of lymphoid cells. PAS stain highlighted the large abnormal cell (red arrow) within the vascular lumen (blue arrow). CD20+large lymphoma B-cell (red arrow) flanked by endothelial cells (blue arrow). Gd, gadolinium; IVLBCL, intravascular large B cell lymphoma.

beta-2 microglobulin levels, an elevated erythrocyte sedimentation rate, anaemia, leucopenia and thrombocytopenia. These characteristics can help differentiate IVLBCL from other neurological diseases.

Regarding imaging characteristics, our findings align with the literature which indicates that IVLBCL often presents as hyperintense multifocal brain lesions, potentially mimicking small infarctions.[15 16] Additionally, we observed that IVLBCL can manifest as hyperintense lesions in both supratentorial and infratentorial structures. Remarkably, nearly half of the patients in our study had longitudinally extensive myelopathy, a finding that corroborates several case reports of intravascular lymphoma.[17 18] This distinctive finding should be considered during the differential diagnosis of longitudinally extensive myelopathy, such as neuroinflammatory diseases, particularly neuromyelitis optica spectrum disorder.[19] Furthermore, our investigation revealed the presence of stroke-like lesions—either infarcts or haemorrhages—within the brains of our patients. This finding introduces additional complexity to the diagnosis of stroke.[20] Although the symptoms and laboratory findings would be different from each other.[21]

IVLBCL can sometimes resemble both primary and secondary CNS lymphoma, as they share many clinical features, including systemic symptoms and neurological deficits.[22–24] A potentially distinctive characteristic of IVLBCL is the presence of longitudinally extensive myelopathy which can serve as a diagnostic clue. Primary CNS lymphoma typically affects the supratentorial brain hemispheres (40%), with less frequent involvement of the posterior fossa and spinal cord. In contrast, secondary CNS lymphoma usually presents with parenchymal participation (40–60%), leptomeningeal disease (20–30%) or a combination of both (10%).[25]

The overall survival rates observed in this study were consistent with those reported in prior research for patients with IVBCL. Liu *et al* reported 1-year and 3-year survival rates of 42.3% and 11.5%, respectively, with a median survival time of 340 days.[3] In comparison, our study revealed 1-year and 2-year overall survival rates of 56.5% and 52.7%, respectively. The survival rate of our patients with CNS involvement is lower than that of those without CNS involvement which is consistent with a previous retrospective study.[26] This could be explained by the fact that patients with CNS involvement usually have more widespread disease and tumour burden than those without CNS involvement.

Several limitations of this study must be acknowledged. The low incidence of IVLBCL limited the sample size which may reduce the power to detect differences in clinical, imaging and laboratory features. Variations in the time from symptom onset to diagnosis, from onset to treatment and in treatment protocols may confound overall survival. Future studies should explore the imaging of patients with IVLBCL, especially MRI of the spine, to provide deeper insights into its unique findings and enhance diagnostic accuracy.

This study highlights the distinct clinical and laboratory features of the IVLBCL with neurological involvement. Neurological symptoms accompanying systemic involvement raise suspicion of such a condition. Longitudinally extensive thoracic myelopathy with faint or without enhancement was the hallmark of the patients with IVLBCL. Early recognition of these differences is crucial for accurate diagnosis and improved patient outcomes. Despite the aggressive nature of the IVLBCL, distinguishing it from other conditions can guide appropriate therapeutic strategies and potentially enhance survival rates.

Author affiliations
[1]Faculty of Medicine Siriraj Hospital, Mahidol University Faculty of Medicine Siriraj Hospital, Bangkok, Thailand
[2]Siriraj Neuroimmunology Center, Division of Neurology, Department of Medicine, Faculty of Medicine Siriraj Hospital, Mahidol University, Salaya, Thailand
[3]Division of Neurology, Department of Medicine, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand
[4]Division of Hematology, Department of Medicine, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand

Acknowledgements We thank Dr Sanya Sukpanichnant for providing us with the pathological imaging.

Contributors Conceptualisation: EU, PL, JJ, WO. Data curation: EU, PL. Formal analysis: PL. Funding acquisition: No funding is required. Investigation: EU, PL, JJ, WO. Methodology: EU, WO. Project administration: EU, JJ. Supervision: JJ, WO. Validation: PL, JJ, WO. Visualisation: EU, PL, WO. Writing, original draft: EU, JJ. Writing, review and editing: EU, PL, JJ, WO. Guarantor: JJ. All authors have read and approved the final version of the manuscript for submission.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Not applicable.

Ethics approval The study protocol was authorised by the Siriraj Institutional Review Board (approval number Si-713/2022).

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data are available upon reasonable request.

ORCID iD
Ekdanai Uawithya http://orcid.org/0009-0008-6262-8158

REFERENCES

1 Fonkem E, Lok E, Robison D, *et al*. The natural history of intravascular lymphomatosis. *Cancer Med* 2014;3:1010–24.
2 Loukhnati M, Lahlimi FE, Tazi I. Intravascular Large B-Cell Lymphoma. *Case Rep Hematol* 2023;2023:5596890.
3 Liu Z, Zhang Y, Zhu Y, *et al*. Prognosis of Intravascular Large B Cell Lymphoma (IVLBCL): Analysis of 182 Patients from Global Case Series. *Cancer Manag Res* 2020;12:10531–40.
4 Ponzoni M, Ferreri AJM, Campo E, *et al*. Definition, diagnosis, and management of intravascular large B-cell lymphoma: proposals and perspectives from an international consensus meeting. *J Clin Oncol* 2007;25:3168–73.

5 Breakell T, Waibel H, Schliep S, *et al*. Intravascular Large B-Cell Lymphoma: A Review with a Focus on the Prognostic Value of Skin Involvement. *Curr Oncol* 2022;29:2909–19.
6 Sukpanichnant S, Visuthisakchai S. Intravascular lymphomatosis: a study of 20 cases in Thailand and a review of the literature. *Clin Lymphoma Myeloma* 2006;6:319–28.
7 Fonkem E, Dayawansa S, Stroberg E, *et al*. Neurological presentations of intravascular lymphoma (IVL): meta-analysis of 654 patients. *BMC Neurol* 2016;16:9.
8 Song K-W, Issa S, Batchelor T. Primary central nervous system lymphoma: epidemiology and clinical presentation. *Ann Lymphoma* 2021;5:16.
9 Zuckerman D, Seliem R, Hochberg E. Intravascular lymphoma: the oncologist's "great imitator". *Oncol* 2006;11:496–502.
10 Armitage JO. Staging non-Hodgkin lymphoma. *CA Cancer J Clin* 2005;55:368–76.
11 Yoon SE, Kim WS, Kim SJ. Asian variant of intravascular large B-cell lymphoma: a comparison of clinical features based on involvement of the central nervous system. *Korean J Intern Med* 2020;35:946–56.
12 Ponzoni M, Campo E, Nakamura S. Intravascular large B-cell lymphoma: a chameleon with multiple faces and many masks. *Blood* 2018;132:1561–7.
13 Ponzoni M, Ferreri AJM. Intravascular lymphoma: a neoplasm of "homeless" lymphocytes? *Hematol Oncol* 2006;24:105–12.
14 Orwat DE, Batalis NI. Intravascular Large B-Cell Lymphoma. *Arch Pathol Lab Med* 2012;136:333–8.
15 Baehring JM, Henchcliffe C, Ledezma CJ, *et al*. Intravascular lymphoma: magnetic resonance imaging correlates of disease dynamics within the central nervous system. *J Neurol Neurosurg Psychiatry* 2005;76:540–4.
16 Popiolek C, Gupta K, Huff ML, *et al*. Intravascular large B-cell lymphoma masquerading as stroke successfully treated with R-Hyper-CVAD. *Leuk Res Rep* 2024;21:100401.
17 Tolvaj B, Hahn K, Nagy Z, *et al*. Life threatening rare lymphomas presenting as longitudinally extensive transverse myelitis: a diagnostic challenge. *Ideggyogy Sz* 2020;73:275–85.
18 Kumar N, Keegan BM, Rodriguez FJ, *et al*. Intravascular lymphoma presenting as a longitudinally-extensive myelitis: diagnostic challenges and etiologic clues. *J Neurol Sci* 2011;303:146–9.
19 Kim HJ, Paul F, Lana-Peixoto MA, *et al*. MRI characteristics of neuromyelitis optica spectrum disorder: an international update. *Neurol (ECronicon)* 2015;84:1165–73.
20 Yamamoto A, Kikuchi Y, Homma K, *et al*. Characteristics of intravascular large B-cell lymphoma on cerebral MR imaging. *AJNR Am J Neuroradiol* 2012;33:292–6.
21 Bhagat R, Shahab A, Karki Y, *et al*. Intravascular Lymphoma-Associated Stroke: A Systematic Review of Case Studies. *Cureus* 2023;15:e50896.
22 Poropatich K, Dittmann D, Chen Y-H, *et al*. A Small Case Series of Intravascular Large B-Cell Lymphoma with Unexpected Findings: Subset of Cases with Concomitant Extravascular Central Nervous System (CNS) Involvement Mimicking Primary CNS Lymphoma. *J Pathol Transl Med* 2017;51:284–91.
23 Schaff LR, Grommes C. Primary central nervous system lymphoma. *Blood* 2022;140:971–9.
24 Malikova H, Burghardtova M, Koubska E, *et al*. Secondary central nervous system lymphoma: spectrum of morphological MRI appearances. *Neuropsychiatr Dis Treat* 2018;14:733–40.
25 Pons-Escoda A, Naval-Baudin P, Velasco R, *et al*. Imaging of Lymphomas Involving the CNS: An Update-Review of the Full Spectrum of Disease with an Emphasis on the World Health Organization Classifications of CNS Tumors 2021 and Hematolymphoid Tumors 2022. *AJNR Am J Neuroradiol* 2023;44:358–66.
26 Seegobin K, Li Z, Alhaj Moustafa M, *et al*. Clinical characteristics, prognostic indicators, and survival outcomes in intravascular lymphoma: Mayo Clinic experience (2003-2018). *Am J Hematol* 2022;97:1150–8.