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## Case Report: Intravascular Large B-Cell Lymphoma of the Brain

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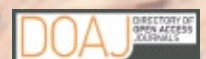
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# Intravascular Large B-Cell Lymphoma of the Brain

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## Case Report

A 61-year-old man with a history of hypertension, dyslipidaemia, and monoclonal gammopathy of undetermined significance was admitted for recurrent strokes.

15 months earlier the patient had presented a sudden left hemiparesis. Brain magnetic resonance imaging (MRI) revealed subacute ischemia in the right corpus callosum. Transthoracic echocardiography, cervical Doppler sonography, electrocardiographic Holter, transoesophageal echocardiography (TOE), immunological tests, and microbiological tests (including tests for syphilis and human immunodeficiency virus) were performed. All of these examinations were normal. He was therefore started on aspirin.

Six months later, the patient presented a recurrence of the left hemiparesis. A new MRI showed new, multifocal, acute infarcts. Compared to the first MRI, the current MRI (Figure 1) showed multiple, hyperintense, multifocal, cortical, or subcortical lesions with hyperintensity on fluid-attenuated inversion recovery (FLAIR).

Due to the possibility of a prothrombotic state anticoagulant treatment was started. Cerebral angiography showed segmentary stenoses of small and medium vessels.

Despite normal inflammatory markers, a possible underlying inflammatory disease (brain vasculitis) as a cause of ischemic damage was considered. The patient was admitted to neurosurgery for cerebrospinal fluid biopsy.

Histopathological examination revealed intravascular proliferation of large atypical lymphoid cells with pleomorphic nuclei and prominent nucleoli (Figure 2a), in the leptomeningeal and cortical vessels (Figure 2b), producing luminal obstruction.

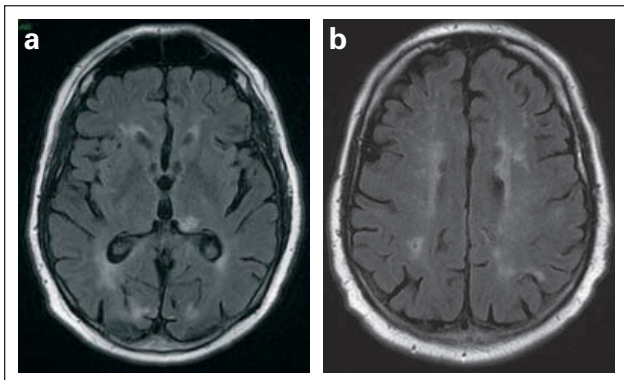


Figure 1. FLAIR MRI depicting the status during the current hospitalisation.

On immunohistochemistry, the neoplastic cells were positive for CD 20 (Figure 2c). The T-lymphocyte marker CD3 showed reactive perivascular T-lymphocytes. The MIB-1 labelling index was > 95 %.

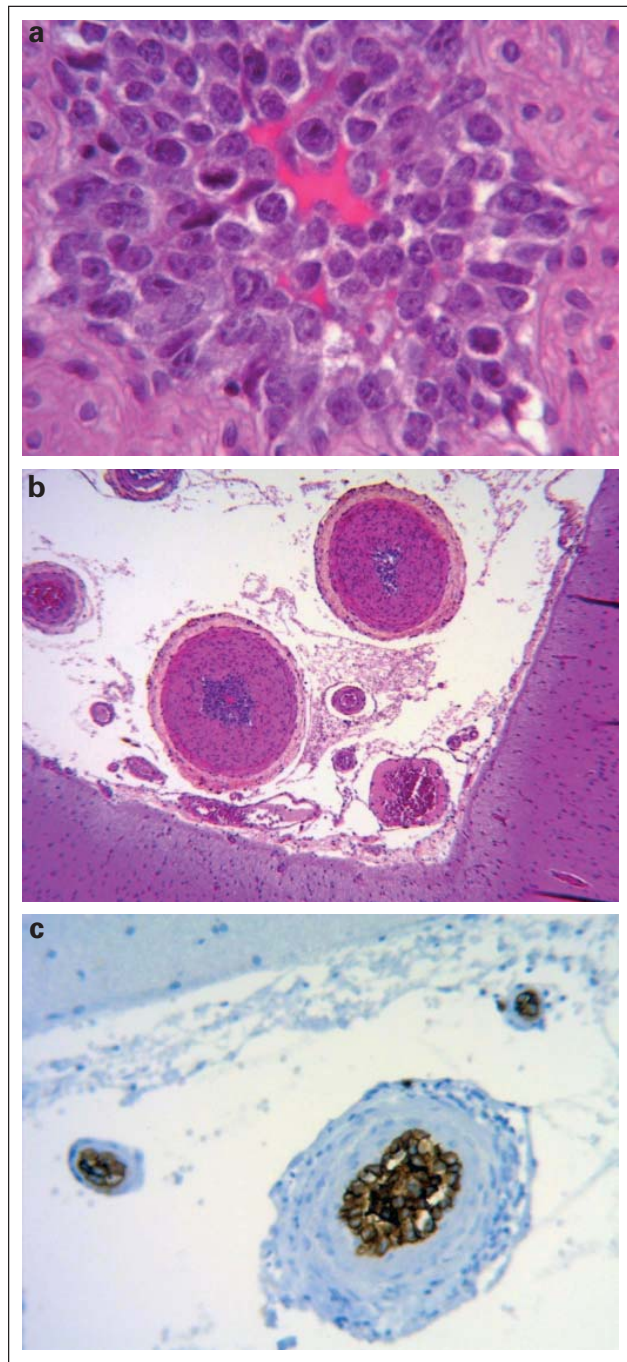


Figure 2. Histopathological and immunohistochemical findings of cerebrospinal fluid biopsy [(a) HES  $\times 400$ ; (b) HES  $\times 50$ ; (c) CD 20  $\times 100$ ].

## ■ Comments

Intravascular large B-cell lymphoma is an uncommon subtype of large-cell lymphoma that is characterised by proliferation of lymphoid cells within the lumina of small and medium blood vessels. The current literature shows that it tends to affect the elderly in their 6<sup>th</sup> to 7<sup>th</sup> decades of life. Presentation is varied, manifesting as rapidly progressive encephalopathy, subacute neurological defects (due to brain infarcts), or headaches.

MRI is the best imaging tool for intravascular lymphoma, revealing abnormalities such as multiple, hyperintense, multifocal, cortical, or subcortical lesions with hyperintensity on T2-weighted and fluid-attenuated inversion recovery (FLAIR) imaging, suggesting small-vessel ischemia. The main differential diagnosis is central nervous system vasculitis.

Conventional cerebral angiography is normal > 50 % of cases but can show multiple segmentary stenoses and dilatations. But it lacks the specificity to differentiate intravascular large B-cell lymphoma from cerebral vasculitis.

Brain biopsy is the diagnostic key of diagnosis if it is large enough, lesions being focal. It shows neoplastic cells with irregular nuclei and prominent nucleoli. Immunohistochemistry shows a CD 20 expression of tumoural cells that are localised in the blood vessels of the leptomeninges, cerebral cortex, and white matter with multifocal vessel occlusion.

The outcome of intravascular lymphoma is very poor with a 1-year mortality rate approximating 80 % from the time of diagnosis. So far, no curative treatment exists.

This case report intends to highlight the key role of histopathology since non-invasive examination did not lead to correct diagnosis.

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### Further Reading:

Cruto C, Taipa R, Monteiro C, et al. Multiple cerebral infarcts and intravascular central nervous system lymphoma: a rare but potentially treatable association. *J Neurol Sci* 2013; 325: 183–5.

Hundsberger T, Cogliatti S, Kleger GR, et al. Intravascular lymphoma mimicking cerebral stroke: report of two cases. *Case Rep Neurol* 2011; 3: 278–83.

Martin-Duverneuil N, Mokhtari K, Behin A, et al. Intravascular malignant lymphomatosis. *Neuroradiology* 2002; 44: 749–54.

Momota H, Narita Y, Miyakita Y, et al. Intravascular lymphoma of the central nervous system presenting as multiple cerebral infarctions. *Nagoya J Med Sci* 2012; 74: 353–8.

Song DK, Boullis NM, McKeever PE, et al. Angiotropic large cell lymphoma with imaging characteristics of CNS vasculitis. *AJNR Am J Neuroradiol* 2002; 23: 239–42.

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