*Case Report*

# Intravascular Lymphomatosis Presenting with Spinal Cord Infarction and Recurrent Ischemic Strokes

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Intravascular lymphomatosis (IVL) is a rare subtype of large B-cell lymphoma that follows an aggressive course with rapidly progressive neurological involvement and potentially fatal outcome.1 We report on a 64-year-old man with progressive myelopathy at T6-T7 and recurrent cerebral infarctions. This case is illustrative of the clinical course that is seen in IVL. It aims to present a timeline of imaging findings that demonstrate the progression of disease and characteristic pathology findings. We emphasize the importance of IVL on the differential diagnosis of spinal cord infarction.

**Key Words:** Stroke—infarction—spinal cord—intravascular lymphomatosis



## Introduction

Intravascular lymphomatosis (IVL) is a progressive disease characterized by the proliferation of lymphoma cells within the lumen of small blood vessels, with a wide variety of neurological manifestations. Diagnosis is usually challenging, often delayed or recognized at autopsy.

## Case Report

A 64-year-old man with hypertension, hyperlipidemia, and degenerative disc disease presented with progressive weakness of the lower extremities, constipation, and urinary dysfunction over a period of 4 weeks. Initial magnetic resonant imaging (MRI) of the cervical, thoracic, and lumbar spine showed severe degenerative disc disease with central canal stenosis at L4-L5. Neurological examination was notable for bilateral lower extremity weakness and gait ataxia. MRI brain showed multifocal acute and subacute scattered infarcts (Fig 1A). Cardiac and arterial imaging was unremarkable. Examination of the cerebrospinal fluid was normal. Computerized tomography (CT) of the abdomen and pelvis was notable for splenomegaly. Laboratory studies showed vitamin B12 and folate deficiency, marginally elevated prothrombin time, and homozygous mutation of the methylenetetrahydrofolate reductase (MTHFR) C677T gene, weakly positive hexagonal phase lupus anticoagulant, reduced antithrombin and protein C activity, elevated factor VIII activity, elevated LDH, positive beta 2 glycoprotein, and polyclonal gammopathy with elevated gamma component, suspicious for multiple myeloma. Infectious workup was noncontributory. Bone marrow biopsy was nondiagnostic with elevated plasma cells of normal morphology and normal flow cytometry. A catheter cerebral angiogram showed no significant changes. He subsequently developed atrial fibrillation and was started on antithrombotic therapy. Despite therapeutic anticoagulation, he continued to deteriorate with increasing confusion, prosopagnosia, dysnomia, color blindness, and lower extremity paraparesis. Repeat MRI brain and spine showed several new cerebral infarctions (Fig 1B), and an acute spinal cord infarction at T6-T7 (Fig 1C-H).

Brain biopsy was consistent with a "double expressor" (myc and bcl2) nongerminal center intravascular LBCL (Fig 1I-K). Patient eventually succumbed to the illness before treatment of IVL was initiated.

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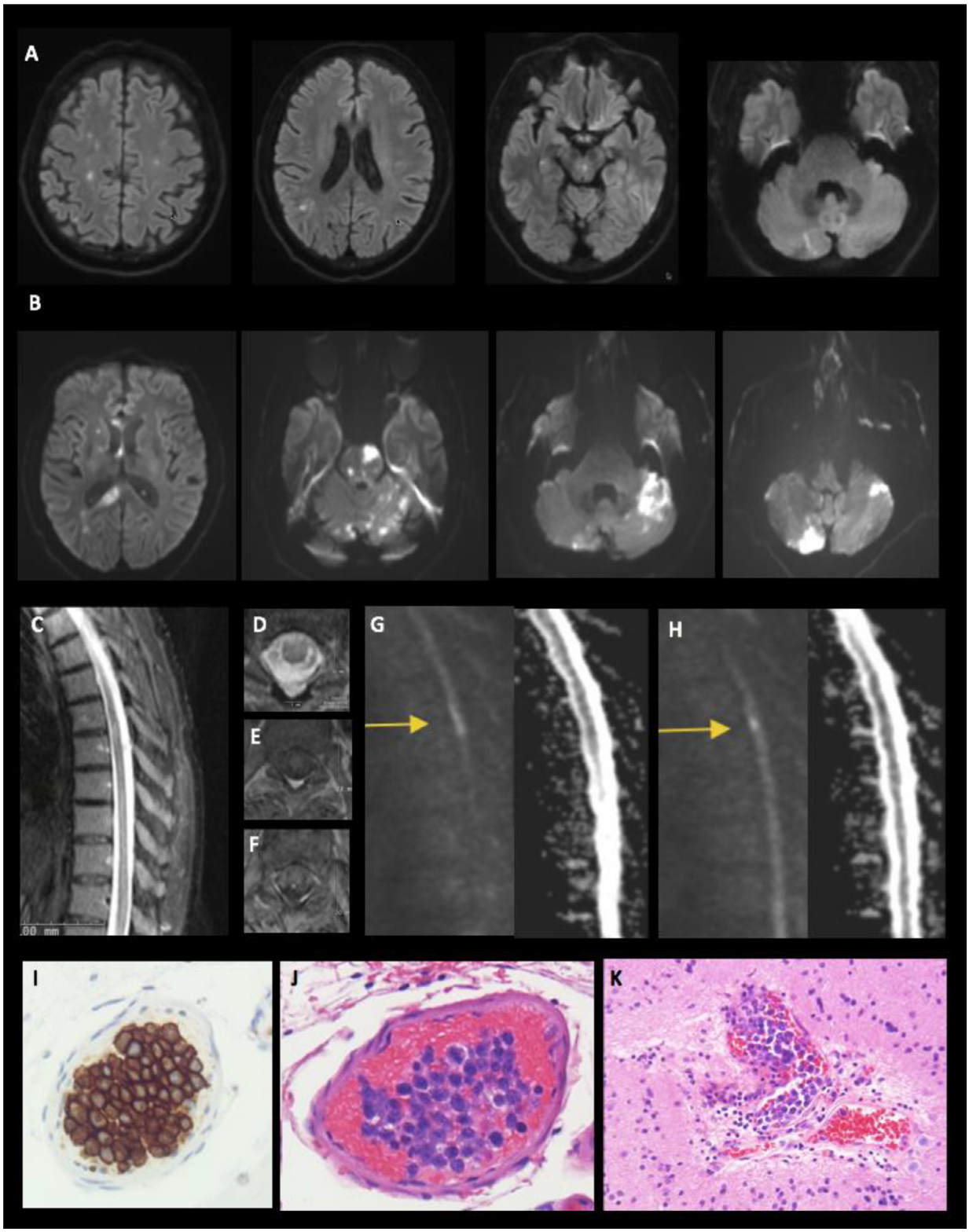


**Figure 1.** *(A) MRI DWI showing several acute infarctions in the deep white matter, brainstem, and cerebellum from patient's initial MRI brain and final MRI brain (B). (C) Sagittal FLAIR sequence with T2 hyperintensity at T6-T7. (D) Axial FLAIR sequence with T2 hyperintensity at T6-T7. (E) Axial T1 and (F) axial T1 postcontrast without corresponding contrast enhancement. (G, H) DWI and ADC sequences with restricted diffusion and subtle ADC correlate at T6-T7. These sequences suggest an acute infarct in the ventral thoracic spinal cord at T6-T7. (I) Histological sections showed medium- and small-sized vessels with intraluminal large CD20-positive B-lymphocytes that were negative for T-cell antigens (not shown), supporting a B-cell lineage malignancy. (J) Large atypical cells with high nuclear to cytoplasmic ratio and large nuclei with vesicular chromatin, which is consistent with lymphoma cells. (K) The surrounding brain parenchyma shows no infiltration by the lymphoma cells.*

## Discussion

CNS involvement of IVL is more common in Western countries compared to Asian countries, which have more systemic involvement (bone marrow, spleen, and liver).[1,2] Spinal cord infarction has been reported as the first manifestation of IVL.[3] Similar to the literature where cord abnormality may only be detected in <60% of patients with spinal cord involvement, our patient had normal initial spine imaging despite progressive paraparesis. When clinical suspicion is high, a repeat MRI may be necessary for early diagnosis and treatment, which could alter the course of the disease.[4–6]

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