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# Fatal Intravascular Large B-cell Lymphoma Mimicking Central Nervous System Vasculitis: A Case Report

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## ABSTRACT

We present a case of a 69-year-old man who presented with one year of cognitive decline, recurrent ischemic strokes, and new onset visual hallucinations. Initially, clinical and radiographic findings raised concern for central nervous system vasculitis, however brain biopsy confirmed a diagnosis of intravascular large B-cell lymphoma.

During hospitalization, Brain MRI revealed multiple acute ischemic strokes in both cerebral hemispheres, a diagnostic brain angiogram showed mild alternating stenosis in branches of right anterior and middle cerebral arteries, and a brain biopsy reported intravascular large B-cell lymphoma.

This case report emphasizes the broad neurologic presentation of intravascular B-cell lymphoma and the urgency of his early diagnosis.

**Keywords:** Fatal intravascular; B-cell lymphoma; Central nervous system vasculitis

## INTRODUCTION

Intravascular large B-cell lymphoma (IVLBCL) is a rare subtype of extranodal non-Hodgkin's lymphoma characterized by proliferation of malignant B lymphocytes within the lumen of small or medium-sized blood vessels[1]. Clinical presentations are variable due to tumors cells invading several different organs, commonly the skin and central nervous system (CNS)[2]. Patients with IVLBCL frequently develop cerebral occlusive ischemia caused by the intravascular foci of malignant lymphocytes mimicking embolic ischemic strokes and central nervous system vasculitis[3].

Due to the heterogenous and aggressive clinical course, rarity of IVLBCL, and lack of specific radiological or laboratory data, the manifestation might be overlooked, and the diagnosis can be difficult, often delayed and misdiagnosed[1,4,5]. This can lead to a delay in treatment which can be devastating as IVCLBCL is rapidly progressive and has a very poor prognosis[6].

We report a case of IVLBCL where the clinical presentation, laboratory, and imaging features mimicked CNS vasculitis.

## CASE PRESENTATION

A 69-year-old man was transferred to our neurology department for evaluation of CNS vasculitis. He had a medical history of rheumatoid arthritis (RA) and ischemic strokes. His RA was well controlled on Metrotexate, prednisone and adalimumab. Strokes were diagnosed two months prior admission and the mechanism was considered embolic stroke of undetermined source (ESUS). He was started on antiplatelet therapy and a high intensity statin, and cardiac loop recorder was implanted.

He presented with left hemiparesis, visual hallucinations, 1-year history of cognitive decline and 60-pound weight loss. He was found to have multiple small acute ischemic strokes in right frontal, bilateral parietal, and bilateral occipital lobes on magnetic resonance imaging (MRI) of the brain (Figure 1A-B). Computed tomography (CT) angiography of brain and neck did not show atherosclerosis or stenosis. Echocardiogram was normal, and interrogation of the loop recorder did not reveal any arrhythmias.

During hospitalization, extensive infectious, autoimmune, and paraneoplastic workup was negative. Lumbar puncture was notable for elevated protein at 83 mg/dl, flow cytometry did not show any abnormality, T-tau protein and 14-3-3 gamma protein were elevated but real-time quaking-induced conversion (RT-QuIC) was negative. EEG showed generalized slowing. CT scan of the chest showed mildly prominent mediastinal lymph nodes. Whole body positron emission tomography (PET) scan was unremarkable.

Two weeks after admission, the patient developed sudden bilateral painless complete vision loss that gradually improved. Brain MRI showed new infarcts in the left frontal and occipital lobes (Figure 1.C-D). Due to concerns about nervous system vasculitis, cerebral angiogram was performed showing mild alternating stenosis in branches of right anterior and middle cerebral arteries (Figure 2). He continued having neurological decline, so he was started empirically on intravenous methylprednisolone without response.

To further investigate the possibility of cerebral vasculitis, he underwent a right frontal lobe biopsy. His biopsy was diagnostic for IVLBCL (Figure 3). The patient and his family declined chemotherapy and opted for hospice care.

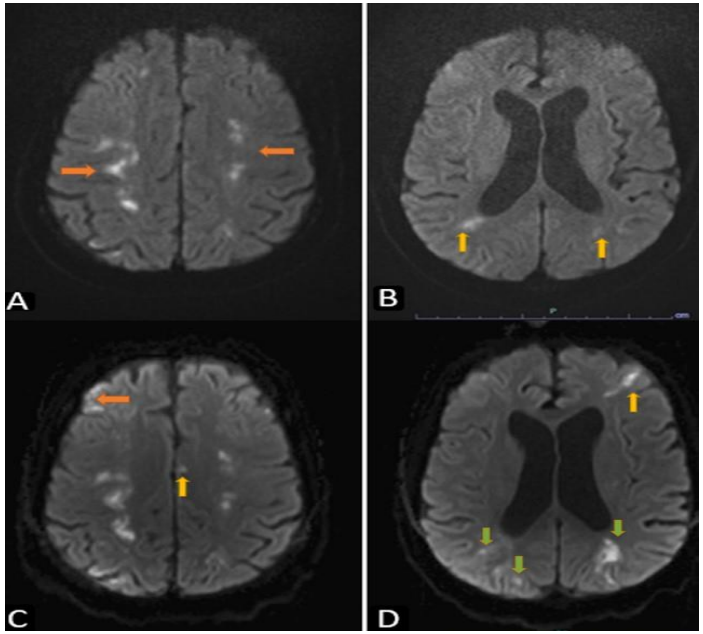


**Figure 1:** Brain MRI at admission and at day 14 of hospitalization.

A: Axial view showing scatter diffusion restriction in DWI consistent with subacute infarctions in the bilateral frontal and parietal lobes (orange arrows). B: Axial view showing scatter diffusion restriction in DWI consistent with subacute infarctions in the periventricular white matter of bilateral posterior temporal lobes (yellow arrows).

C: Brain MRI on day 14 of admission showing new foci of diffusion restriction in left medial frontal lobe (yellow arrow) , and right fontal lobe (orange arrow) consistent with acute infarcts. D: Brain MRI on day 14 of admission showing new foci of diffusion restriction in left subcortical frontal lobe (yellow arrow), and bilateral superior occipital lobes (green arrows) consistent with acute infarcts.

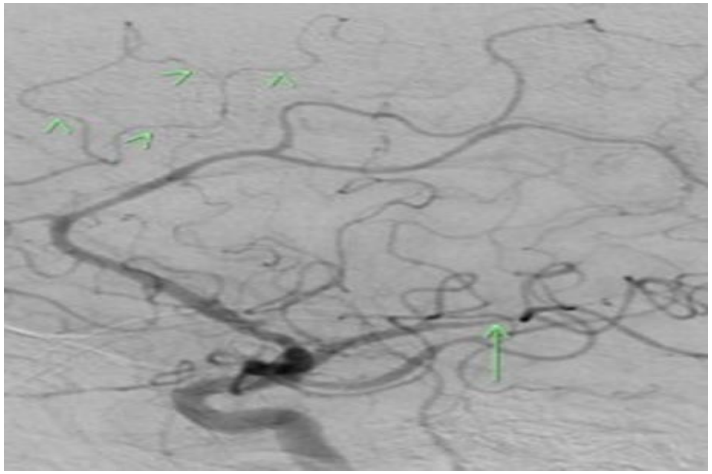

**Figure 2:** Brain angiogram showing mild alternating areas of stenosis in distal ACA branches (head arrows) and a short segment of mild stenosis of right proximal M2 branch (arrow).

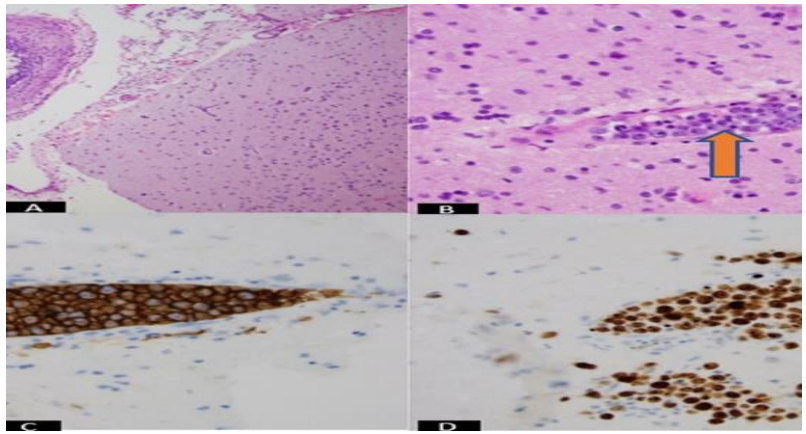

**Figure 3:** Histopathologic examination of the brain biopsy tissue shows leptomeningeal (A) and cortical penetrating vessels involved by enlarged, malignant-appearing lymphoid cells (B, orange arrow) . The cells were positive for CD20 (C) and showed high proliferative index by Ki67 (D) on immunostains, supportive of a large B-cell lymphoma (A: hematoxylin and eosin stain, overall 100X magnification; B: hematoxylin and eosin stain, overall 400X magnification; C: CD20 immunostain, overall 400X magnification; D: Ki67 immunostain, overall 400X magnification)

## DISCUSSION

IVLBCL is a rare disease and almost half of cases have central nervous system (CNS) involvement, which is associated with worse clinical outcomes and survival rates as appreciated in our patient[2]. The most common CNS manifestations are cognitive impairment/dementia, seizures, and ischemic strokes. In regards to differential diagnoses, it can mimic cerebral vasculitis, encephalopathy, or infections of the CNS. This wide variety of neurological presentations can lead to misdiagnoses and delay treatment[2].

Clinically and radiologically, IVLBCL can mimic CNS vasculitis. Interestingly, the segmental areas of stenosis and dilatation referred to as 'bead pattern' in brain angiogram, classically associated with vasculitis, are also seen in 45% of IVLBCL cases which makes the diagnosis more difficult[6,7].

We noticed a progressive neurologic decline even after starting steroids, which has been documented in others case reports[6,7]. This may reflect a lack of inflammation and favor the proliferation of malignant B lymphocytes within the lumen of blood vessels as main pathophysiology of symptoms[1].

Brain biopsy is the diagnostic procedure of choice in suspected IVLBCL affecting the CNS, as CSF analysis, laboratory, and radiologic sings lack specificity and sensitivity to differentiate IVLBCL from cerebral vasculitis. In this case, there was a delay in the diagnoses, which is a common denominator according to published literature[1,3-6]. Regarding treatment, immunochemotherapy can substantially lead to the prevention of devastating strokes and may induce durable complete remission as demonstrated in other case reports[6].

Our thorough review of the literature revealed that only a few cases, similar to our patient, had clinical, laboratory and imaging features that could be seen in either CNS vasculitis or IVLBCL[6-7].

## CONCLUSION

In conclusion, we hope this case report encourages the early consideration of IVLBCL for patients with clinical manifestation of CNS vasculitis, as the treatment is different, and remission can be accomplished.

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