

Intravascular Large B-Cell Lymphoma and the Use of R-Hyper-CVAD in Cases with Central Nervous System Involvement

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Introduction

Intravascular Large B Cell Lymphoma (IVLBCL) is exceptionally rare. This lymphoma presents with a wide range of symptoms that vary depending upon the organ systems affected and it can be quite challenging to diagnose. It is characterized by proliferation of large lymphoma cells within the lumina of small blood vessels [1]. R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisone) is classically considered the therapy of choice in this disease, but efficacy is unclear especially in cases with Central Nervous System (CNS) involvement. This is because this regimen does not cross the blood brain barrier, while R-Hyper-CVAD does [2]. Thus, when a patient with recurrent strokes and IVLBCL was referred to our clinic for management, it was ultimately decided to utilize R-Hyper-CVAD for management. The central nervous system involvement in IVLBCL poses significant diagnostic and therapeutic challenges. CNS symptoms can range from cognitive impairments and psychiatric disturbances to focal neurological deficits and seizures. The presence of lymphoma cells within the CNS complicates treatment, as the blood-brain barrier limits the efficacy of many standard chemotherapeutic agents [3].

Description

The patient we describe was a 56-year-old male who had presented for stroke symptoms at a local hospital multiple times. These symptoms included hearing loss, right facial paresthesia, vertigo, blurry vision, impaired depth perception and tinnitus. Various MRIs of the brain repeatedly demonstrated subacute changes in numerous regions of the brain: including the pons, frontal lobe, parietal lobe and occipital lobe. Thromboembolism and hypercoagulability workups were negative and no source for these symptoms was immediately identified. One repeat angiogram was however suggestive for vasculitis and prednisone was started although later workup for vasculitis was negative as well.

The patient at this time also had a CT chest/abdomen/pelvis which demonstrated a right kidney mass that was concerning for renal cell carcinoma. Once the patient was stabilized, he underwent right partial nephrectomy and IVLBCL was diagnosed along with concurrent oncocytoma. 8 cycles of R-Hyper-CVAD were given over 8 months and the patient had an excellent response. Further imaging over the next 4 years has demonstrated no recurrence and he has had no more stroke symptoms.

IVLBCL has a guarded prognosis. This is certainly not aided by delays in diagnosis. Over half of patients with IVLBCL are diagnosed post-mortem and median survival time in 182 cases studied was 340 days [4]. R-CHOP is the most studied chemotherapy regimen in IVLBCL, but given the rarity of the disease, sample sizes are often small in most research. Since CNS involvement in IVLBCL has been rarely explored, use of R-Hyper-CVAD was chosen in our patient because of the guarded prognosis and the severity of his stroke symptoms [5]. It was felt that by providing therapy that crossed the Blood Brain Barrier (BBB), he would be best treated this way. R-Hyper-CVAD has been previously utilized to treat IVLBCL with pontine involvement, however this patient had no neurological symptoms unlike the patient we describe [6].

Conclusion

Overall, IVLBCL presents in a heterogenous manner that may include stroke symptoms. We treated a patient presenting this way with R-Hyper-CVAD and he has remained without recurrence for 5 years since diagnosis. Further research needs to be performed on therapies of choice in IVLBCL, as data is currently limited particularly in cases with CNS involvement.

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Received: 23 July, 2024, Manuscript No. JBL-24-142871; Editor assigned: 25 July, 2024, PreQC No. JBL-24-142871 (PQ); Reviewed: 08 August, 2024, QC No. JBL-24-142871; Revised: 12 February, 2025, Manuscript No. JBL-24-142871 (R); Published: 19 February, 2025, DOI: 10.37421/2165-7831.2025.15.337

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How to cite this article: Popiolek, Christopher, Ranju Gupta. "Intravascular Large B-Cell Lymphoma and the Use of R-Hyper-CVAD in Cases with Central Nervous System Involvement." *J Blood Lymph* 15 (2025): 337.