ISSN 2689-8268 | Volume 6

American Journal of Surgery and Clinical Case Reports

Case Report Open Access

Intravascular Large B-Cell Lymphoma (IVLBCL) Presenting with CNS Involvement in Patient with Chronic Lymphocytic Leukemia

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Received: 13 Apr 2023
Accepted: 30 May 2023
Published: 08 June 2023
J Short Name: AJSCCR

Citation:

Balodis A. Intravascular Large B-Cell Lymphoma (IVLBCL) Presenting with CNS Involvement in Patient with Chronic Lymphocytic Leukemia. Ame J Surg Clin Case Rep. 2023; 6(12): 1-4

Keywords:

Magnetic Resonance Imaging; Intravascular Large B-Cell Lymphoma

1. Abstract

1.1. Background: Intravascular lymphoma (IVL) is a rare, often fatal disease characterized by intraluminal proliferation of lymphoid cells within blood vessels. Intravascular Large B-Cell Lymphoma (IVLBCL) is frequently found in different organs, but the skin and central nervous system (CNS) are the most affected ones. Routine radiological examination and additional diagnostic tools, such as high-resolution, three-dimensional (3D) FLAIR sequence, T2-weighted (T2W) and T1-weighted (T1W) contrast-enhanced magnetic resonance imaging (MRI), MRI diffusion and perfusion imaging studies are useful to confirm the diagnosis in cases where it is not easy to perform histopathological examinations [1].

1.2. Case Report: We present a clinical case of a 63-year-old female patient who presented with progressive cognitive deficit as mild motor aphasia, acalculia, agraphia, dyslexia and mild ideomotor apraxia without any signs of other organ involvement. It was known that the patient had 5-year anamnesis of chronic lymphocytic leukemia, stage II and splanchnic venous thrombosis 2 months ago. Blood biochemical analysis without any significant change, except leukocytosis in full blood count (leukocytes 15,2 x 10*9/L, where neutrophils were in normal range, but lymphocytes were increased). Head magnetic resonance imaging (MRI) showed pathological signal areas on the left parietal lobe with hypointense signal on contrast-enhanced T1-weighted, hyperintense signal on contrast-enhanced T2-weighted and FLAIR-weighted MRI images with restricted diffusion and slightly lower ADC value. Magnetic resonance perfusion imaging studies did not show an increase in cerebral blood flow or in cerebral blood volume. In this case, the patient's diagnosis was made based on typical radiological features.

1.3. Conclusions: Intravascular large B-cell lymphoma is a rare subtype of extra nodal large B-cell lymphoma with an aggressive clinical course. The diagnosis is challenging, as it presents without any obvious tumour mass or lymphadenopathy. [2] As the clinical presentation is variable without specific characteristics, mostly biopsy or even autopsy confirms the definite diagnosis. But in some cases, the diagnosis can be made by typical radiological findings in the restricted availability of brain biopsy.

2. Introduction

Intravascular lymphoma (IVL) is a rare, often fatal disease characterized by the intraluminal proliferation of lymphomatous cells within the blood vessels [3]. Intravascular large B-cell lymphoma (IVLBCL) accounts for 1% of B-cell lymphomas. From the Surveillance, Epidemiology, and End Results (SEER) data, the incidence of IVLBCL is estimated to be 0.095 per 1000 000 per year. The increase of IVLBCL incidence is due to an improved awareness of the disease and more precise diagnostic facilities [6]. The median age at the time of the disease presentation is 70 years (ranging 34-90 years). Men and women are equally affected [7].

IVLBCL is frequently found in multiple organs, but the skin and CNS are the most affected ones. [5] Overall, the prognosis is poor with a high mortality rate, average survival times in some cases between 5 and 7 months. However, recent data suggest that 50% 5-year survival can be achieved if treatment is initiated in the early stages of the disease [4]. The accurate diagnosis of isolated CNS

involvement of IVLBCL is challenging and depends primarily on histopathological examination due to the lack of specific clinical manifestations, laboratory markers, and radiological features. Appropriate treatment can improve outcomes, making a timely and correct diagnosis crucial for patients with IVLBCL [8].

The main differential diagnosis for intravascular large B-cell lymphoma includes intravascular natural killer cell (NK) lymphomas, intravascular T-cell lymphomas, and intralymphatic ALK-negative Anaplastic Large Cell Lymphomas (ALCL) [4]. Also IVLBCL should be differentiated from central pontine myelinolysis, posterior reversible encephalopathy syndrome, inflammatory diseases, such as infections, autoimmune vasculitis, IgG4-related diseases and age-related changes [10].

3. Case Report

We present a clinical case of a 63-year-old female patient who presented with progressive cognitive deficit as mild motor aphasia, acalculia, agraphia, dyslexia and mild ideomotor apraxia without any signs of other organ involvement. It was known that the patient had 5-year anamnesis of Chronic Lymphocytic Leukaemia (CLL), II stage and splanchnic venous thrombosis 2 months ago. Blood biochemical analysis without any significant change, except leukocytosis in full blood count (leukocytes 15,2 x 10*9/L, where neutrophils were in normal range, but lymphocytes were increased). Additionally, an analysis on anti-HIV 1/2, anti-HCV, HbsAg, RPR, and TPHA syphilis tests were negative. Lumbar puncture showed no significant changes - cell count 0, protein 0.57 g/L. Initial head Computed Tomography (CT) showed a small hypodense zone in the left parietal lobe.

Head Magnetic Resonance Imaging (MRI) was indicated for further investigation. MRI of the brain showed pathological signal areas on the left parietal lobe with hypointense signal and patchy enhancement on contrast-enhanced T1-weighted (Figure 1), hyperintense signal on T2-weighted (Figure 2) and FLAIR-weighted (Figure 3) MRI with restricted diffusion (Figure 4) and slightly lower ADC value (Figure 5). Magnetic resonance perfusion imaging studies did not show an increase in cerebral blood flow (Figure 6) or in cerebral blood volume (Figure 7).

The diagnosis was made based on clinical picture, comorbidities, and MRI findings, as brain biopsy was not available. Unfortunately, the patient died in 3 months after the IVLBC was diagnosed.

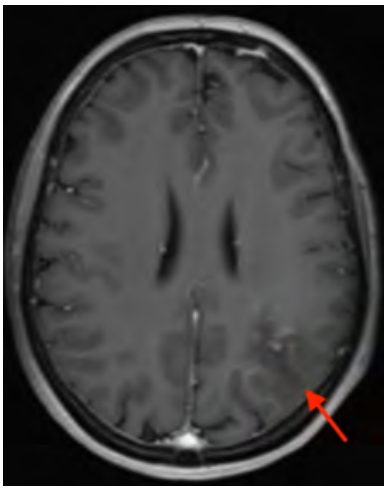


Figure 1: Axial contrast-enhanced T1-weighted magnetic resonance imaging (MRI) showing hypointense signal areas with patchy enhancement on the left parietal lobe.

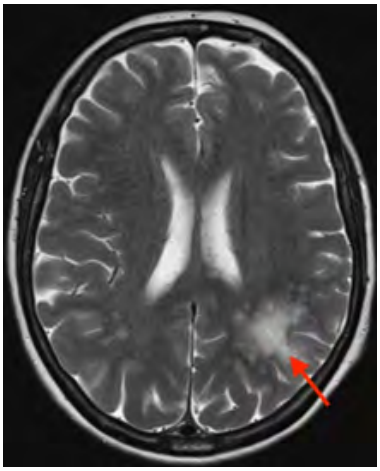


Figure 2: Axial contrast-enhanced T2-weighted MRI showing hyperintense signal areas on the left parietal lobe with small peripheral edema.

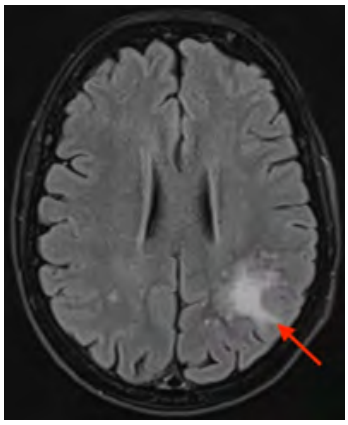
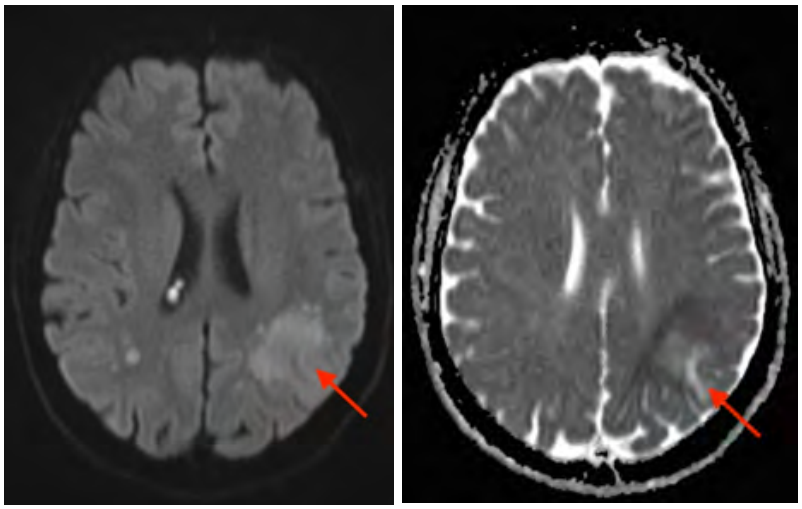
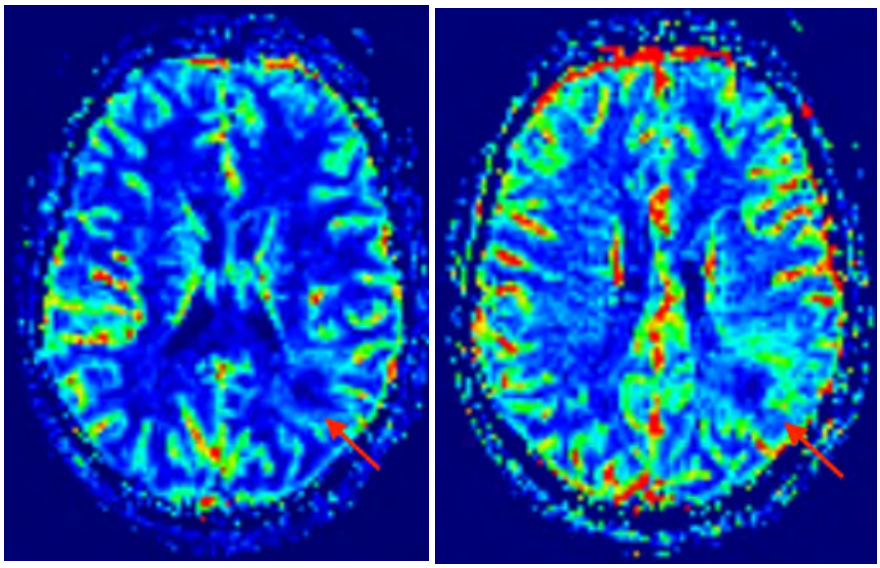


Figure 3: Axial FLAIR-weighted MRI showing hyperintense signal areas on the left parietal lobe with small peripheral edema.



Figures 4 and 5: Axial diffusion weighted (DW) MRI showing restricted diffusion with slightly lower ADC value.



Figures 6 and 7: Magnetic resonance perfusion imaging (PWI) studies did not show an increase in cerebral blood flow (CBF) or in cerebral blood volume (CBV).

4. Discussion

Intravascular large B-cell lymphoma is a rare subtype of extra nodal large B-cell lymphoma with an aggressive clinical course. [2] In this case, despite timely diagnosis and applied therapy, the patient died 3 months after IVLBC was diagnosed. The disease progresses mainly with nonspecific symptoms such as fever, respiratory symptoms, and general fatigue which makes timely diagnosis very difficult. The main clinical sign of IVLBCL is a lack of lymphadenopathy. More than 60% of patients with IVLBCL are reported to develop neurological symptoms, such as encephalop-

athy, stroke, seizure, myelopathy, radiculopathy, and neuropathy, during the clinical course of the disease. [8] Presentation of intravascular lymphoma arising as Richter's syndrome in the setting of previously stable CLL is extremely rare. To the best of our knowledge, there are only several reports of IVLBCL associated with CLL [11, 12]. Richter's syndrome occurs in 2–8% of patients with CLL, typically manifesting as de novo or transformed diffuse large B-cell lymphoma (DLBCL) [13]. Abnormal findings on brain MRI are found approximately 90% of patients with IVLBCL. [9] Brain MRI findings in patients with IVLBCL have been categorized into 4 patterns: (1) Nonspecific white matter lesions; (2) Infarct-like lesions; (3) Hyperintense lesions in the pons on T2-weighted imaging (T2WI); (4) Meningeal thickening and/or enhancement. [8] Characteristic MRI findings for IVLBCL are - T1WI: multifocal hypointense lesions; T2WI: hyperintensities in deep white matter with mild surrounding edema; FLAIR: homogeneously iso-/hypointense, may be hyperintense; DWI: restricted diffusion with low ADC values; PWI: low CBV and CBF ratios; T1WI C+: variable enhancement [14, 15]. Lymphoma confirmed by histological analysis is a common diagnostic standard around the world. [16] In this case, diagnosis was made based on clinical presentation, anamnesis and typical radiological characteristics as brain biopsy was not available.

5. Conclusion

Intravascular large B-cell lymphoma is a rare subtype of extra nodal large B-cell lymphoma with an aggressive clinical course. [2] As the clinical presentation is variable without specific characteristics, mostly biopsy or even autopsy confirms the definite diagnosis. But in some cases, diagnosis can be made by typical radiological findings in restricted availability of brain biopsy. Awareness of this disease and high suspicion can lead to correct diagnosis.

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