

Secondary CNS Lymphoma: A Case of Diffuse Large B-Cell Lymphoma Diagnosed through CNS Involvement

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Abstract

There are 2 major subtypes of CNS lymphoma: secondary CNS lymphoma (involvement of the CNS by systemic lymphoma) and Primary CNS Lymphoma (PCNSL). Two-thirds of secondary CNS lymphomas have leptomeningeal involvement, while parenchymal CNS lymphoma represents one-third of secondary CNS lymphomas and almost 100% of PCNSLs. We present a case of a 49-year-old male patient that was diagnosed with systemic lymphoma that presented initially with neurological symptoms (secondary CNS lymphoma). Lymphoma comprises a heterogeneous group of diseases; important advances have been made in diagnosis and treatment. Imaging features may help distinguish between primary and secondary CNS lymphoma, playing an important role in planning therapy and prognostication.

Keywords: Therapy; Cancer; Lymphoma; Imaging; Brain lesions

Introduction

The World Health Organization International Classification of Disease recognizes more than 50 types of lymphoma based on histopathologic, immunohistochemical, cytogenetic, and molecular analyses [1]. Lymphoma of the CNS consists of 2 major subtypes: secondary CNS involvement by systemic lymphoma, the most common, and PCNSL, when the lymphoma is restricted to the brain, leptomeninges, spinal cord, or eyes, without evidence of it outside the CNS [2].

The risk of CNS involvement in aggressive NHL is 2%-27%, while patients with Hodgkin lymphoma are at risk of 0.5%. An increase of CNS involvement is increased in patients with extranodal involvement and those with primary or acquired immunodeficiency disorders [2].

The incidence rates of PCNSL are increasing among immunocompetent patients, and immunocompromised patients have an increased risk of developing PCNSL. Early diagnosis of CNS lymphoma is crucial for proper management in both immune-competent and immune-compromised [3].

Case Presentation

A 49-year male patient was transferred to our hospital with a 2-week history of convergent strabismus of the left eye, and weakness on the left side of his body which had worsened significantly over the previous 10 days. The patient had been previously diagnosed with hypertension and was not under treatment of any other condition when admitted. He had no history of congenital immunodeficiency disease, previous organ transplantation, or immunosuppressive therapy, and did not have AIDS. His family history was unremarkable for cancer or any immunodeficiency. Further investigation revealed a weight loss of 20 kg in 7 months and pancytopenia. The patient was submitted to a bone marrow biopsy that revealed infiltration by diffuse large B cells lymphoma.

A contrast-enhanced Magnetic Resonance Image (MRI) scan of the brain demonstrated expansive/infiltrating deep frontal masses associated with edema, the biggest one on the left cerebral hemisphere (Figures 1 and 2). Postgadolinium T1-weighted MR imaging showed intense, homogeneous enhancement of the deep left-frontal tumor (Figure 3). In addition, the MRI revealed bilateral lesions within the

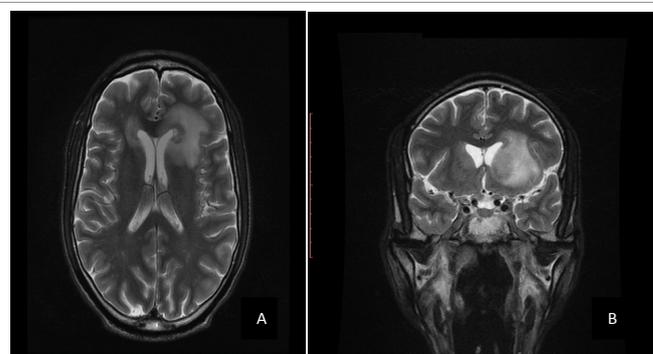


Figure 1: (A) T2-weighted MR imaging axial; (B) Coronal depicts a deep left-frontal mass.

cavernous sinus, involving the V, VI, VII, VIII, IX and X cranial nerves and the walls of the fourth ventricle. Infiltration of the pituitary stalk was also observed (Figure 3).

Discussion

Lymphoma may be unifocal, multifocal, or diffuse, affect isolated lymph nodes or any organ system, and demonstrate a range of imaging appearances at almost every site [1]. Central nervous system lymphoma (CNSL) is an aggressive and rare brain neoplasm that may involve the brain, meninges, spinal cord, and eyes. CNSL is divided into 2 subtypes, Primary Central Nervous System Lymphoma (PCNSL) and Secondary Central Nervous System Lymphoma (SCNSL). In PCNSL, the Central Nervous System (CNS) is primarily involved without systemic

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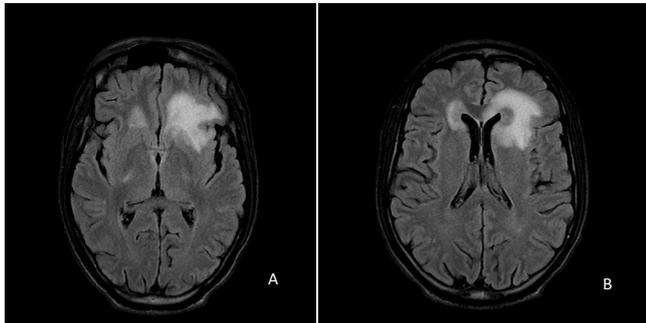


Figure 2: Axial T2-weighted fluid-attenuated inversion recovery (FLAIR) MR imaging depicts the extent of edema to better advantage.

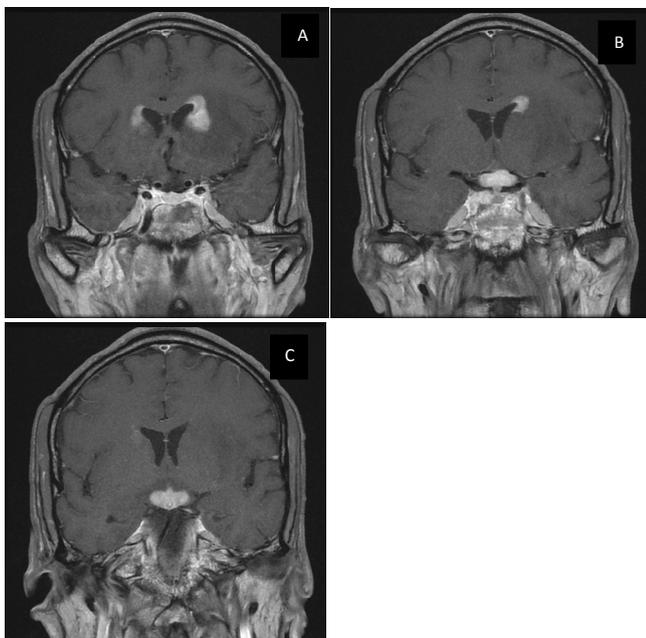


Figure 3: (A and B) Postgadolinium T1-weighted MR imaging shows intense, homogeneous enhancement of the deep frontal tumors; (B and C) Bilateral lesions involving the cavernous sinus, and infiltration of the pituitary stalk.

lymphoma presence. SCNSL is defined as secondary CNS involvement in patients with systemic lymphoma; this type has also been referred to as metastatic lymphoma [3].

The incidence of SCNSL in the general population ranges from 4 to 23%. The factors that most significantly influence the incidence rate of SCNSL include the variant of primary lymphoma, the involvement of more than one extranodal site, a serum lactate dehydrogenase level greater than three times the normal limit, an advanced stage of the systemic disease, and a high International Prognostic Index [4].

The characteristic clinical presentation of SCNSL is a new onset headache (50%), palsies of cranial nerves III, IV, VI, and VII, changes in mental status (29%), and even coma and seizures (23%-29%). It typically presents within 6 months of diagnosis of the primary lymphoma, which is typically confirmed with CSF studies and imaging [4].

CNS involvement in aggressive NHL tends to occur early at a median

of 5-12 months after the primary diagnosis of NHL. Approximately two-thirds of the patients present with leptomeningeal spread and one-third, with parenchymal disease. Leptomeningeal spread, similar to leptomeningeal metastases from any cause, often involves the cranial nerves, spinal cord, or spinal roots and may present as cranial or spinal neuropathies [2].

There are variable SCNSL appearances at the time of initial diagnosis. It can appear as a solitary lesion, a single lesion in either brain parenchyma, focal meningeal, or ependymal localization, or single cranial nerve involvement. The continuous spread through several neuronal structures was considered as a solitary lesion. It can also appear as multiple lesions when there is a combination of 2 or more solitary lesions mentioned above [3].

The diffuse infiltrative type is characterized by extensive continuous infiltration of both white and gray matter by non-enhancing and/or enhancing tumorous involvement (from homogeneous to patchy, worm-like, stripy, misty, etc.), which spread along large white matter tracts and continuously affected at least 2/3 of 1 cerebral hemisphere; and/or different extents of both supratentorial and infratentorial regions; and/or different extents of both hemispheres [3].

The meningeal/ependymal involvement appears as a thickening and enhancing meningeal/ependymal surfaces, which can be smooth, irregular, or nodular, while cranial nerve involvement as a thickening of cranial nerves and their T2 hyperintense and enhancing portions, which can be smooth, irregular, or nodular [3]. The imaging technique of choice to detect leptomeningeal metastasis is contrast-enhanced MR imaging. CT is less sensitive, especially in patients with hematologic malignancies [1].

Although CNS lymphomas may have characteristic imaging findings on traditional CT and MR imaging, none of these will unequivocally differentiate CNS lymphoma from other brain lesions. A visible tumor on imaging is essential to raise the suspicion of CNS lymphoma, which then can lead to an early histologic diagnosis based on cytology of the CSF or brain biopsy [1].

Conclusion

When CNS lymphoma is suspected, contrast-enhanced MR is the imaging technique of choice. Diagnostic imaging plays a critical role in the initial evaluation, monitoring, and follow-up of lymphoma patients. Knowing potential variations of appearance and pitfalls in imaging interpretation will allow the radiologist to provide added value to the report. In some cases, the MRI appearance of lymphoma is tricky as we have demonstrated separately. A multidisciplinary approach for the next research is recommended.

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