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Case Report

Multifocal Lateral and Fourth Ventricular Primary Central Nervous System Lymphoma: Case Report and Literature Review

Multifokal Lateral ve Dördüncü Ventriküler Primer Merkezi Sinir Sistemi Lenfoması: Olgu Sunumu ve Literatür Derlemesi

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ABSTRACT

We report a 66-year-old man with 2-month history of dizziness and 1-month history of diplopia. Cranial magnetic resonance imaging (MRI) disclosed two solid masses, one in the right lateral ventricle and another in the fourth ventricle. A surgical biopsy was performed on the basis of safety. The diagnosis of large B-cell lymphoma was made postoperatively. The patient recovered without additional deficits and was then commenced on chemotherapy and remained well 6 months after the diagnosis. Primary B-cell lymphomas should always be included in the list of differential diagnosis of intraventricular tumors. Here we report an extremely rare case of primary central nervous system lymphoma with multifocal ventricular involvement.

KEYWORDS: Primary CNS lymphoma, Multifocal, Cerebral ventricle neoplasms

ÖZ

İki aylık baş dönmesi ve 1 aylık çift görme öyküsü olan 66 yaşında bir erkek hasta sunuyoruz. Kraniyal manyetik rezonans görüntüleme (MRG), biri sağ lateral ventrikül ve diğeri dördüncü ventrikülde olmak üzere iki solid kitle gösterdi. Emin olmak için cerrahi biyopsi yapıldı. Postoperatif olarak büyük B hücreli lenfoma tanısı kondu. Hasta ek bir defisit olmadan iyileşti ve kemoterapiye başlandı. Hasta tanıdan 6 ay sonra iyi durumdadır. Primer B hücreli lenfomalar intraventiküler tümörlerin ayırıcı tanısında daima dikkate alınmalıdır. Burada multifokal ventriküler tutulumlu çok nadir bir primer merkezi sinir sistemi lenfoma olgusu sunuyoruz.

ANAHTAR SÖZCÜKLER: Primer MSS lenfoma, Multifokal, Serebral ventrikül neoplazmları

INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is a rare malignant tumor, and PCNSLs located in the cerebral ventricles in immunocompetent patients are extremely rare. The standard treatment for PCNSL is biopsy followed by systemic chemotherapy with or without whole brain radiotherapy or intrathecal chemotherapy (4, 5). Here we report a case of PCNSL located in both lateral and fourth ventricles and that was successfully diagnosed by surgical exploration and biopsy. The patient was then commenced on chemotherapy and remained well 6 months after the diagnosis. In conclusion, primary B-cell lymphomas should always be included in the list of differential diagnosis of intraventricular tumors.

CASE REPORT

A 66-year-old man presented to our hospital with 2-month history of dizziness and 1-month history of diplopia. Neurological examination demonstrated incomplete abduction of the left eye and complete left peripheral facial paralysis. The rest of the clinical examination revealed normal findings. His initial blood test was unremarkable. Lumbar puncture with cytological studies revealed no abnormal cell. Cranial magnetic resonance imaging (MRI) disclosed two solid masses, one in the right lateral ventricle and another in the fourth ventricle both with contrast enhancement and mild surrounding edema (Figure 1A-C). No intralesional calcification or hemorrhage was found. According to these radiological features, the diagnosis of ependymoma/metastasis/PCNSL was suspected. A normal computed tomography (CT) scanning chest, abdomen further helped to exclude an extra-cranial malignant lesion. A surgical exploration and biopsy was performed. Postoperative histopathological examination showed infiltration of medium and large cells with hyperchromatic nuclei. Immunohistochemistry revealed that tumor cells express CD79a and CD20 (B-cell markers) (Figure 2A,B). Thus, diagnosis of large B-cell lymphoma was made. Postoperatively, the patient recovered without additional deficits and was then commenced on chemotherapy and remained well 6 months after the diagnosis.

DISCUSSION

Intra-ventricular PCNSLs are very rare tumors and so far, less than 10 cases have been described in literature (2, 3, 6, 8-10, 12), most of which with single cerebral ventricular involvement. We report an extremely rare case that PCNSLs were located both in lateral and fourth ventricle. According to review of literature, PCNSLs are mostly located in the brain hemisphere (38%), thalamus/basal ganglia (16%), corpus callosum (14%) and peri-ventricular regions (12%) (5), with a single tumor in 60–70% of patients and multiple in the remainder (4). For tumors in the fourth ventricle, medulloblastoma (commonly in chidhood), ependymoma and astrocytoma is most frequent (11). The typical imaging features of PCNSL is not characteristic, usually showing hypo- or isointense lesions on T1-weighted MRI, iso- or hyperintense lesions on T2-weighted MRI and most lesions show moderate-to-marked contrast enhancement on MR imaging with variable surrounding edema (7). It was generally accepted that surgical gross resection does not result in a clinical benefit and should thereby be avoided (1, 5). The diagnosis of PCNSL is usually suggested to be established by biopsy and further treated by chemotherapy and radiotherapy. To conclude, primary

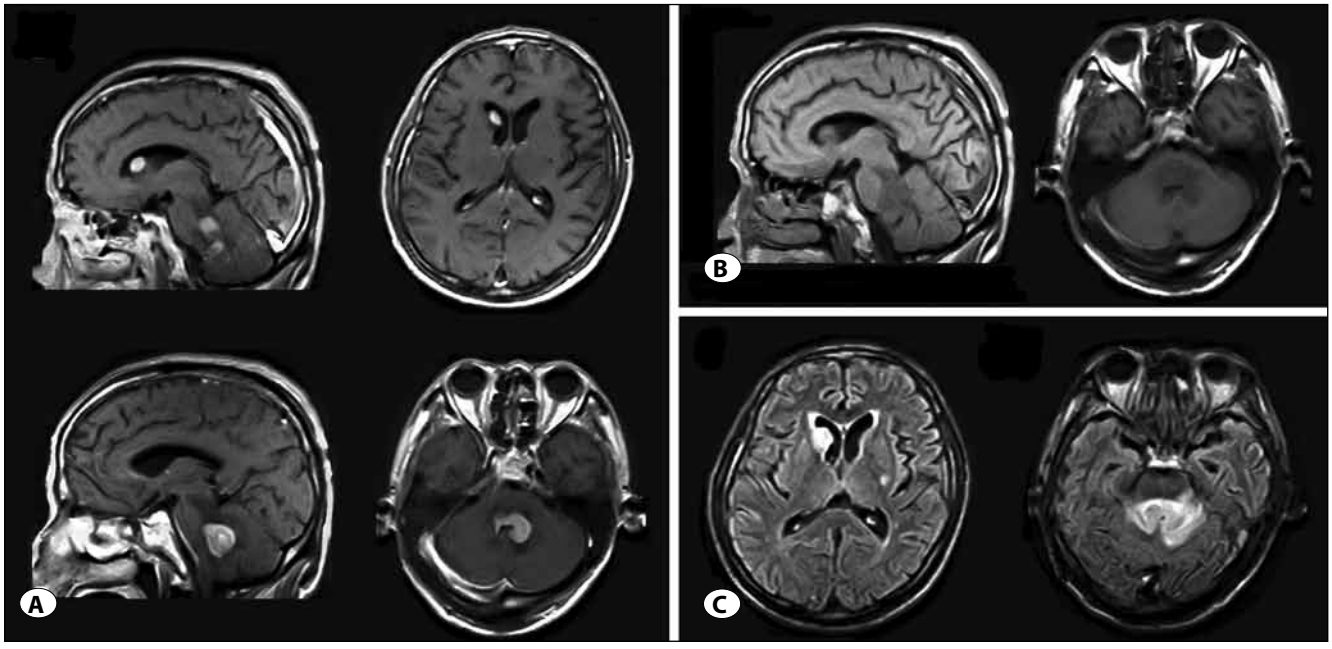

Figure 1: A) Sagittal (left) and axial (right) T1-weighted contrast MRI showed two well-demarcated, enhanced lesions, one in the right lateral ventricle and another in the fourth ventricle. **B)** The lesions are hypointense on T1-weighted MRI (left: sagittal, right: axial) and **C)** hyperintense on FLAIR-weighted MRI.

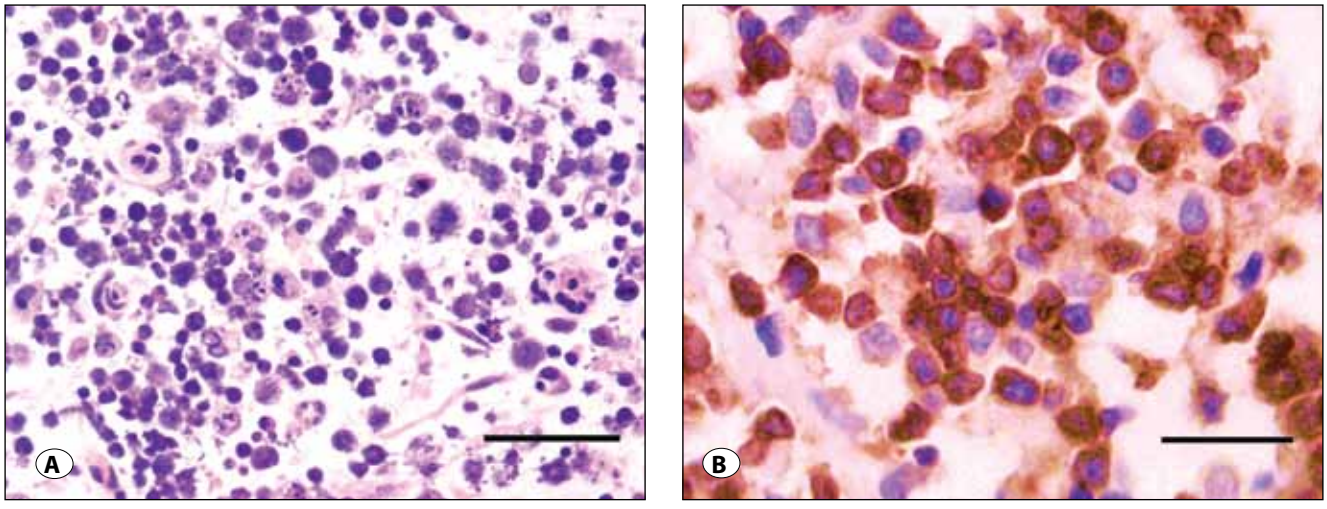

Figure 2: Permanent section of the lesion in the fourth ventricle showed **A)** infiltration of lymphoma cells **B)** with a B cell marker. (**A:** hematoxylin-eosin, bar=100µm; **B:** CD79a, bar=50µm).

B-cell lymphomas should always be included in the list of differential diagnosis of intra-ventricular tumors whether single or multifocal. When PCNSL is suspected, biopsy and chemotherapy/radiotherapy is mostly suggested in the diagnosis and treatment process.

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