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SNI: Neuro-oncology

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Case Report

Immunocompetent patient with isolated primary fourth ventricle lymphoma. Unusual diagnosis, their pitfalls, and challenges

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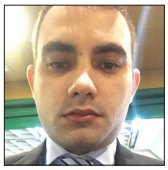

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Received : 29 June 2022
Accepted : 24 August 2022
Published : 14 October 2022

DOI
10.25259/SNI_584_2022

Quick Response Code:

ABSTRACT

Background: Primary central nervous system lymphoma (PCNSL) is an uncommon lesion and represent 4% of all central nervous system (CNS) cancers. There have been few reports of localized isolated lymphoma developing in the fourth ventricle, with only 8 previous cases described. We present a case of an immunocompetent patient with isolated fourth ventricle lymphoma who did not have diffusion-weighted imaging (DWI) restriction.

Case Description: A 45-year-old man presented a history of headache, vomiting and weigh loss. Upon clinical examination, he presented bilateral papilledema, multidirectional nystagmus, and gait imbalance. Magnetic resonance imaging showed a solid tumor in fourth ventricle with 1.8 × 1.6 × 1.1 cm. The patient was submitted to a suboccipital telovelar approach. The pathological study showed a neoplasm composed of loose round cells. Immunohistochemistry showed positivity for CD-45 and CD-23. The diagnosis of primary CNS lymphoma of the fourth ventricle was certified. Patient was sent to complementary treatment with hematologist and radiotherapy and chemotherapy were started.

Conclusion: PCNSL is a rare and aggressive pathology with high rates of mortality and recurrence. It requires a multidisciplinary team and multiple therapies to control the disease and deliver better quality of life and prognosis to the patient.

Keywords: Cancer, Central nervous system, Fourth ventricle, Lymphoma

INTRODUCTION

Primary central nervous system lymphoma (PCNSL) is an uncommon lesion. They represent 4% of all CNS cancers. The PCNSL is an aggressive form of extranodal non-Hodgkin's lymphoma, which represent 6% of the total.[4,6,12] PCNSL can occur in immunosuppressed patients or in immunocompetent patients, however, it is more common in the first group, being most frequently observed during the fifth and sixth decades of life.[4,6]

The anatomical involvement is in deep brain structures such as corpus callosum and basal ganglia. The supratentorial location represents 87% of all cases. In posterior fossa, the brainstem and cerebellum are

the most common sites of presentation.[6] There have been few reports of localized isolated lymphoma developing in the fourth ventricle, with only eight previous cases described.[1,3,7-11,13]

CASE REPORT

A previously healthy 45-year-old man was admitted to the neurosurgical department with a history of headache, vomiting, and weight loss. The symptoms had started 2 months before admission, and the patient sought medical assistance in others two hospitals. On clinical examination, he presented bilateral papilledema, multidirectional nystagmus, and gait imbalance. Complementary examinations were negative for HIV and the defense cells count was normal.

During the radiological investigation, magnetic resonance imaging (MRI) showed a solid tumor in the fourth ventricle with 1.8 × 1.6 × 1.1 cm of size and with the hyper signal on T2 and high contrast uptake on T1, however, with no diffusion-weighted imaging (DWI) restriction [Figure 1]. Among the possible diagnoses, choroid plexus tumors, metastatic tumors, or hemangioblastoma were discussed.

Because of the intracranial hypertension syndrome and the presence of an expansive lesion of the fourth ventricle, the patient was submitted to a suboccipital telovelar approach. During the surgery, a solid gray lesion adherent to the cerebellar vermis and infiltrated the floor of the fourth ventricle was seen, with higher adherence to the roof of the ventricle, however, the tumor appeared to be low vascularized and was pale in color. After debulking, an extracapsular resection was realized. After the surgery, the patient was submitted to a new MRI which showed complete tumor resection [Figure 2].

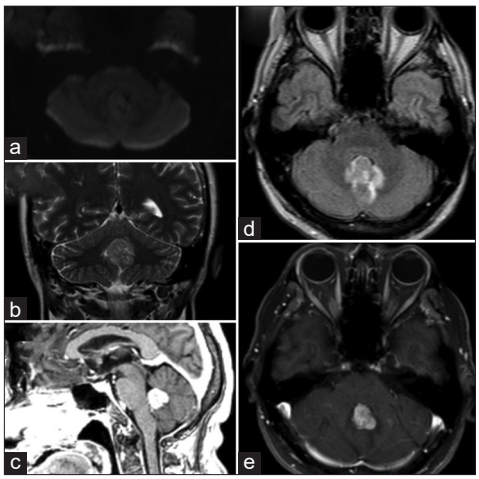

Figure 1: (a) Axial DWI shows no restriction of lesion. (b) Coronal T2 show high signal. (c) Sagittal T1 show high contrast uptake. (d) (Axial Flair) show high signal and (e) (Axial T1) show high contrast uptake

The pathological study showed a neoplasm composed of loose round cells. Immunohistochemistry showed positivity for CD-45, CD-23, and Ki67 in 90% of the cells which concluded the diagnosis of primary CNS lymphoma of the fourth ventricle [Figure 3].

With the lymphoma diagnosis, the patient was sent to complementary treatment with a hematologist, and radiotherapy and chemotherapy were started. Since then, the patient keeps in ambulatorial assistance.

DISCUSSION

PCNSL is an aggressive form of non-Hodgkin lymphoma which is localized in the CNS. They are uncommon lesions that represent approximately 6% of all Lymphomas.[2,4,6,12] In general, these lesions are localized in deep brain structures such as corpus callosum and basal ganglia.[4,6] The presence

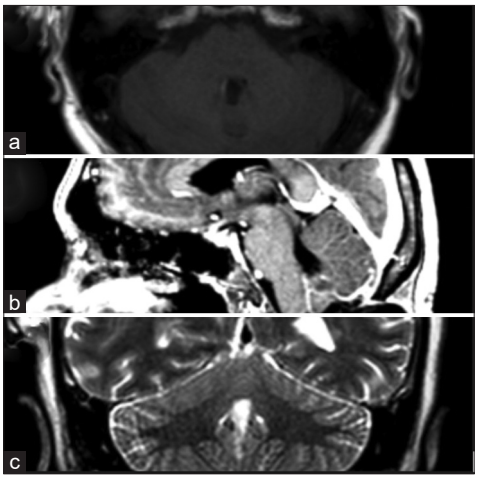

Figure 2: (a) (Axial T1), (b) (Sagittal T1), and (c) (Coronal T2) show the complete lesion resection.

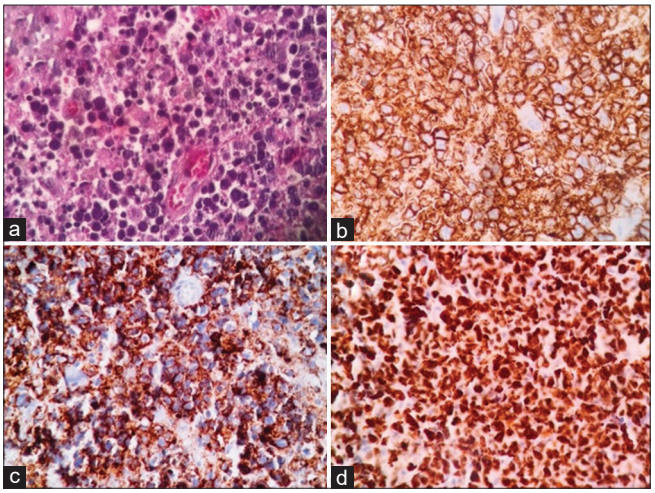

Figure 3: (a) (HE analysis) – shows big lymphocytic cells with hyperchromatic nuclei. (b-d) Show CD20, CD45, and Ki67 markers.

Table 1: Reported cases of primary fourth ventricle lymphoma.

Case number	Author, Year	Gender, Age	Treatment	Outcome
1	Werneck *et al*.[13]	Female, 17	Intrathecal Methotrexate	48 h
2	Haegelen *et al*.[8]	Female, 33	GTR. Chemotherapy. Radiotherapy	7 months
3	Hill *et al*.[9]	Male, 69	Biopsy. Chemotherapy	3 months
4	Liao *et al*.[11]	Male, 77	GTR	9 months
5	Bokhari *et al*.[3]	Male, 50	GTR. Methotrexate. Radiotherapy	18 months
6	Fabiano *et al*.[7]	Female, 60	GTR. De Angelis Protocol	6 months
7	Alabbdulsalam *et al*.[1]	Male, 18	GTR. Chemotherapy. Radiotherapy	18 months
8	Hsu *et al*.[10]	Male, 61	GTR. Chemotherapy	3 months
9	Authors Case	Male, 45	GTR. Chemotherapy and Radiotherapy in progress	4 months

of isolated fourth ventricle lesion is rare, with only eight cases previously reported [Table 1].[1,3,7-11,13] The first report was dated 1977 by Werneck *et al*.[13]

Immunosuppression is a risk factor for the growth of this pathology and however is more prevalent in descriptions of cases in immunocompetent patients. The onset in adulthood is more prevalent, nevertheless, the mean age of presentation is 65 years[6,12]

Clinical presentation is variable.[2] Several signals and symptoms can occur depending on the anatomical location of the tumor difficulty the diagnosis and the suspicion. The most common presentations are unspecific as focal neurologic deficits, behavior changes, and high intracranial syndrome symptoms.[4]

The radiologic criteria of evaluation can help with suspicion. If possible, MRI is mandatory because of some specific changes observed as bellow described. In deep brain structures or on multifocal presentation, the presence of contrast uptake and DWI restriction increase the possibility of diagnosis.[6,12] The real reason is not understood, but some PCNSLs do not present this characteristic radiological alteration; however, the possible explanation is a late necrotic/ischemic tumor, given the absence of hemorrhage and the little vascularization of the lesion observed during the operation.

Despite the radiologic information, pathologic confirmation is necessary and can be made for many possible approaches; however, the stereotaxic biopsy and navigation-guided needle biopsy are the most cited because of the minimally invasive technique.[5,12] Here, an important consideration is the use of corticosteroids. These medications can camouflage the pathological results and late the diagnosis because they can falsify the pathological result. In an ideal situation, the patient must be free of corticosteroids 2 weeks before biopsy.[5]

The lymphoma subtype diagnosed is in 90% of cases B-cell Lymphomas. Less commonly can be present in Burkitt or T-Cell forms. In general, the histologic aspect includes centroblastic cytology and perivascular tropism. CD markers offer a specific diagnosis. CD20, CD19, and CD79 for B-Cell type are the most commonly finds.[2,5,6]

The management is challenging. A multidisciplinary team is necessary, and the stratification depends on age, comorbidities, performance status, and neurologic conditions. Chemotherapy and radiotherapy can be used isolated or combined.[2]

Surgical approaches are discouraged. The previous analysis showed no benefit in outcome when used alone or combined with adjuvant therapy. Another explanation is that here the disease is multifocal and has infiltrative nature, besides can induce permanent neurological deficits. From this perspective, the surgical approaches are not adequate for incomplete resection.[4,12]

The prognosis depends on many variables. Age and performance status are independent prognostic factors. Some authors also describe Ki-67, MUM1, and bcl-6 markers as possible prognosis definers[6] After the application of chemotherapy and radiotherapy in the defined list of treatments for these patients, there was an improvement in survival time; however, there is still a considerable rate of lesion recurrence and mortality that encourage new research and treatments.[2,4,6,12]

Within recent gains in lymphoma therapy, the targeted treatment with immunomodulators has to be noted as an important weapon, mainly reducing tumor recurrence rates.[5] The German High-Grade Non-Hodgkin Lymphoma Study Group Trials showed that the use of rituximab decreased CNS recurrence. In the Korean study, only one case of CNS recurrence was observed in 80 cases on total.[2]

CONCLUSION

Despite expansive lesions of fourth ventricle being considered more commonly ependymomas, medulloblastomas, and metastasis, it is possible the diagnosis of lymphoma. In this case, no DWI restriction was observed, which led to microsurgical resection as the first-choice therapy due to the low possibility of lymphoma.

PCNSL is a rare and aggressive pathology with high rates of mortality and recurrence. It is necessary for a multidisciplinary team and multiple therapies to control the disease and deliver better quality of life and prognosis to the patient.

Surgery is necessary to confirm the diagnosis; however, minimally invasive such as stereotaxic biopsy and navigation-guided needle biopsy is important tools for accessing tumor tissue for diagnostic confirmation. Chemotherapy and radiotherapy keep as the principal way of treatment, nevertheless, recent studies showed the importance of immunotherapy in overall survival and recurrence of pathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest

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How to cite this article: Holanda TS, Pimentel IM, Gosch GO, Tavora DG, Bandeira LA, Filho FL. Immunocompetent patient with isolated primary fourth ventricle lymphoma. Unusual diagnosis, their pitfalls, and challenges. Surg Neurol Int 2022;13:463.