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(CASE REPORT)

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Lymphoma of the cerebellopontine angle mimicking metastasis lesion: A case report

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Abstract

Primary lymphoma of the cerebellopontine angle (CPA) is rare and uncommon in the central nervous system, accounting for approximately 0.2-2% of all brain tumors. To our knowledge, there have only been 32 cases reported worldwide so far. Here, we report a rare case of B-cell lymphoma in a 56-year-old woman who presented with seizure, dysphagia, and dizziness and showed a lesion involving the left CPA, right frontal horn and septum pellucidum on magnetic resonance imaging (MRI). The primary diagnosis was metastatic tumor; however, Stereotactic biopsy on left CPA lesion was done, and the histological features confirmed the diagnosis of B-cell malignant lymphoma. The patient was treated with chemotherapy, with complete resolution of symptoms after the first chemotherapy session.

Keywords: Malignant lymphoma; Cerebellopontine angle; Metastatic tumor; stereotactic biopsy.

1. Introduction

Primary central nervous system lymphomas (PCNSLs) are uncommon and very rare, accounting for approximately 0.2%–2 percentage of all brain tumors [1]. They usually present in supratentorial locations with a predilection for deep white mater such as corpus callosum, basal ganglia, thalami, and paraventricular region [2]. Cerebellopontine angle (CPA) location of PCNSLs is rare and can be mistaken for other diseases such as vestibular schwannoma or meningioma, which constitute vast majority of mass lesions in the CPA. To our knowledge, only 32 cases have been reported so far in the literature [3, 4]. We report a case of CPA lymphoma with other associated localization, diagnosed by stereotactic biopsy, and we discuss the presentation, radiologic characteristics, preoperative differential diagnosis and treatment.

2. Case report

A 56-year-old women patient with one-year history of headache presented gradually dizziness and ataxia. Two months before her admission she presented multiple episodes of seizures, vomiting and swallowing dysfunction.

A physical examination disclosed that the patient's consciousness was intact with partial palsy of the left facial nerve (House-Brackman grade III) and left cerebellar syndrome. All routine investigations were within normal limits. Computed Tomography (CT) and Magnetic resonance imaging (MRI) brain showed left CPA mass lesion extending to fourth ventricle, associated with small lesion in right frontal horn and septum pellucidum (Figure 1, 2).

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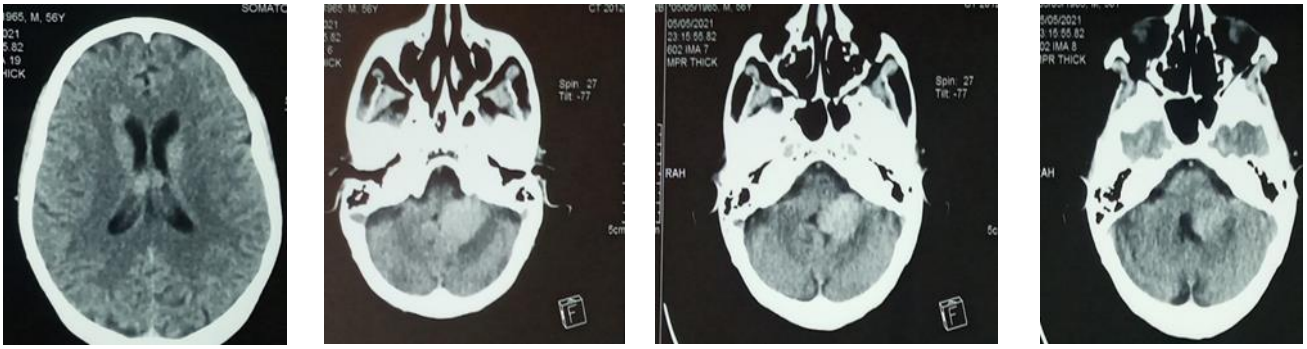

Figure 1 CT scan show left CPA mass lesion extending to fourth ventricle, associated with small lesion in right frontal horn and septum pellucidum

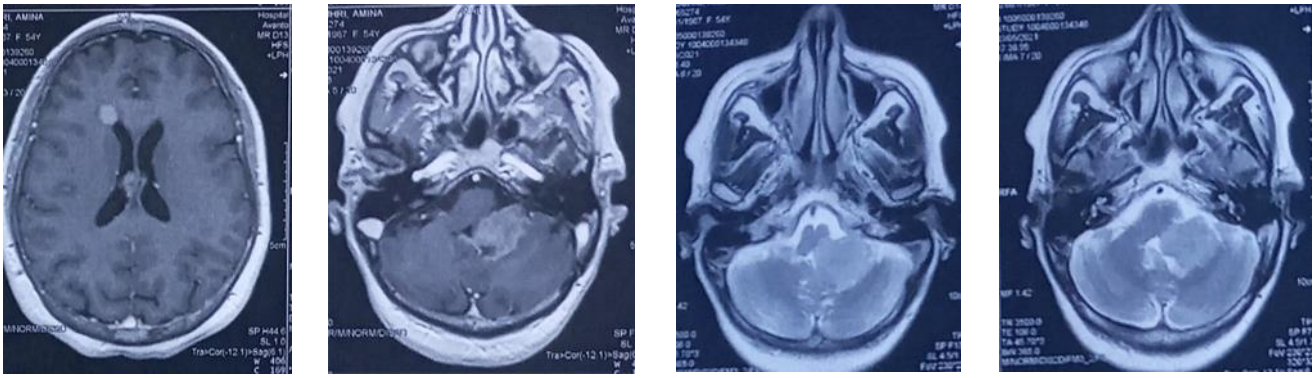

Figure 2 MRI show a left CPA mass lesion extending to fourth ventricle, associated with small lesion in right frontal horn and septum pellucidum

Radiological impression was metastatic lesions and the patient was taken for stereotactic biopsy [figure 3], the biopsied sample consisted of gray-white soft tissue and histopathological examination revealed fragmented bits of glial tissue harboring a cellular lesion arranged in diffuse sheets, comprising large cells having vesicular nuclei and moderate cytoplasm. Dilated blood vessels were seen. On immunohistochemistry, tumor cells were positive for CD20 (L26 clone) and BCL6, BCL12, MUM1 and Ki67 was 90%. A final diagnosis of diffuse large B-cell lymphoma (DLBCL) without germinal center type was rendered.

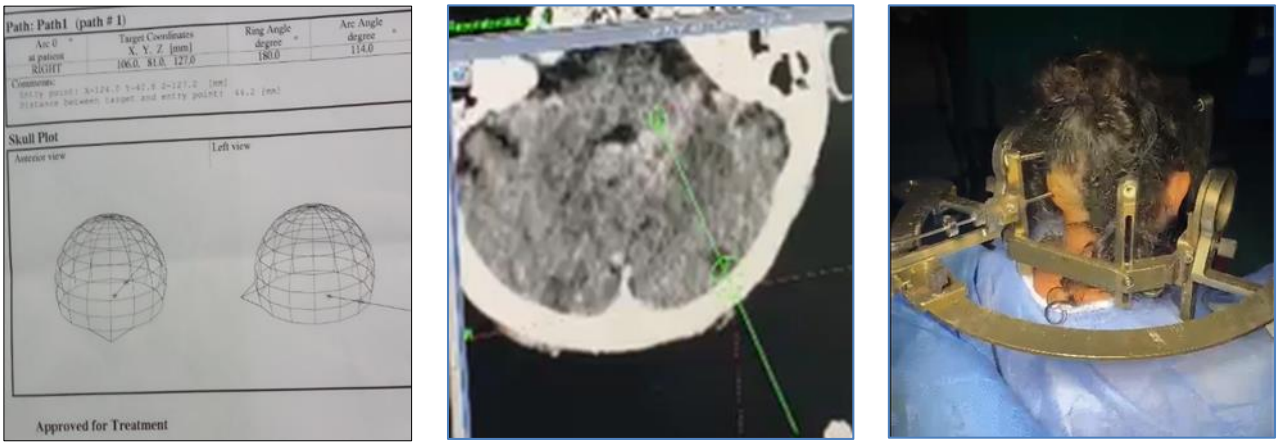

Figure 3 Stereotaxic identification using CT scan

After having been informed of the pathological diagnosis, the patient underwent chemotherapy with four cycles of cyclophosphamide, doxorubicin, vincristine, and prednisolone in oncological department. The evolution was marked by improvement of swallow dysfunction and gait disturbances.

3. Discussion

Lesions involving the intracranial Cerebellopontine Angle (CPA) are mostly benign. The major differential diagnoses of a CPA mass mainly include acoustic schwannoma (80%-90%), meningioma (5%-10%), and epidermoid inclusion cysts (5%-7%). Lymphoma of central nervous system is a relatively rare, highly malignant tumor that accounts for only 0.2-2% of intracranial tumors [1, 2]. It can be classified as primary and secondary depending on the involvement of extracranial sites. Primary because no lymphoma outside the central nervous system is detected [13].

Primary central nervous system lymphoma (PCNSL) presents as a single lesion in 60% to 70% of patients, and diffuse large B-cell lymphoma accounts for the majority (90%) of cases [5], most commonly located in the hemisphere (38%), thalamic/basal ganglia (16%), corpus callosum (14%), periventricular area (12%), or cerebellum (9%) [6]. Cerebellopontine angle lymphoma is very rare; however, it must be kept in differential diagnosis, as accurate diagnosis is imperative for proper management of the patient. Only 32 cases have been reported so far in the literature.

It has been reported to present between the ages of 16 days and 90 years, but this condition is predominantly one of the fifth and sixth decades, with a male to female ratio of 1.5–2.7 [8, 9].

Regarding the locations of cerebellopontine angle lymphoma, 22 cases (68.75%) were in the left side, 14 cases (43.75%) were in right side, and two cases (6.3%) were bilateral [10]. And clinical manifestations are related to the anatomical structures surrounding the lesion and volumes of the tumors. PCNSLs of the CPA typically present with trigeminal, facial, audio vestibular and cerebellar dysfunction, including hearing loss, tinnitus, facial palsy, vertigo and ataxia [7, 11, 18]. Other neurological symptoms include headache, nausea, vomiting, hoarseness and nystagmus [18, 15, 16].

The CT characteristics of Central Nervous System lymphomas have been reported to be indistinguishable from acoustic Neurinomas and Meningiomas [3]. However, in contradistinction to most acoustic Neurinomas, there is less bony erosion of the internal auditory canal, especially in primary CPA lymphoma. On MRI, they are typically homogeneously isointense on T1WI [7, 1], and homogeneously isointense/hypo intense with mild perifocal edema on T2WI [4, 12]. After contrast administration, they show an intense homogeneous enhancement [16]. In our case, the MRI appearance of the CPA lymphoma appeared isointense on T1WI, isointense on T2WI. After contrast administration, the lesion presented an intense homogeneous enhancement.

When CT or MRI suspects Primary lymphoma of Cerebellopontine angle, conclusive diagnosis should be made by histological or cytological examination of tumor [14, 17]. In our case, the suspect diagnosis were metastasis because of presence of two other lesions in right frontal horn and septum pellucidum, and in the literature only one case of 32 reported the preoperative diagnosis was metastasis [19]

CSF sampling by lumbar puncture can be performed at the time of initial assessment [12].If CSF cytology is successful to establish a definitive diagnosis of malignant lymphoma, surgery can be avoided [14, 20].But if it failed to confirm the diagnosis, tumor excision or biopsy should be performed.

Since radical tumor resection has no advantage on survival and may cause more neurologic deficits, radical decompression should be discouraged [12, 14, 15]. It is advisable to proceed with stereotactic biopsy for brain lesions with a radiographic appearance consistent with Primary lymphoma [21]. Through review of the literature, tumor resection was performed in 80.6% of the patients, and stereotactic biopsy was achieved in only 13.8% of the patients.

Once the diagnosis of primary lymphoma of Cerebellopontine angle is established, more effective treatment should be delivered. However, the optimal treatment of PCNSL has yet to be defined [21]. High-dose methotrexate (HD-MTX)–based induction chemotherapy is considered standard for newly diagnosed primary lymphoma [21].

4. Conclusion

Although malignant lymphomas rarely occupy the CP angle, it should be considered in the differential diagnosis of CP angle tumors. It is desirable to obtain a frozen section in all CP angle tumors during surgery to identify this tumor with preference to stereotactic biopsy, because an aggressive total removal is not necessary but radiation therapy should additionally be performed in this tumor.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

The authors declare that they have no conflicts of interest.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors'.

Statement of informed consent

The patient gave his informed consent to publish his case.

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