

Primary Central Nervous System Lymphoma in the Fourth Ventricle

XinYi^{1,2#}, Shuwei Qiu^{1#}, Xiaoming Rong¹, Mohammad Imran Ahmed Ibrahim¹, Qingyu Shen^{1*} and Yuefei Deng^{3*}

¹Department of Neurology, Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University, PR China

²Department of Neurology, the first affiliated Hospital of Jishou University, Jishou, PR China

³Department of Neurosurgery, Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University, PR China

#These authors contributed equally to this work

*Corresponding authors: Yuefei Deng, Department of Neurosurgery, Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University, No. 107 West Yanjiang Road, Guangzhou 510120, PR China, Tel: +8613503000739; Fax: 86-20-81332833; -E-mail: flyneurosurgery@163.com

Qingyu Shen, Department of Neurology, Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University, PR China, Tel: +8613609710406, E-mail: Qingyushen123@126.com

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Abstract

Primary central nervous system lymphoma (PCNSL), a highly malignant and infrequent tumor, is rarely found within the fourth ventricle. Here we report a case of isolated lymphoma within the fourth ventricle: a 61-year-old male presented with headache, mental disorder, abnormal gait and urinary incontinence. Based on the clinical symptoms and neuroimage, He was initially diagnosed as choroid plexus papilloma. However, post-surgery pathology corrected the diagnosis as a diffuse large B-cell lymphoma. To the best of our knowledge, only nine cases are documented in the literature and all of them were initially diagnosed as other tumours or not determined. Therefore, Thisis unconventional location and atypical clinical manifestation of PCNSL in the fourth ventricle should be included on the list of differential diagnosis of the fourth ventricle tumors.

Case Report

A 61-year-old man presented to our hospital complaining of headache for one month and somnolence, abnormal gait and urinary incontinence for one week. During admission, he was in a state of somnolence and physical examination only revealed the mild cervical resistance. His past medical history and laboratory tests including routine blood parameters, blood biochemistry (including lactate dehydrogenase (LDH) HIV testing), routine coagulation parameters and erythrocyte sedimentation rate were normal as well.

Brain magnetic resonance imaging (MRI) revealed one irregular nodule occupying the fourth ventricle with a hypo intensity on T1 weighted image (T1WI) and is intensity on T2 weighted image (T2WI) (Figure 1). Post-contrast T1WI showed well-defined and marked homogenous enhancement without necrosis. Furthermore, contrast enhancement image demonstrated that the primary nidus of the mass originated from the roof of the fourth ventricle, which was surrounded with choroid plexus (Figure 2). The intraventricular location of the mass and MRI manifestation led us to clinical diagnosis as choroid plexus papilloma (CPP). PET-CT of the whole body showed high metabolism in the fourth ventricle. The special location of the mass prompted our surgical team to choose surgical excision over stereotactic biopsy.

Interestingly, post-surgery pathology diagnosed as a diffuse large B-cell lymphoma (Figure 3). The patient refused to undergo radiotherapy or chemotherapy immediately after the surgery.

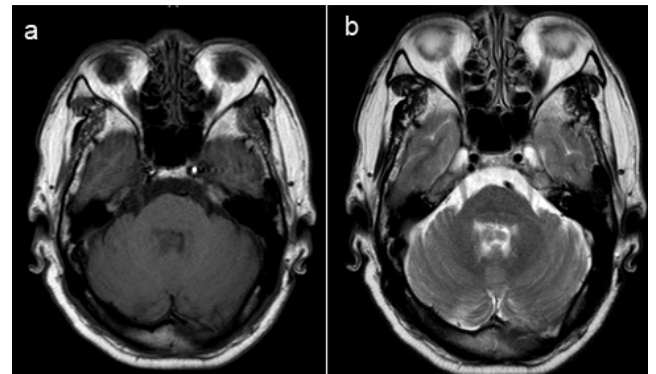


Figure 1: Axial brain MR T1WI demonstrating a hypointense mass lesion (a), T2WI isointensity (b), occupying within the fourth ventricle.

At 6-week follow-up after the resection, brain MRI demonstrated enhanced nodules in the third ventricle and midbrain aqueduct area, which were considered as implantation metastases. Consequently, the patient was administrated with chemotherapy combined with radiotherapy. Another month later, these lesions amazingly disappeared on brain MRI. Currently, 20 months post-surgery, the patient is still under active follow-up.

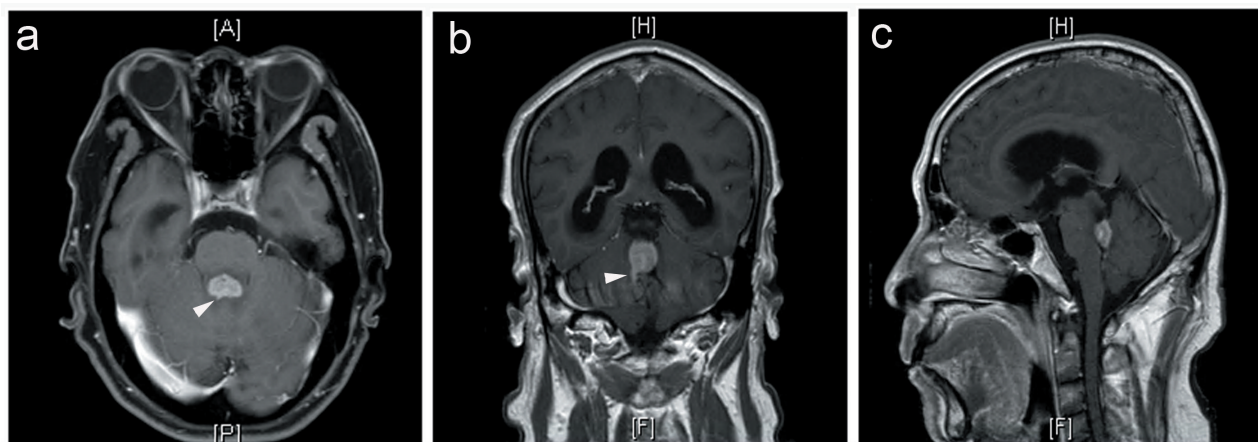


Figure 2: MR T1WI contrast enhancement demonstrating that the mass lesion is well-defined and marked homogenous enhancement with the lesion nidus originating from the roof of the fourth ventricle (arrow). a: axial; b: coronal; c: sagittal.

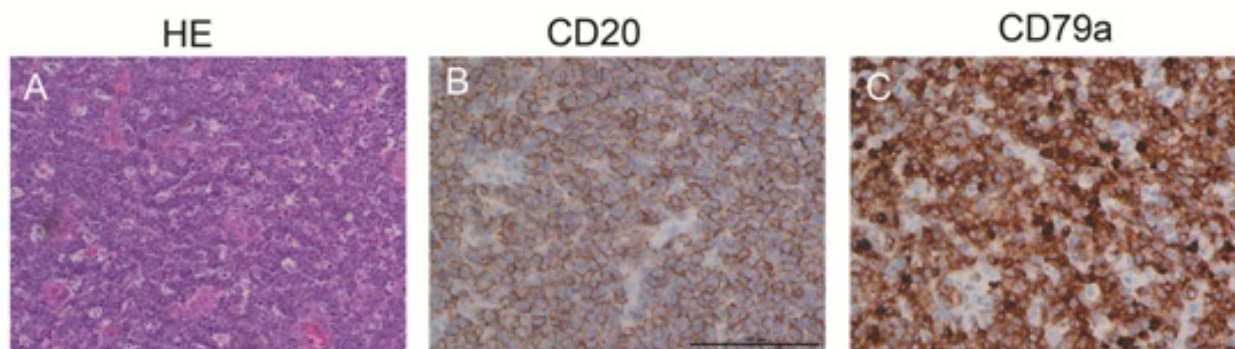


Figure 3: Pathological assays: The tumor was composed of large neoplastic lymphoid, and aggregations of mature lymphocytes also seen (Hematoxylin and Eosin stain) (A) and immunohistochemical stain were positive for CD20B-cell marker) and CD79a (B and C).

Discussion

PCNSLs are uncommon among intracranial tumours and their familiar locations are cerebral hemispheres, basal ganglia and the corpus callosum. The tissue of fourth ventricle tumour is mainly ependymal and choroidal. The List of the fourth neoplastic lesions includes ependymoma, choroid plexus papilloma, sub ependymoma, low-grade astrocytoma, and medulloblastoma. PCNSLs within fourth ventricle are relatively seldom seen in immune-competent patients. To the best of our knowledge, only nine cases of PCNSL located at fourth ventricle have been reported in the literature (Table 1) [1-9].

Given the sensitivity of PCNSLs to chemotherapy and radiotherapy, surgical resection is not recommended for PCNSLs, the early recognition of PCNSL is heavily warranted. Usually, PCNSLs on MRI appears with hypo intense/isointense on T1WI and isointense on

T2WI. Early identification of PCNSLs via neuroimaging is necessary but is not specific in signal characteristics. In our case, the initial diagnosis was CPP, which is usually located in the lateral and fourth ventricles. However, CPP usually appears isointense on CT and isointense to the white matter on MR T1WI with lobulated contour and occasionally calcifications and intralesional flow voids could also be observed. These characteristics may facilitate the differential diagnosis of PCNSL. According to Summary of the nine reported cases and our case of primary isolated lymphoma within the 4th ventricle a consideration of PCNSL should be warranted if contrast enhancement image shows homogeneous enhancement without necrosis. Besides, Magnetic Resonance Spectrum (MRS), in combination with hyper intensity on CT [10] is helpful for differential diagnosis between PCNSL and other brain tumors. By the way, due to the special location, excision was the preferred choice for PCNSL in the fourth ventricle.

Authors, year	Age/sex	Clinical presentation	Initial diagnosis	MRI	Surgical treatment	Diagnosis	Extend treatment	Follow-up
Werneck et al. [1] 1977	17/F	Meningitis	Meningitis	N/A	N/A	Primary CNS lymphoma (necroscopy)	N/A	N/A
Haegelen et al. [2] 2001	33/F	Headaches, Vertigo and static cerebellar syndrome	N/A	A homogenous fourth ventricular lesion without hydrocephalus.	Excision	High-grade B-cell lymphoma	Chemotherapy, autologous stem cell transplantation and WBRT	7 months without recurrence
Hill et al. [3] 2009	69/M	Vomiting, mild nausea, anorexia and weight loss.	N/A	A homogeneously enhancing mass in the caudal portion of the fourth ventricle without hydrocephalus.	Excision	High-grade B-cell lymphoma	Chemotherapy	3 months without recurrence
Bokhari et al. [4] 2013	50/M	Vomiting, moderate nausea, headache and disorders of consciousness	Medulloblastoma or ependymoma	A strongly enhancing mass occupying the inferior of the fourth ventricle with hydrocephalus.	Excision	High-grade B-cell lymphoma	Chemotherapy and WBRT	18 months without recurrence
Rao et al. [5] 2013	59/M	Vomiting, vertigo, tremors of both upper limbs and head and unsteady gait	Hemangioblastoma or low grade glioma	Isointense on T1-weighted, hypointense on T2-weighted sequences with contrast enhancement	Excision	Diffuse large B-cell lymphoma	Chemotherapy	8 months without recurrence
Liao CH et al. [6] 2013	77/M	Intermittent vertigo, nausea, vomiting, unsteady gait	Metastasis or high-grade glioma	A homogeneously enhancing tumor in the fourth ventricle with hydrocephalus	Excision	Diffuse large B-cell lymphoma	No further treatment	9 months without recurrence
Fabiano et al. [7] 2014	60/F	Diplopia	N/A	A homogeneously enhancing mass within the fourth ventricle without hydrocephalus	Excision	Diffuse large B-cell lymphoma	Chemotherapy and WBRT	6 months without recurrence
Gossman et al. [8] 2014	66/M	Progressive gait disturbances and diplopia	N/A	A homogeneously enhancing tumor in the fourth ventricle.	Excision	Large B-cell lymphoma	Further oncological treatment	N/A
Huang-I Hsu et al. [9] 2015	61/M	Headache, dizziness, progressively unsteady gait	Lymphoma or ependymoma	A homogeneously enhancing tumor in the fourth ventricle	Excision	Diffuse large B-cell lymphoma	Chemotherapy and radiotherapy	3 months without recurrence
This case	61/M	Headaches, disorders of consciousness, abnormal gait and urinary incontinence	Choroid plexus papilloma	A homogeneously enhancing tumor in the fourth ventricle with hydrocephalus	Excision	Diffuse large B-cell lymphoma	Chemotherapy and radiotherapy	20 months without recurrence and following up

N/A: Not Available; WBRT: Whole Brain Radiotherapy.

Table 1: Summary of the nine cases and our case of primary isolated lymphoma within the 4th ventricle.

Conclusion

We conclude that primary central nervous system lymphoma should be included on the list of differential diagnosis of the fourth ventricle tumour.

Acknowledgement

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References

1. Werneck LC, Hatschbach Z, Mora AH, Novak EM (1977) Meningitis caused by primary lymphoma of the central nervous system. Report of a case. *Arquivos de Neuro-Psiquiatria* 35: 366-372.
2. Haegelen C, Riffaud L, Bernard M, Morandi X (2001) Primary isolated lymphoma of the fourth ventricle: Case report. *Journal of neuro-oncology* 51: 129-131.
3. Hill CS, Khan AF, Bloom S, Mc Cartney S, Choi D (2009) A rare case of vomiting: Fourth ventricular b-cell lymphoma. *Journal of neuro-oncology* 93: 261-262.
4. Bokhari R, Ghanem A, Alahwal M, Baeesa S (2013) Primary isolated lymphoma of the fourth ventricle in an immunocompetent patient. *Case Reports in Oncological Medicine* 2013: 614658.
5. Rao RN, Mishra D, Agrawal P, Kumar R (2013) Primary b-cell central nervous system lymphoma involving fourth ventricle: A rare case report with review of literature. *Neurology India* 61: 450-453.
6. Liao CH, Lin SC, Hung SC, Hsu SP, Ho DM, et al. (2014) Primary large b-cell lymphoma of the fourth ventricle. *Journal of clinical neuroscience* 21: 180-183.
7. Fabiano AJ, Syriac S, Fenstermaker RA, Qiu J (2014) Primary fourth ventricular b-cell lymphoma in an immunocompetent patient. *Clinical neuropathology* 33: 94-97.
8. Grossman R, Nossek E, Shimony N, Raz M, Ram Z (2014) Intraoperative 5-aminolevulinic acid-induced fluorescence in primary central nervous system lymphoma. *Journal of Neurosurgery* 120: 67-69.
9. Hsu HI, Lai PH, Tseng HH, Hsu SS (2015) Primary solitary lymphoma of the fourth ventricle. *International Journal of Surgery Case Reports* 14: 23-25.
10. Sutherland T, Yap K, Liew E, Tartaglia C, Pang M, et al. (2012) Primary central nervous system lymphoma in immunocompetent patients: a retrospective review of MRI features. *Journal of Medical Imaging and Radiation Oncology* 56: 295-301.