Primary Central Nervous System Lymphoma in the Fourth Ventricle

Xin Yi1,2#, Shuwei Qiu1#, Xiaoming Rong1, Mohammad Imran Ahmed Ibrahim1, Qingyu Shen1* and Yuefei Deng3*

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

Rec date: Jan 06, 2017; Acc date: Feb 02, 2017; Pub date: Feb 04, 2017

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, Thesis 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 forth 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 an 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.
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, subependymoma, 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 (neuroscopy) | 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 homogenously 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**

This work was supported by National Natural Science Foundation of China (No. 81471290 to Qingyu Shen and No. 81402065 to Shuwei Qiu) and Natural Science Foundation of Guangdong (No.52013010015652 to Qingyu Shen).
References


