A cerebellar neurocytoma with an excellent response to radiotherapy: a case report

Siwei Chen

Peking University First Hospital

Hongzhou Duan

Peking University First Hospital

Ran Liu (✉ dyyxys@126.com)

Peking University First Hospital <https://orcid.org/0000-0003-0819-4543>

Jingjing Luo

Peking University First Hospital

Hui Wang

Peking University First Hospital

Shuang Zhang

Peking University First Hospital

Haiqiang Jin

Peking University First Hospital

Jing Bai

Peking University First Hospital

Feng Gao

Peking University First Hospital

Guiping Zhao

Peking University First Hospital

Yining Huang

Peking University First Hospital

Case report

Keywords: Extraventricular neurocytoma (EVN), Cerebellum, Radiotherapy

Posted Date: February 26th, 2020

DOI: <https://doi.org/10.21203/rs.2.24594/v1>

License: © ⓘ This work is licensed under a Creative Commons Attribution 4.0 International License.

[Read Full License](#)

Abstract

Background: Extraventricular neurocytoma (EVN) is a rare central neurocytoma with neoplasms occurring in the brain parenchyma outside the ventricular system with similar biological behaviours and histopathological characteristics. Cases of EVN reported in the brainstem and cerebellum are fairly rare. In addition, very few cases with radiotherapy as the only treatment have been reported, and their outcomes were unclear.

Case presentation: We reported an EVN of the brainstem and cerebellum confirmed by pathology in a 43-year-old male presenting with unprovoked nausea and dizziness. The patient showed a favourable outcome with only radiotherapy during the 2-year follow-up.

Conclusions: Patients who have EVN may have a favourable prognosis with radiotherapy without surgery.

Background

Central neurocytomas (CNs) are benign central nervous system tumours that are ranked as grade II according to the 2007 World Health Organization classification (1). This tumour was first described by Hassoun and colleagues in 1982 (2) and was characterized by its neuronal origin and its location within the lateral ventricles (3). In 1989, Ferroel reported tumours in the parenchyma outside the ventricular system with a similar histopathologic morphology and named them extraventricular neurocytomas (EVNs) (4). In adults, EVNs are most likely to occur within the frontal lobe, followed subsequently by the temporal, parietal and occipital lobes, but rarely occur in the brainstem and cerebellum (5). We present a patient with an EVN in the posterior fossa who showed a good response to radiotherapy.

Case Presentation

A 43-year-old male presented with unprovoked nausea for 2 months and dizziness for 1 month. He developed blurred vision and diplopia 2 weeks later. Physical examination demonstrated horizontal and downbeat nystagmus, as well as diplopia, when he looked downwards. Cranial magnetic resonance imaging (MRI) revealed an ill-defined lesion in the dorsal part of the pons and the cerebellum around the fourth ventricle (Fig. 1) with a homogeneous low signal intensity on T1-weighted images, a high signal intensity on T2-weighted images, and mild homogenous Gd enhancement (Fig. 1). Lumbar puncture showed no obvious abnormality except that the protein level in the cerebrospinal fluid (CSF) was 0.48 g/L. AQP-4 antibody was negative both in the serum and CSF. Flow cytometry showed no abnormal subtypes of lymphocytes in CSF. The maximum SUV of the lesion was 10.5–12.3 on positron emission tomography-computed tomography (PET-CT). The patient denied biopsy and received glucocorticoids for 1 month. Follow-up MRI revealed a well-circumscribed $2.0 \times 1.3 \times 2.1$ cm mass in the vermis with evident enhancement 2 months later with marked perilesional oedema (Fig. 2). Stereotactic core needle biopsy was performed to make a diagnosis. Monotonous clear round cell proliferation without invasive characteristics was observed, clearly separated from cerebellar tissue. Immunohistochemical staining

showed strong positivity for synaptophysin (Syn), as well as neuron-specific enolase (NSE), NeuN and D2-40. Glial fibrillary acidic protein (GFAP) was focally positive (+/-), while EGFR, CD34, inhibin, S100, Oligo2, IDH1 and p53 were all negative. The Ki67 labelling index was < 1% (Fig. 3). The histopathological results supported the diagnosis of EVN. Three weeks later, without any intervention, repeated cranial MRI showed enlargement of the tumour to 3.0 × 2.4 × 2.4 cm with a cystic component. Despite the rapid progression of the tumour, the patient accepted only radiotherapy (2 Gy*25, total 50 Gy) and rejected surgery. One month after radiotherapy, the patient's symptoms were relieved. At the 8-month follow-up, MRI showed that the tumour size was obviously reduced (Fig. 4). At the 12-month and 24-month follow-ups, the cranial MRI results were similar to those obtained 8 months after radiotherapy.

Discussion

At present, there are almost 201 cases of EVN reported, mostly from Asian populations (6). Among these cases, 28% were in the frontal lobe, 18% were in the temporal lobe, and only 13 cases of EVNs were located in the cerebellum (7). At symptom onset, the lesion was located in the area postrema, which mimicked neuromyelitis optica spectrum disorder (8). However, the AQP-4 antibody was negative in both the cerebral spinal fluid and serum, and the lesion was not responsive to steroid therapy. Two months after the steroid treatment, the lesion was enlarged with marked oedema. The diagnosis of EVN was established based on the pathological results. Additionally, glioma, lymphoma and oligodendroglioma were excluded by the immunohistochemistry results.

Although neurocytomas are tumours of WHO grade II, indicating that they have better outcomes, some EVNs may have more infiltrative and aggressive progression, leading to a poor prognosis and a high rate of recurrence. These EVNs were given the attribute name "atypical" and are characterized by a Ki-67 index > 2% or MIB-1 > 3% and/or with atypical histological features (9, 10). Neurocytomas with high FDG uptake also showed an increased proliferative index associated with atypical histological features (11). In the current case, the tumour showed benign features with Ki67 < 1% (12). However, the tumour grew rapidly after needle biopsy. Some reports have also indicated that although neurocytomas are considered indolent, they may have aggressive biologic behaviour, including postoperative rapid regrowth (13, 14). Increased glucose metabolism may also indicate high proliferative activity of the brain tumour (15).

Gross total resection (GTR) is the preferred strategy for EVNs, with the best outcome and a low recurrence rate (5, 9). However, because some EVNs are located close to eloquent areas, surgery often results in subtotal resection (STR). Radiotherapy is recommended for atypical EVNs, among which only 13% have the chance to undergo GTR (16). The effectiveness of GTR and STR combined with adjuvant radiotherapy was reported to be comparable (5, 9, 16). Among the cases with treatment details available, 48.4% underwent gross total resection (GTR), 15.6% underwent subtotal resection (STR), and 28.7% received adjuvant radiation. Only 4 patients received biopsy and radiation as the only therapy. The current case is the only report of a cerebellar EVN that showed a good response to only radiotherapy without STR. However, little is known about the prognosis of EVN under radiotherapy as the only therapy.

Conclusions

We report a case of EVN localizing in the cerebellum. The patient showed a favourable prognosis with radiotherapy without surgery.

Abbreviations

EVN

Extraventricular neurocytoma

CN

Central neurocytoma

MRI

Magnetic resonance imaging

CSF

Cerebrospinal fluid

PET-CT

Positron emission tomography-computed tomography

Syn

Synaptophysin

NSE

Neuron-specific enolase

GFAP

Glial fibrillary acidic protein

GTR

Gross total resection

STR

Subtotal resection

Declarations

Ethics approval and consent to participate

Not applicable

Consent for publication

Written informed consent for publication of the details and clinical images was obtained from the patient.

Availability of data and materials

Not applicable, entire data is shown within the manuscript/figures.

Competing interests

The authors declare that they have no competing interests.

Funding

Not applicable.

Authors' contributions

SC, HD, RL took care of this patient, performed stereotactic core needle biopsy and wrote the manuscript. JL, HJ, JB, FG, GZ and YH analysed this case and directed treatment. HW and SZ carried on the pathology analysis.

Acknowledgements

Not applicable.

References

1. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, et al. The 2007 WHO classification of tumours of the central nervous system. *Acta Neuropathol.* 2007;114(2):97-109.
2. Hassoun J, Gambarelli D, Grisoli F, Pellet W, Salamon G, Pellissier JF, et al. Central neurocytoma. An electron-microscopic study of two cases. *Acta Neuropathol.* 1982;56(2):151-6.
3. Dedushi K, Kabashi S, Ugurel MS, Ramadani N, Mucaj S, Zeqiraj K. Magnetic Resonance Imaging of a Case of Central Neurocytoma. *Acta Inform Med.* 2016;24(6):419-21.
4. Ferreol E, Sawaya R, de Courten-Myers GM. Primary cerebral neuroblastoma (neurocytoma) in adults. *J Neurooncol.* 1989;7(2):121-8.
5. Sweiss FB, Lee M, Sherman JH. Extraventricular neurocytomas. *Neurosurg Clin N Am.* 2015;26(1):99-104.
6. Wang J, Song DL, Deng L, Sun SY, Liu C, Gong DS, et al. Extraventricular neurocytoma of the sellar region: case report and literature review. *Springerplus.* 2016;5(1):987.
7. Mallick S, Benson R, Rath GK. Patterns of care and survival outcomes in patients with an extraventricular neurocytoma: An individual patient data analysis of 201 cases. *Neurol India.* 2018;66(2):362-7.
8. Wingerchuk DM, Banwell B, Bennett JL, Cabre P, Carroll W, Chitnis T, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology.* 2015;85(2):177-89.
9. Patil AS, Menon G, Easwer HV, Nair S. Extraventricular neurocytoma, a comprehensive review. *Acta Neurochir (Wien).* 2014;156(2):349-54.
10. Moriguchi S, Yamashita A, Marutsuka K, Yoneyama T, Nakano S, Wakisaka S, et al. Atypical extraventricular neurocytoma. *Pathol Int.* 2006;56(1):25-9.

11. Ohtani T, Takahashi A, Honda F, Ishiuchi S, Kurihara H, Inoue T, et al. Central neurocytoma with unusually intense FDG uptake: case report. *Ann Nucl Med*. 2001;15(2):161-5.
12. Cho M, Joo JD, Kim BH, Choe G, Kim CY. Hypothalamic Extraventricular Neurocytoma (EVN) in a Pediatric Patient: A Case of EVN Treated with Subtotal Removal Followed by Adjuvant Radiotherapy. *Brain Tumor Res Treat*. 2016;4(1):35-9.
13. Warmuth-Metz M, Klein R, Sorensen N, Solymosi L. Central neurocytoma of the fourth ventricle. Case report. *J Neurosurg*. 1999;91(3):506-9.
14. Wharton SB, Antoun NM, Macfarlane R, Anderson JR. The natural history of a recurrent central neurocytoma-like tumor. *Clin Neuropathol*. 1998;17(3):136-40.
15. Kruer MC, Kaplan AM, Etzl MM, Jr., Carpentieri DF, Dickman PS, Chen K, et al. The value of positron emission tomography and proliferation index in predicting progression in low-grade astrocytomas of childhood. *J Neurooncol*. 2009;95(2):239-45.
16. Xu L, Ouyang Z, Wang J, Liu Z, Fang J, Du J, et al. A clinicopathologic study of extraventricular neurocytoma. *J Neurooncol*. 2017;132(1):75-82.

Figures

Figure 1

Baseline brain MRI showed an ill-defined lesion around the fourth ventricle on a T2 weighted image (a). A contrast T1-weighted image showed mild enhancement of the lesion on the axial view (b).

Figure 2

HE staining and immunohistochemical staining ($\times 20$). (a) HE staining showed monotonous clear round cell proliferation without invasive characteristics, clearly separated with the cerebellar tissue and the delicate fibrillary matrix similar to neuropil. Immunohistochemical staining of the mass showed diffuse strong positivity of Syn (b). GFAP was focally positive (+/-) (c). Ki67<1% (d).

Figure 3

Five months after onset. Contrast MRI revealed a well-circumscribed 3.0 x 2.4 x 2.4 cm mass in the vermis with evident enhancement and perilesional oedema on the axial (a) and sagittal (b) views.

Figure 4

Contrast cranial MRI 8 months after radiotherapy. There was mild enhancement in the fourth ventricle on the axial (a) and sagittal (b) views.