

Spontaneous Regression of Tumor in an Optic Pathway Glioma Patient With Diencephalic Syndrome: Case Report and Literature Review

Rutao Luo

Beijing Children's Hospital, Capital Medical University and National Center for Children's Health

Tianlei Zhang

Beijing Children's Hospital, Capital Medical University and National Center for Children's Health

Wei Yang

Beijing Children's Hospital, Capital Medical University and National Center for Children's Health

Yuanqi Ji

Beijing Children's Hospital, Capital Medical University and National Center for Children's Health

Hailang Sun

Beijing Children's Hospital, Capital Medical University and National Center for Children's Health

Dapeng Li

Beijing Children's Hospital, Capital Medical University and National Center for Children's Health

Ming Ge (✉ geming88@126.com)

Beijing Children's Hospital, Capital Medical University

Case report

Keywords: optic pathway glioma, diencephalic syndrome, Spontaneous regression

Posted Date: December 16th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-1133705/v1>

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

[Read Full License](#)

Abstract

Background: Diencephalic syndrome (DS) can cause failure to thrive in pediatrics, which is mostly found in optic pathway gliomas (OPGs) patients. OPGs patients with DS always show a poor outcome.

Case presentation: We present the case of an OPG patient with DS who got a spontaneous regression without any treatment. A 6-month-old girl presented with failure to thrive for two months and visual dysfunction with bilateral horizontal nystagmus. MRI demonstrated a 42mm*37mm*36mm enhanced and lobulated lesion located in the sellar region with a clear boundary with surrounding tissues. OPG and DS was diagnosed according to her radiological examination and manifestation. Conservative follow-up was given. There was a spontaneous decrease in tumor size during follow-up. Symptom improves and the patient continues to have a good quality of life despite a moderate dysfunction of her left eye.

Conclusions: Conservative observation can be used as a treatment for some OPG patients, body weight may be a marker of the growth of tumor in OPG patients with DS.

Background

Optic pathway gliomas(OPGs)are a serious of special glioma originating in the visual pathway, usually found in children under 10 years old. OPGs can occur in any part of the visual pathway, including optic nerve, optic chiasma, optic tract, etc. It can also invade the adjacent hypothalamus, pituitary gland, third ventricle, lateral ventricle and other structures. Generally, OPGs in children are well differentiated, low grade. However, biological behaviors of OPGs are very unstable. OPGs in some patients can remain stable for a long time, while some can grow or spread rapidly, and some can even be found shrink or disappear naturally^[1-3]. Besides, OPGs can lead to various endocrine changes, including diencephalic syndrome, a rare cause of failure to thrive^[4]. The mechanism of the tumors' biological behavior and diencephalic syndrome (DS) is still not clear.

In this paper we present the case of a pediatric patient with an OPG and diencephalic syndrome, radiological regression and body weight growth was found during the follow-up time. The patient didn't receive any treatment, and had an excellent quality of life.

Case Presentation

History and Examination

A 6-month-old girl presented with poor appetite and no significant increase in body weight for two months. Also, she got horizontal nystagmus two days before his initial visit. Physical examination showed the patient got bilateral horizontal nystagmus and left esotropia. Poor visual acuity was found on the left side, but good acuity on the right side. MRI demonstrated a 42mm*37mm*36mm enhanced and lobulated lesion located in the sellar region with a clear boundary with surrounding tissues (Fig. 1A1, B1). The lesion had an abnormal signal with a large cystic component at the bottom. The optic chiasm

and optic nerve nearby could not be recognized. OPG and diencephalic syndrome was diagnosed by the history and characteristics of the MRI. Considered the low age and low body weight of the patient, patient's parents refused surgical treatment. Close follow-up was suggested.

One month later, horizontal nystagmus of the patient disappeared. However, the body weight declined continuously and came to the lowest point of 4.5 kg at his age of 1 year old (Fig. 2A). The patient's appetite and food intake increased gradually after the age of 1, and diencephalic syndrome improved obviously. The MRI of the patient at her age of 1 year and 11 months old showed the size of the tumor reduced to 38mm*35mm*23mm (Fig. 1A2, B2). A newly generated cystic component seemed like arachnoid cyst formed at the left temporal pole.

The girl's weight grew obviously to 10kg at her age of 2 years and 2 months. It seemed like she recovered from DS (Fig. 2B). Her parent didn't find a remarkable decrease of her vision and her horizontal nystagmus never reappeared. Flash visual evoked potential (FVEP) showed she had a moderate retardation of her left eye and a normal signal of the right eye.

Discussion

DS was a syndrome can lead to failure to thrive in pediatric patients, usually caused by neoplastic lesions located in the region of anterior hypothalamus. DS was closely related to the OPGs^[5]. Some scholars believed that patients with OPG with diencephalic syndrome have a poor prognosis. Gropman believes that the average survival time of untreated OPGs patients with DS was usually less than one year^[6]. Rakotonjanahary reported that girls with OPG and DS had a poor prognosis^[7]. In this case, without any treatment, the child did not have the aggravation of diencephalic syndrome and tumor growth. However, the tumor decreased and DS of the patient was improved, which was a relatively rare case.

Scholars had noticed the natural regression of tumors including some tumors of central nerve system. In 1992, Brzowski first reported a case of a 2-year-old boy with NF-1 who developed a natural regression of optic glioma^[8]. In early cases natural regression were mostly observed in patients with NF-1, so regression was thought to exist merely in these patients. Biological behavior was also regarded to be different in patients with NF-1 or not^[3]. However, in fact, patients with OPG without NF-1 also had natural regression. Parsa reported 13 children with optic glioma with natural regression, 9 of whom did not have NF-1^[9]. He believed that tumors shrank after a long period of chemotherapy or radiation should also be divided into natural regression, and that these treatments might trigger programmed death of tumor cells.

The mechanism of natural regression of OPG has not been clearly studied, and some scholars proposed some conjectures. Some researchers thought regression of tumors were mostly due to the shrink of cystic components. However, we had found the decrease of tumor cells in several cases. Other studies also found regression of astrocytoma located in other positions^[10, 11]. Some believed slow proliferation could be caused by enhanced immunocompetence and immune recognition of tumors^[9, 12]. Even though NF-1

did not show close relationship with regression of the OPGs in some studies, we thought it still playing an important role.

Since the mechanism of glioma spontaneous regression is currently unknown, it is controversial whether OPG patients can be treated with conservative observation for a period of time. Parsa thought optic pathway gliomas should be divided into hamartomas rather than neoplastic, and therefore should not be treated, even when there is an evidence of clinical or radiographic progression^[13, 14]. However, some scholars believed this kind of tumor still belong to the neoplastic^[15]. Alvord studied the outcome of OPG patients who didn't receive any treatment, found that most of them got a worse result^[16]. During the period of observation tumors could progress and cause irreversible consequences, decision should be made carefully. Some scholars speculated whether the growth of tumors could be judged by visual function of OPG patients. Study found that the reduction of tumor size was not always positively correlated with the patients' visual function, the vision of some OPG patients with natural regression could remain stable or even deteriorate^[17]. Some scholars believed that there was no strong correlation between tumor size and the visual function^[18]. During the treatment of this case, we found the original nystagmus of the patient disappeared and the emaciation caused by diencephalic syndrome gradually improved, but the left eye vision did not improve. Body weight curve of this patient was obviously different from children of the same age and was closely related to the growth of the tumor (Fig. 3). We thought visual acuity could not be used as a marker for the choice of conservative observation. Body weight may be a marker of tumor progression of OPG patients with diencephalic syndrome. Even though patients with diencephalic syndrome always showed a worse outcome, for children with recent weight gain, close follow-up may be a choice.

Conclusion

We believe that conservative observation can be used as a treatment for some OPG patients, it is not yet possible to distinguish which tumors will naturally recede. To our knowledge, this is the first case that spontaneous regression occurred in OPG patient accompanied with diencephalic syndrome. We think body weight may be a marker of the growth of tumor in this group of patients. Conservative treatment may be suit for those who got a recent weight gain. However, close follow-up is still necessary. Active treatment (chemotherapy, surgery, radiotherapy) should be considered when vision function getting worse or other serious symptom occur.

List Of Abbreviation

OPGs Optic pathway gliomas

DS diencephalic syndrome

FVEP flash visual evoked potential

Declarations

Availability of data and materials

The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

Ethical approval and consent to participate

The study protocol was approved by the Institutional Review Board of Beijing Children's Hospital, Capital Medical University and the procedures followed were in accordance with institutional guidelines. All participants had been informed before recruited and informed consent were signed and collected.

Consent for publication

Consent for publication has been signed by this children's parents.

Competing interest

We identify that no financial interests or affiliations with institutions, organization, or companies are mentioned or have impacts on the views expressed in the article. The authors declare that they have no competing interest.

Funding

This study was granted by the Beijing Hospitals Authority Clinical Medicine Development of special funding support, code: XMLX202144.

Author's contributions

MG proposed the conception and revised the manuscript.

RTL designed the work and was a major contributor in writing the manuscript.

TLZ mainly analyzed and interpreted the patient data.

WY undertook part of the work in data analysis.

HLS helped to interpret image and pathology data patient.

DPL made major contribution in follow up and revised the manuscript.

YQJ helped to draft the work and revised manuscript.

All authors read the manuscript and were informed before the manuscript was submitted.

Acknowledgements

Not applicable.

References

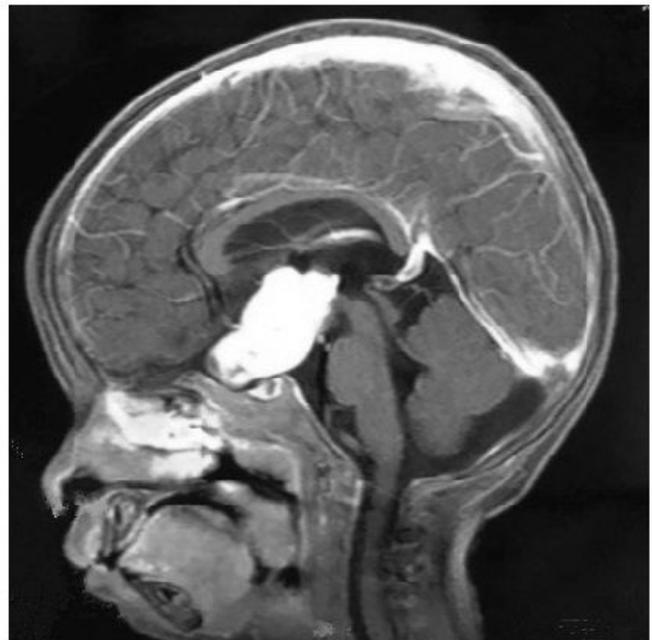
1. Chalouhi N, Jabbour P, Daou B, Starke RM, Shields B, Hasan DM. A New Protocol for Anticoagulation With Tirofiban During Flow Diversion. *Neurosurgery*. 2016. 78(5): 670-4.
2. Traber GL, Pangalu A, Neumann M, et al. Malignant optic glioma - the spectrum of disease in a case series. *Graefes Arch Clin Exp Ophthalmol*. 2015. 253(7): 1187-94.
3. Opocher E, Kremer LC, Da DL, et al. Prognostic factors for progression of childhood optic pathway glioma: a systematic review. *Eur J Cancer*. 2006. 42(12): 1807-16.
4. Tosur M, Tomsa A, Paul DL. Diencephalic syndrome: a rare cause of failure to thrive. *BMJ Case Rep*. 2017.
5. Waga S, Shimizu T, Sakakura M. Diencephalic syndrome of emaciation (Russell's syndrome). *Surg Neurol*. 1982. 17(2): 141-6.
6. Gropman AL, Packer RJ, Nicholson HS, et al. Treatment of diencephalic syndrome with chemotherapy: growth, tumor response, and long term control. *Cancer*. 1998. 83(1): 166-72.
7. Rakotonjanahary J, De Carli E, Delion M, et al. Mortality in Children with Optic Pathway Glioma Treated with Up-Front BB-SFOP Chemotherapy. *PLoS One*. 2015. 10(6): e0127676.
8. Brzowski AE, Bazan C, Mumma JV, Ryan SG. Spontaneous regression of optic glioma in a patient with neurofibromatosis. *Neurology*. 1992. 42(3 Pt 1): 679-81.
9. Parsa CF, Hoyt CS, Lesser RL, et al. Spontaneous regression of optic gliomas: thirteen cases documented by serial neuroimaging. *Arch Ophthalmol*. 2001. 119(4): 516-29.
10. Lenard HG, Engelbrecht V, Janssen G, Wechsler W, Tautz C. Complete remission of a diffuse pontine glioma. *Neuropediatrics*. 1998. 29(6): 328-30.
11. Rao S, Constantini S, Gomori JM, Siegal T, Epstein F. Spontaneous involution of an intra-axial brain stem lesion: a case report. *Pediatr Neurosurg*. 1995. 23(5): 279-81; discussion 282.
12. Kernan JC, Horgan MA, Piatt JH, D'Agostino A. Spontaneous involution of a diencephalic astrocytoma. *Pediatr Neurosurg*. 1998. 29(3): 149-53.
13. Parsa CF, Givrad S. Pilocytic astrocytomas as hamartomas: implications for treatment. *Br J Ophthalmol*. 2008. 92(1): 3-6.
14. Parsa CF. Why optic gliomas should be called hamartomas. *Ophthalmic Plast Reconstr Surg*. 2010. 26(6): 497.
15. Liu GT, Katowitz JA, Rorke-Adams LB, Fisher MJ. Optic pathway gliomas: neoplasms, not hamartomas. *JAMA Ophthalmol*. 2013. 131(5): 646-50.
16. Alvord EC, Lofton S. Gliomas of the optic nerve or chiasm. Outcome by patients' age, tumor site, and treatment. *J Neurosurg*. 1988. 68(1): 85-98.

17. Thiagalingam S, Flaherty M, Billson F, North K. Neurofibromatosis type 1 and optic pathway gliomas: follow-up of 54 patients. *Ophthalmology*. 2004. 111(3): 568-77.
18. Liu GT, Lessell S. Spontaneous visual improvement in chiasmal gliomas. *Am J Ophthalmol*. 1992. 114(2): 193-201.

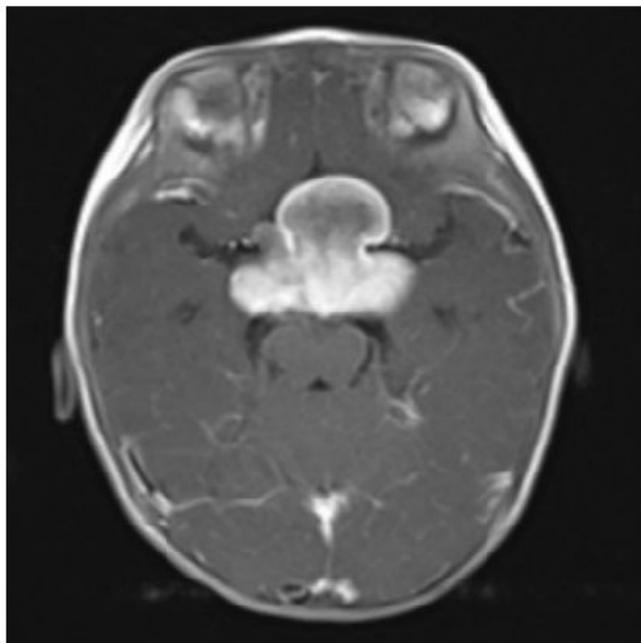
Figures



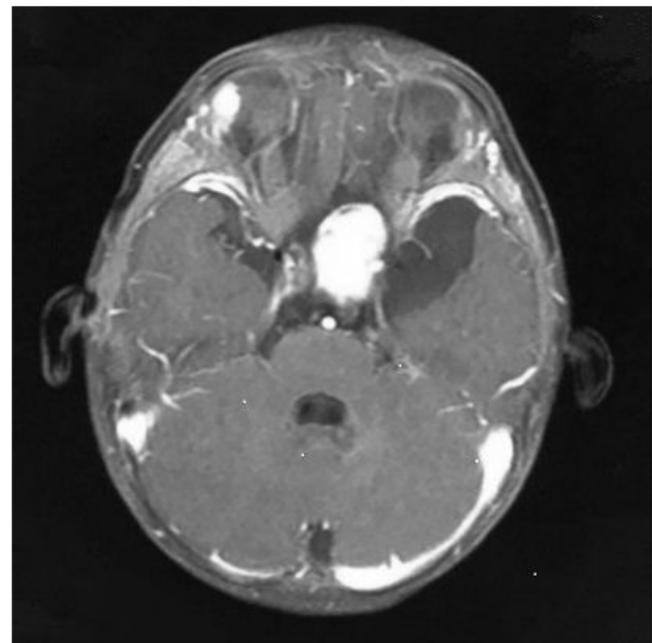
A1



A2



B1



B2

Figure 1

A1 and B1 were sagittal and axial enhanced MRI of the patient at her age of 6 months and 20 days. A2 and B2 were the sagittal and axial image when she was 1 year and 11 months old. Tumor size shrunk obviously from 42mm*37mm*36mm to 38mm*35mm*23mm. A newly generated cystic component seemed like arachnoid cyst (18mm*35mm*26mm) formed at the left temporal pole,



A



B

Figure 2

A. At the age of one year, the child weighed 4.5kg, showing extreme weight loss and malnutrition due to subcutaneous fat loss. B. At the age of 2 years and 2 months, the child weighed 10kg, and the malnutrition was significantly improved.

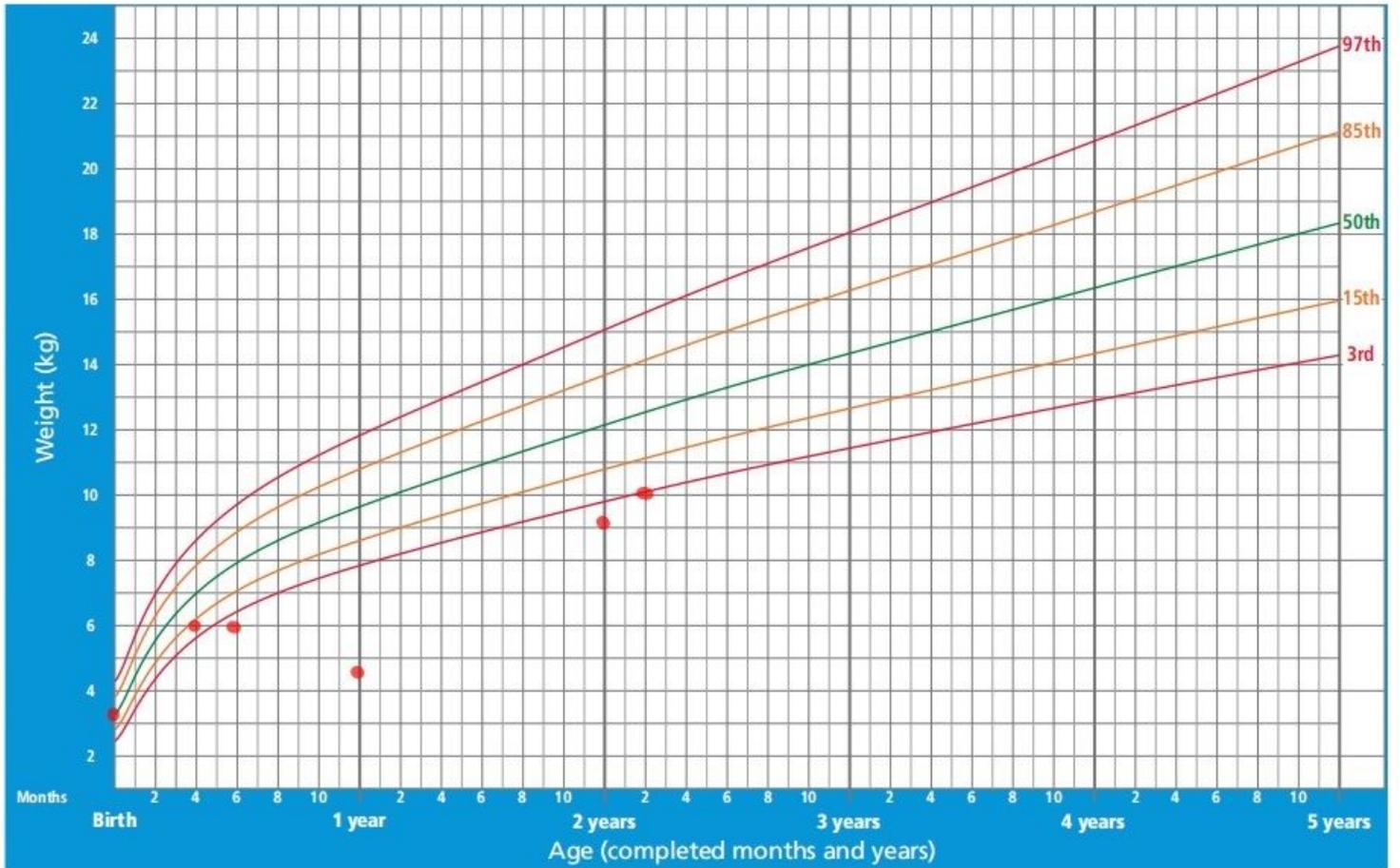


Figure 3

Comparison between the patient and the percentile of children with the same age and gender in the change trend chart. The patient's birth weight was 3.35kg, the weight did not increase from 4 to 6 months, and the lowest weight was 4.5kg at 1 year old. The weight increased after 1 year old, 9.3kg at 2 years old, and 10kg at 2 years and 2 months old.