Int J Clin Exp Med 2017;10(5):8410-8414
www.ijcem.com /ISSN:1940-5901/IJCEM0050048

Case Report

Lateral ventricular chordoid glioma in a pediatric patient: a case report and literature review

Fan Chen[1*], Cong Li[2*], Xiaoliang Liu[1*], Xiaoyu Yang[1*], Zhen Guo[3*], Dongdong Wang[1*], Dawei Chen[1*]

Departments of [1]Neurosurgery, [2]Nephrology, [3]Cardiovascular Medicine, First Hospital of Jilin University, Changchun 130021, China. [*]Equal contributors.

Received February 3, 2017; Accepted March 17, 2017; Epub May 15, 2017; Published May 30, 2017

Abstract: Chordoid glioma is a rare tumor exhibiting both glial and chordoid features. This entity is generally considered to occur exclusively in the region of the third ventricle, however, that arising in the lateral ventricle is extremely rare. Herein, we report a case of lateral ventricular chordoid glioma in a pediatric patient. Additionally, relevant literatures were reviewed. An intracranial space-occupying lesion was discovered in a 10-year-old asymptomatic boy. Brain computed tomography (CT) and magnetic resonance imaging (MRI) revealed a nodular mass in the frontal horn of the right lateral ventricle. Histopathological examination and immunohistochemical staining were consistent with a diagnosis of chordoid glioma. No adjuvant radiotherapy or chemotherapy was performed. Preoperative identification of chordoid gliomas depending on neuroradiological profiles is challenging, and definitive diagnosis still needs histopathological evidence. Surgical resection should be the choice of treatment, and prognosis is favorable.

Keywords: Chordoid glioma, lateral ventricle, pediatric, case report

Introduction

Chordoid glioma is a rare intracranial neoplasm, which exhibits both glial and chordoid features [11, 15, 21]. This entity is generally considered to occur exclusively in the region of the third ventricle and hypothalamus; however, chordoid glioma in the lateral ventricle is extremely rare [1, 12, 16, 20]. Up to date, less than one hundred cases with chordoid glioma have been reported in previous literatures; the majority of these patients were adult with a female predominance, and there were only 4 pediatric cases [3, 4, 7, 15]. Due to the rarity of chordoid glioma, its clinical manifestations, radiological features, histopathological characteristics, and diagnosis are not well elucidated.

Herein, we report a case of lateral ventricular chordoid glioma in a pediatric patient. Additionally, relevant literatures were reviewed.

Case report

A 10-year-old boy presented to us with an incidentally discovered intracranial space-occupying lesion. No significant symptom was complained. Physical examinations were all normal. Brain computed tomography (CT) scanning showed an ill-defined isodense mass in the right frontal horn of lateral ventricle (**Figure 1A**). Brain magnetic resonance imaging (MRI) revealed a nodular mass in the right frontal horn of lateral ventricle, which appeared slight hyperintensity on T1-weighted imaging, slight hyperintensity on T2-weighted imaging, and isointensity on diffusion-weighted imaging (DWI). After administration of contrast medium, heterogeneous enhancement was notable (**Figure 1B-E**). A suspected diagnosis of subependymal giant-cell astrocytoma was made, and surgical resection was scheduled.

A supratentorial craniotomy was performed under the guidance of a MRI-assisted navigation system. The intraoperative findings showed that the tumor was reddish in color and hard in nature. Because of the tumor's adhesion to the lateral wall of the lateral ventricle and septum pellucidum and the ill-defined margins, *en bloc* resection was difficult to achieve, and thus the tumor was subtotally removed. Postoperatively, the boy developed psychiatric symptoms mani-

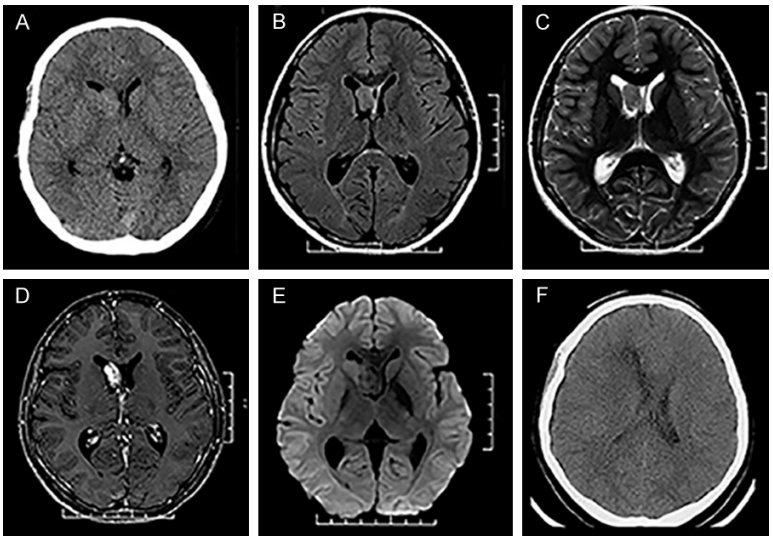

Figure 1. Radiological profiles of the boy with chordoid glioma. Brain CT scanning showed an ill-defined isodense mass in the right frontal horn of lateral ventricle (A). Brain MRI revealed a nodular mass in the right frontal horn of lateral ventricle, which appeared slight hyperintensity on T1-weighted imaging (B) and slight hyperintensity on T2-weighted imaging (C). After administration of contrast medium, heterogeneously remarkable enhancement was observed (D). The tumor was isointensity on diffusion-weighted imaging (E). The follow-up CT 6 months after surgery showed no recurrence (F).

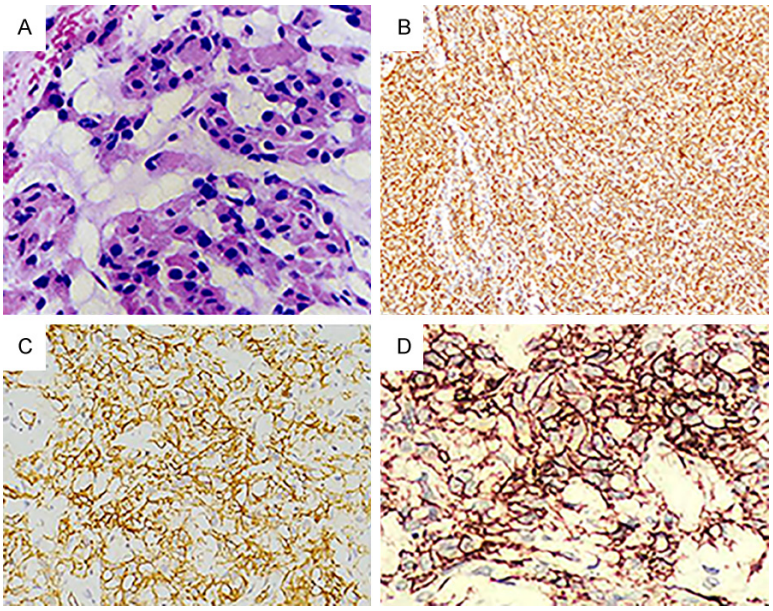

Figure 2. Histopathological examination of the resected specimen. Microscopically, the tumor is composed of clusters and cords of epithelioid cells and abundant myxoid and mucinous stroma; the nuclei are medium-sized and isomorphous (A, hematoxylin and eosin stain). Immunostaining shows the tumor was positive for vimentin (B), GFAP (C), and EMA (D).

festing as dysphoria and hyperactivity that aggravated during the night. Intravenous injection of small-dosage diazepam was administered, and these psychiatric symptoms disappeared 6 days later.

Histopathological examination showed cords and clusters of tumor cells and the nuclei were medium-sized and isomorphous (**Figure 2A**). Immunohistochemical staining showed immunopositivity for glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), and vimentin (**Figure 2B-D**). The Ki-67 proliferation fraction was approximately 2%. These findings were consistent with a diagnosis of chordoid glioma.

No adjuvant radiotherapy or chemotherapy was performed. During a follow-up period of 6 months, no tumor progression or recurrence was noted (**Figure 1F**).

Discussion

Chordoid glioma was firstly described by Brat *et al.* in 1998 [2]. In the 2007 version of the World Health Organization classification of central nervous system tumors included chordoid glioma in the category of neuroepithelial tumors of uncertain histogenesis, corresponding to grade II [14]. This entity is extremely rare, and up to date there have been only 86 cases identified in English literatures. The majority of the previously reported chordoid gliomas were located in the third ventricle. The unusual locations included left thalamus [8], left temporo-parietal lobe [4], and the occipital horn of the right lateral ventricle [7]. Chordoid gliomas are more commonly detected in adults with female predominance [22]. The onset age ranged from 5 to 72 years, with a median age of 44 years; till

Table 1. Summary of chordoid gliomas reported in literatures and the current case

Characteristics	Number (n)	Percentage (%)
Gender		
Male	36/87	41.38%
Female	51/87	58.62%
Age		
Pediatric	5/87	5.7%
Adult	82/87	94.3%
Location		
Third ventricle	83/87	95.4%
Left thalamus	1/87	1.15%
Left temporo-parietal lobe	1/87	1.15%
Right occipital horn of lateral ventricle	1/87	1.15%
Right frontal horn of lateral ventricle	1/87	1.15%
MRI T1-weighted imaging		
Hypointensity	6/27	22.22%
Isointensity	7/27	25.93%
Hyperintensity	13/27	48.15%
Heterogeneous intensity	1/27	3.7%
MRI T2-weighted imaging		
Hypointensity	5/32	15.63%
Isointensity	8/32	25%
Hyperintensity	16/32	50%
Heterogeneous intensity	3/32	9.38%
MRI enhancement		
Homogeneous	30/40	75%
Heterogeneous	10/40	25%
Immunehistochemical staining		
GFAP diffuse positivity	70/71	98.59%
GFAP focal positivity	1/71	1.41%
Vimentin positivity	51/51	100%
EMA diffuse positivity	7/58	12.07%
EMA focal positivity	35/58	67.31%
S-100 diffuse positivity	22/46	47.83%
S-100 focal positivity	14/46	30.43%
CD34 diffuse positivity	31/41	75.61%
CD34 focal positivity	9/41	21.95%
Synaptophys infocal positivity	2/26	7.69%
Cytochrome diffuse positivity	6/22	27.27%
Cytochrome focal positivity	14/22	63.64%
NSE diffuse positivity	1/8	12.5%
NSE focal positivity	3/8	37.5%
Surgical resection extent		
Gross total resection	39/81	48.15%
Subtotal resection	33/81	40.74%
Biopsy	9/81	11.11%
Outcome		
No recurrence	8/81	9.9%
Recurrence	73/81	90.1%

now, there are only four pediatric cases [3, 4, 7, 15]. The present 10-year-old boy is the fifth pediatric patient diagnosed with chordoid glioma. There were 51 females and 36 males reported in literatures, the male-to-female ratio being 1:1.42.

The clinical manifestations of chordoid gliomas are nonspecific, and the symptoms are associated with the space-occupying compression that is related to the location and size of the tumor. As chordoid gliomas mostly occur in the third ventricle, the most common manifestation is obstructive hydrocephalus. When the tumor compresses the hypothalamus and optic nerve system, the patient may develop endocrinal disorders and visual disturbances. Interestingly, in the current case, the boy was asymptomatic, and the silent clinical course may indicate an indolent nature of chordoid glioma.

Radiologically, chordoid gliomas have no characteristic features. On CT, these tumors are usually well-defined, iso- to hyperdense, and homogeneously enhanced [1, 11]. Additionally, in previous literatures, three cases showed tumor apoplexy [4, 5, 10]. On magnetic resonance T1-weighted imaging, chordoid gliomas appeared isointensity in 48.15% cases, hyperintensity in 25.93% cases, hypointensity in 22.22% cases, and heterogeneous intensity in 3.70% cases. On T2-weighted imaging, chordoid gliomas appeared isointensity in 25.00% cases, hyperintensity in 50.00% cases, hypointensity in 15.63% cases, and heterogeneous intensity in 9.38% cases. After administration of gadoliniun contrast agent, the homogeneous and heterogeneous enhancement was noted in 75% and 25% cases, respectively. The majority of tumors were solid, and cystic component was observed in

26% cases. Calcification was rarely seen [5, 6, 9], and only two cases were found with significant peritumoral edema [13, 23]. In relevant studies, magnetic resonance spectroscopy of chordoid glioma demonstrated increased Choline peak and decreased NAA peak, indicating a histologically low-grade nature [17]. The radiological findings in our case were consistent with those in the previous reports.

The definitive diagnosis of chordoid gliomas still depends on histopathological evidence. Microscopically, the tumor is composed of clusters and cords of epithelioid cells, abundant myxoid and mucinous stroma, and capillary network; the nuclei are medium-sized and isomorphous with mild nuclear pleomorphism, and there is generally no mitosis, vascular endothelial hyperplasia, or necrosis. Throughout the tumor tissue, there were diffuse infiltrates of lymphocytes and plasma cells [2]. The immunohistochemical staining showed diffuse positivity for GFAP in 70 of the 71 available cases (98.59%) and focal positivity for GFAP in one case (1.41%). All the 51 cases with vimentin staining were positive (100%). The positive rate for EMA, S-100 protein, CD34, synaptophysin, cytochrome, and neuron specific enolase (NSE) was 79.38%, 78.26%, 97.56%, 7.69%, 90.91%, and 50%, respectively. Except for the only case showing a high Ki-67 proliferative fraction of 20% [18], all the tumors exhibited a low Ki-67 percentage (≤5%) indicating a benign biological behavior. The detailed information of literature review was summarized in **Table 1**.

The histogenesis of chordoid gliomas is still controversial. Brat *et al.* found the majority of these entities were located in the third ventricle and the ultrastructural features was similar to those described for the secretory ependymal cells; thus, they speculated chordoid glioma may represent a specific subtype of ependymoma [2]. Sato *et al.* proposed that chordoid glioma in the third ventricle may originate from tanycytes-a specialized form of ependymal cells [19]. Noteworthily, the location of chordoid glioma in the current case is quite unusual, and we considered it might be related to the heterotopia of the tumor cells in the ventricular system.

Due to the rarity of this entity, the optimal treatment has not yet been outlined. Surgical resection is the mainstream treatment, and maximal safe resection should be the goal. Although chordoid gliomas are benign tumors with the possibility of surgical cure and favorable prognosis, the specific location complicates surgical removal and imparts a significant risk of neurologic injury. The selection of specific approach should be individually designed depending on the growth pattern of tumors [7, 8]. The efficacy of adjuvant radiotherapy and chemotherapy is still unclear. In literatures, a total of 81 patients underwent surgical treatment, and the mean follow-up period was 21.59 months. Gross total resection was achieved in 39 (48.15%) cases, and no adjuvant radiotherapy or chemotherapy was administrated in these cases; during follow-up period, no recurrence was noted. Subtotal resection was achieved in 33 (40.74%) cases and biopsy was performed in 9 (11.11%) cases; recurrence or progression of remnant tumor was noted in 8 (19.05%); 14 among these 42 cases underwent radiotherapy, among which 6 patients (42.86%) were observed with recurrence. However, due to the limited clinical evidence, the definitive role of radiotherapy and the relevant prognosis cannot be concluded and still needs further research involving a much larger cohort.

Conclusion

Clinical manifestations and radiological features of chordoid gliomas are non-specific. Preoperative identification of chordoid gliomas is challenging, and definitive diagnosis still needs histopathological examinations. Surgical resection should be the choice of treatment and the approach should be individually designed. Following adequate resection, chordoid gliomas may be associated with a favorable prognosis.

Disclosure of conflict of interest

None.

Address correspondence to: Dawei Chen, Department of Neurosurgery, First Hospital of Jilin University, 71 Xinmin Avenue, Changchun 130021, China. Tel: 0086-043185398815; Fax: 0086-04318539-8815; E-mail: cdw2006@tom.com

References

[1] Ampie L, Choy W, Lamano JB, Kesavabhotla K, Mao Q, Parsa AT, Bloch O. Prognostic factors for recurrence and complications in the surgi-

cal management of primary chordoid gliomas: a systematic review of literature. Clin Neurol Neurosurg 2015; 138: 129-136.
[2] Brat DJ, Scheithauer BW, Staugaitis SM, Cortez SC, Brecher K, Burger PC. Third ventricular chordoid glioma: a distinct clinicopathologic entity. J Neuropathol Exp Neurol 1998; 57: 283-290.
[3] Castellano-Sanchez AA, Schemankewitz E, Mazewski C, Brat DJ. Pediatric chordoid glioma with chondroid metaplasia. Pediatr Dev Pathol 2001; 4: 564-567.
[4] Goyal R, Vashishta RK, Singhi S, Gill M. Extraventricular unusual glioma in a child with extensive myxoid change resembling chordoid glioma. J Clin Pathol 2007; 60: 1294-1295.
[5] Horbinski C, Dacic S, Mclendon RE, Cieply K, Datto M, Brat DJ, Chu CT. Chordoid glioma: a case report and molecular characterization of five cases. Brain Pathol 2009; 19: 439-448.
[6] Iwami K, Arima T, Oooka F, Fukumoto M, Takagi T, Takayasu M. Chordoid glioma with calcification and neurofilament expression: case report and review of the literature. Surg Neurol 2008; 71: 115-120.
[7] Jain D, Sharma MC, Sarkar C, Suri V, Rishi A, Garg A, Vaishya S. Chordoid glioma: report of two rare examples with unusual features. Acta Neurochir (Wien) 2008; 150: 295-300.
[8] Jin WK, Kim JH, Choe G, Kim CY. Chordoid glioma: a case report of unusual location and neuroradiological characteristics. J Korean Neurosurg Soc 2010; 48: 62-65.
[9] Jung TY, Jung S. Third ventricular chordoid glioma with unusual aggressive behavior. Neurol Med Chir (Tokyo) 2007; 46: 605-608.
[10] Kawasaki K, Kohno M, Inenaga C, Sato A, Hondo H, Miwa A, Fujii Y, Takahashi H. Chordoid glioma of the third ventricle: a report of two cases, one with ultrastructural findings. Neuropathology 2009; 29: 85-90.
[11] Ki SY, Kim SK, Heo TW, Baek BH, Kim HS, Yoon W. Chordoid glioma with intraventricular dissemination: a case report with perfusion MR imaging features. Korean J Radiol 2015; 17: 142.
[12] Kobayashi T, Tsugawa T, Hashizume C, Arita N, Hatano H, Iwami K, Nakazato Y, Mori Y. Therapeutic approach to chordoid glioma of the third ventricle. Neurol Med Chir 2013; 53: 249-255.
[13] Liu WP, Cheng JX, Yi XC, Zhen HN, Fei Z, Li Q, Zhang X. Chordoid glioma: a case report and literature review. Neurologist 2011; 17: 52-56.
[14] Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, Scheithauer BW, Kleihues P. The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol 2007; 114: 97-109.
[15] Morais BA, Menendez DF, Medeiros RS, Teixeira MJ, Lepski GA. Chordoid glioma: case report and review of the literature. Int J Surg Case Rep 2015; 57: 168-171.
[16] Ni HC, Piao YS, Lu DH, Fu YJ, Ma XL, Zhang XJ. Chordoid glioma of the third ventricle: four cases including one case with papillary features. Neuropathology 2013; 33: 134-139.
[17] Qixing F, Peiyi G, Kai W, Xuzhu C, Xiangde M, Jianping D. The radiological findings of chordoid glioma: report of two cases, one case with MR spectroscopy. Clin Imaging 1900; 39: 1086-1089.
[18] Sanches P, Yamashita S, Freitas CCMD, Resende LADL. Glioma cordoide do terceiro ventrículo: descrição de um novo caso; Chordoid glioma of the third ventricle: a new case report. Radiol Bras 2012; 45: 288-290.
[19] Sato K, Kubota T, Ishida M, Yoshida K, Takeuchi H, Handa Y. Immunohistochemical and ultrastructural study of chordoid glioma of the third ventricle: its tanycytic differentiation. Acta Neuropathol 2003; 106: 176-180.
[20] Smith AB, Smirniotopoulos JG, Horkanyne-Szakaly I. From the radiologic pathology archives: intraventricular neoplasms: radiologic-pathologic correlation. Radiographics 2013; 33: 21-43.
[21] Soffietti R, Ruda R, Reardon D. Rare glial tumors. Handb Clin Neurol 2016; 134: 399-415.
[22] Vanhauwaert DJ, Clement F, Van DJ, Deruytter MJ. Chordoid glioma of the third ventricle. Acta Neurochir 2008; 150: 1183-1191.
[23] Vij M, Jaiswal S, Jaiswal AK, Jain M, Behari S. Chordoid glioma of the third ventricle: a case report with review of literature. Neurol India 2011; 59: 469-471.