Surgical management and clinical outcomes of cerebellar liponeurocytomas-a report of 7 cases and a pooled analysis of individual patient data

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Research Article

Keywords: Cerebellar liponeurocytomas, gross total resection, follow-up, recurrence, radiotherapy

Posted Date: November 11th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-1047305/v1>

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Abstract

Objective

Cerebellar liponeurocytomas (CLPNs) are very rare. Limited studies described this disease and their treatment protocol remain unclear. To better understand the disease, we review the clinical features and outcomes, and propose a treatment protocol based on previously reported cases and cases from our institute.

Methods

The clinical data were obtained from 7 patients with pathologically confirmed CLPNs, who accepted surgical treatment in our institute between November 2011 and June 2021. We also reviewed the literature and 75 patients with CLPNs were identified between September 1993 and June 2021. Risk factors for Progression-free survival (PFS) were evaluated in the pooled cohort.

Results

The authors' cohort included 4 males and 3 females with a mean age of 43.9 ± 14.5 (range: 29-64 years). 3 cases are located in lateral ventricle and 4 cases are located in cerebellum. All 7 cases achieved gross total resection (GTR) and radiotherapy was administered to 2 cases. After a mean follow-up of 44.9 ± 44.4 months, all patients remained well with no recurrence or death. For the reported 75 patients, there are 35 males and 40 females with a mean age of 46.2 ± 13.6 years (range: 6-77 years). Biopsy, gross total resection (GTR) and non-GTR were achieved in 1 (1.3%), 50 (66.7%), and 24 (32%) patients, respectively. Radiotherapy was administered to 16 cases and chemotherapy was administered to only 1 case. After a mean follow-up of 47.5 ± 51.5 months, 3 patients died and tumor recurrence occurred in 17 patients. Multivariate Cox analysis revealed that non-GTR predicted a poor PFS ($p=0.030$). Kaplan-Meier analysis showed that GTR was significantly associated with better PFS ($p=0.0084$). PFS rates at 1, 5, 10 years were 92.7%, 78.0%, 23.8% respectively.

Conclusions

Cerebellar liponeurocytomas (CLPNs) are very rare brain tumors. Although they have favorable clinical prognosis, the recurrence is relatively high. GTR should be the first choice and close follow-up is necessary. Postoperative radiotherapy could not improve PFS in this study. A larger cohort is needed to verify our findings.

Introduction

Cerebellar liponeurocytoma (CLPN) is very rare. It was first reported in 1978 by Bechtel et al[7]. CLPN was firstly defined as a distinct entity according to the WHO 2000 classification of tumors of central nervous system[57]. In 2021, the lesion was categorized as a neuronal or neurocytic tumor and classified as WHO grade II tumor[45]. It usually occurred in adults and has a relatively favorable clinical outcome[44]. Although several cases have been reported previously, the treatment protocol still remained uncertain. Therefore, we retrospectively reviewed clinical data of 7 cases with CLPNs in our institute and reviewed the literature. We aimed to describe the clinical characteristics of CLPN, evaluate the risk factors for progression-free survival (PFS), and propose a treatment protocol for CLPN.

Material And Methods

We retrospectively reviewed the clinical data and radiographs of 7 cases with CLPNs who accepted surgical treatment at Beijing Tiantan Hospital between November 2011 and June 2021. The clinical data included age, sex, symptoms, duration of symptoms, tumor location, the extent of tumor resection, pathological results, radiotherapy and chemotherapy etc. The extent of tumor resection defined as gross total resection (GTR) and non-GTR based on pre- and postoperative MRIs. The pathological diagnosis of CLPNs was determined by the Department of Neuropathology at Beijing Neurosurgical Institute. This study was approved by the internal review board of Beijing Tiantan Hospital. For the literature review, we searched the PubMed database using the keywords "liponeurocytoma(s)" between September 1993 and June 2021 and a total of 75 cases were included. The risk factors were evaluated with univariate and multivariate Cox regression analysis of the pooled cases. The Kaplan-Meier method was used to determine the PFS differences with p-values calculated from the log-rank test. Analyses were performed using SPSS statistical software with significance set at $p < 0.05$.

Results

The authors' cohort include 4 (57.1%) males and 3 (42.9%) females, with a mean age of 43.9 ± 14.5 years (range: 29-64 years). The mean duration of symptoms was 10.7 ± 7.9 months (range: 1-24 months). Preoperative symptoms included headache (6/7, 85.7%) and gait difficulty (1/7, 14.3%). 4 (57.1%) cases were located in cerebellum and 3 (42.9%) cases located in lateral ventricle. The family histories of all patients were unremarkable. On MRI, all 7 patients showed heterogeneous signals of T1WI and T2WI and heterogeneous enhancement was displayed in 6 (85.7%) patients. Gross-total resection (GTR) were achieved in all patients. After a mean follow-up of 44.9 ± 44.4 months, all patients remained well with no recurrence or death. Immunohistochemically, Ki-67 index were relatively low for 6 available patients. The clinical data are summarized in Table 1.

Table 1
Clinical characteristics of 7 cases with CLPNs

Case	Sex	Age	symptoms	Duration of symptoms (months)	Location	Treatment	Ki-67 (%)	Multifocality	Recurrence	Outcome	PFS (months)	FU (months)
1	F	48	Headache	18	Cerebellum	GTR	<5	No	No	Alive	120.8	120.8
2	M	29	Headache	1	Lateral ventricle	GTR	5	No	No	Alive	83.4	83.4
3	F	64	Headache	24	Cerebellum	GTR+RT	7.5	No	No	Alive	57.3	57.3
4	F	30	Headache	12	Lateral ventricle	GTR	7	No	No	Alive	29.6	29.6
5	M	57	Gait difficulty	8	Cerebellum	GTR+RT	5	No	No	Alive	13.1	13.1
6	M	50	Headache	6	Cerebellum	GTR	5.5	No	No	Alive	5.3	5.3
7	M	29	Headache	6	Lateral ventricle	GTR	NA	No	No	Alive	4.6	4.6
PFS: progression-free survival; FU: follow up; F: female; M: male; NA: not available; GTR; gross total resection; RT: radiotherapy												

For the literature review, 75 cases of CLPNs were identified (Table 2). Among them, 35 cases were males and 40 cases were females with a mean age of 46.2 ± 13.6 years (range: 6-77 years). On Magnetic Resonance Imaging (MRI), 58 cases (77.3%) were located in infratentorial region (including cerebellum, CPA area and fourth ventricle) and 17 cases (22.7%) located in supratentorial region (including lateral ventricle and frontal lobe). Biopsy, gross total resection (GTR) and non-GTR were achieved in 1 (1.3%), 50 (66.7%), and 24 (32%) patients, respectively. Radiotherapy was administered to 16 cases and chemotherapy was administered to only 1 case. After a mean follow-up of 47.2 ± 50.4 months, tumor recurrence occurred in 17 (26.6%) patients (Table 3).

Table 2
Clinical data of 75 cases with cerebellar liponeurocytoma from September 1993 to June 2021

Authors and year	Sex/age (years)	Location	Multifocality	Ki-67 (%)	Treatment	Recurrence	Outcome	PFS (months)	FU (months)
Hirono et al., 2021[34]	F/44	Cerebellum, third and fourth ventricle	Yes	2	Non-GTR	Yes	Alive	7	7
Al-Umran et al., 2021[4]	M/24	Lateral ventricle	No	10	GTR+RT	No	Alive	12	12
Abuzneid et al., 2021[1]	F/50	Cerebellum	No	NA	GTR	No	Alive	1	1
Deora et al., 2019[21]	M/55	Cerebellum	No	4.5	Non-GTR	No	Alive	48	48
Hamzaoglu et al., 2018[32]	F/55	Cerebellum	No	2	GTR	No	Alive	-	-
Gembruch et al., 2018[26]	M/39	Cerebellum	No	2	GTR	No	Alive	15	15
Chiaromonte et al., 2018[17]	F/35	Cerebellum	No	6	GTR	No	Alive	48	48
Cai et al., 2018[14]	M/11	Frontal lobe	No	1.5	GTR	No	Alive	74	74
Börekci et al., 2018[11]	F/62	Lateral ventricle	No	NA	GTR	No	Alive	6	6
Ben Nsir et al., 2018[10]	F/24	Lateral ventricle	No	3	GTR	No	Alive	12	12
Xu et al., 2017[69]	M/29	Lateral ventricle	No	3.5	GTR	No	Alive	24	24
	F/48	Cerebellum	No	1.5	GTR	No	Alive	60	60
Xu et al., 2017[68]	F/49	Cerebellum	No	1	GTR	NA	NA	NA	NA
	F/45	Cerebellum	No	<1	GTR	No	Alive	54	54
	M/29	Lateral ventricle	No	5	GTR	No	Alive	17	17
Tucker et al., 2017[65]	F/41	Cerebellum	No	4	GTR	NA	NA	NA	NA
Sivaraju et al., 2017[61]	M/37	Cerebellum	Yes	6	Non-GTR+RT	No	Alive	24	24
Hermann et al., 2017[33]	F/77	Cerebellum	No	12	GTR	No	Alive	24	24
Wolf et al., 2016[67]	F/37	Cerebellum	No	<1	Non-GTR	NA	NA	NA	NA
Pikis et al., 2016[55]	F/72	Cerebellum	No	12.5	GTR	No	Alive	36	36
Nzegwu et al., 2016[47]	F/6	Cerebellum	No	NA	GTR	No	Alive	-	-
Takami et al., 2015[64]	M/59	Cerebellum	No	2	Non-GTR	No	Alive	24	24
Scoppetta et al., 2015[60]	M/42	Cerebellum	Yes	NA	Biopsy	NA	NA	NA	NA
Radke et al., 2015[56]	F/59	Cerebellum	No	3	Non-GTR	Yes	Alive	180	204
Kakkar et al., 2015[39]	M/35	Cerebellum	No	3.5	GTR	No	Alive	-	-
Ruiz Ginés r et al., 2014[59]	M/33	Lateral ventricle	No	<1	GTR	NA	NA	NA	NA
Oudrhiri et al., 2014[49]	F/64	Cerebellum	No	<1	Non-GTR	No	Alive	24	24
Karabagli et al., 2014[40]	M/34	Lateral ventricle	No	1.5	GTR	No	Alive	24	24
Non-GTR: non-gross total resection, *: patients died of postoperative complications, including intraventricular bleeding, intracranial hemorrhage and pneumonia and sepsis.									

Authors and year	Sex/age (years)	Location	Multifocality	KI-67 (%)	Treatment	Recurrence	Outcome	PFS (months)	FU (months)
Ben Nsir et al., 2014[9]	F/59	Cerebellum	No	3	GTR	No	Alive	18	18
Pelz et al., 2013[54]	F/54	Cerebellum	Yes	NA	Non-GTR	Yes	Alive	24	48
Dey et al., 2013[22]	F/30	Cerebellum	No	3	Non-GTR	Yes	Alive	6	6
Beizig et al., 2013[8]	F/45	Cerebellum	No	NA	GTR	No	Alive	18	18
Guan et al., 2012[30]	M/50	Cerebellum	No	NA	GTR	NA	NA	NA	NA
Chung et al., 2012[18]	M/49	Cerebellum	No	13.68	Non-GTR +RT	No	Alive	12	12
Anghileri et al., 2012[6]	F/40	Cerebellum	No	1.5	GTR	Yes	Alive	96	132
	F/59	Cerebellum	No	1.5	Non-GTR	Yes	Alive	15	120
Gupta et al., 2011[31]	F/45	Lateral ventricle	No	3	GTR	NA	NA	NA	NA
den Hollander et al., 2011[20]	M/43	Lateral ventricle	No	NA	GTR	NA	NA	NA	NA
Chakraborti et al., 2011[15]	M/36	Lateral ventricle	No	<1	GTR	Yes	Alive	112	136
	M/30	Lateral ventricle	No	<1	GTR	NA	NA	NA	NA
	M/32	Lateral ventricle	No	<1	GTR	NA	NA	NA	NA
	F/45	Cerebellum	No	<1	GTR	NA	NA	NA	NA
Pankaj et al., 2010[51]	M/35	Lateral ventricle	No	NA	GTR	No	Alive	-	-
Patel et al., 2009[53]	F/40	Cerebellum	No	NA	GTR	No	Alive	36	36
Pasquale et al., 2009[52]	M/64	Cerebellum	No	15	GTR	Yes	Alive	42	90
Limaiem et al., 2009[43]	M/42	Cerebellum	No	2	GTR	Yes	Alive	120	132
Châtillon et al., 2009[16]	F/42	Cerebellum	No	10	GTR+RT	No	Alive	36	36
Kuchelmeister et al., 2006[42]	M/35	Lateral ventricle	No	NA	Non-GTR	NA	NA	NA	NA
Owler et al., 2005[50]	M/34	Fourth ventricle	No	NA	GTR+RT	No	Alive	8	8
Buccoliero et al., 2005[12]	M/64	Cerebellum	No	~10	GTR+RT	No	Alive	5	5
Aker et al., 2005[2]	F/49	Cerebellum	No	<1	Non-GTR +RT	Yes	Dead	2	19
Valéry et al., 2004[66]	F/74	Cerebellum	No	5	GTR+RT	No	Alive	12	12
Rajesh et al., 2003[58]	M/30	Lateral ventricle	No	NA	Non-GTR	-	Dead*	-	-
Kachhara et al., 2003[38]	F/62	Cerebellum	No	NA	GTR	NA	NA	NA	NA
Jenkinson et al., 2003[37]	F/51	Cerebellum	No	3	Non-GTR +RT	Yes	Alive	12	15
Fung et al., 2003[25]	F/56	Cerebellum	No	NA	Non-GTR	No	Alive	6	6

Non-GTR: non-gross total resection, *: patients died of postoperative complications, including intraventricular bleeding, intracranial hemorrhage and pneumonia and sepsis.

Authors and year	Sex/age (years)	Location	Multifocality	KI-67 (%)	Treatment	Recurrence	Outcome	PFS (months)	FU (months)
Elshihabi et al., 2003[24]	F/45	Cerebellum	No	<2.5	Non-GTR +RT+CT	No	Alive	36	36
Akhaddar et al., 2003[3]	F/43	Cerebellum	No	NA	GTR	No	Alive	16	16
Montagna et al., 2002[46]	F/53	Cerebellum	No	NA	Non-GTR +RT	NA	NA	NA	NA
Cacciola et al., 2002[13]	M/61	Cerebellum	No	NA	GTR	No	Alive	12	12
Taddei et al., 2001[63]	M/61	Cerebellum	No	NA	GTR	No	Alive	6	6
Jackson et al., 2001[36]	M/66	Cerebellum	No	3	GTR+RT	NA	NA	NA	NA
George et al., 2001[28]	F/59	Lateral ventricle	No	5.8	Non-GTR	Yes	Alive	67	67
Alkadhi et al., 2001[5]	M/38	Cerebellum	No	NA	GTR	No	Alive	15	15
	M/53	Cerebellum	No	NA	Non-GTR	Yes	Alive	108	180
Orlandi et al., 1997[48]	M/55	Cerebellum	No	NA	GTR	No	Alive	8	8
Soylemezoglu et al., 1996[62]	F/48	Cerebellum	No	<5	Non-GTR +RT	No	Alive	42	42
	M/53	CPA area	No	<5	GTR	Yes	Alive	132	132
	F/59	CPA area	No	<5	GTR+RT	No	Alive	60	60
Giangaspero et al., 1996[29]	M/37	Cerebellum	No	<1	GTR	Yes	Dead*	120	132
	M/36	Cerebellum	No	<1	GTR	Yes	Alive	120	180
	F/57	Cerebellum	No	<1	Non-GTR +RT	No	Alive	24	24
Ellison et al., 1993[23]	F/50	Cerebellum	No	<1	GTR	NA	NA	NA	NA
Davis et al., 1993[19]	F/49	Cerebellum	No	NA	Non-GTR	Yes	Alive	60	60
	M/53	Cerebellum	No	NA	Non-GTR+RT	NA	Dead*	-	6
Non-GTR: non-gross total resection, *: patients died of postoperative complications, including intraventricular bleeding, intracranial hemorrhage and pneumonia and sepsis.									

Table 3
Summary of clinical characteristics of cerebellar liponeurocytoma from literature and our institute

Variable	Reported cases (n=75)		Current series	Overall
	No. of Available cases	Value		
Mean age, years	75	46.2±13.6	43.9±14.5	46.0±13.6
Sex (M/F)	75	35/40	4/3	39/43
Supratentorial/Infratentorial	75	17/58	3/4	20/62
Multifocality	75	4	0	4
GTR	75	50 (66.7%)	7 (100%)	57 (76%)
Radiotherapy	75	16 (21.3%)	2 (28.6%)	18 (22.0%)
Chemotherapy	75	1 (1.3%)	0	1 (1.2%)
Mean PFS, months	53	40.0±40.7	44.9±44.4	40.6±40.8
Mean FU, months	54	47.5±51.5	44.9±44.4	47.2±50.4
Recurrence	57	17 (29.8%)	0	17 (26.6%)
Death	59	4 (6.8%)	0	4 (6.1%)
Values are presented as the number of patients (%), and mean values are presented as mean±SD.				

The mean follow-up in the pooled cohort (including 7 cases from our institute and 53 cases from the literature) was 47.2±50.4 months. PFS rates at 1, 5, 10 years were 92.7%, 78.0%, 23.8% respectively. The univariate Cox regression analysis showed that GTR (p=0.015) had significantly prolonged PFS than non-GTR. Other risk factors, including age (p=0.800), sex (p=0.325), tumor location (p=0.782) and radiotherapy (p=0.708), were not significant. Then we added sex, GTR and radiotherapy to the multivariate cox analysis. GTR (p=0.030) was the only risk factor to predict a better PFS (Table 4). Kaplan-Meier analysis showed that compared to non-GTR (median PFS: 67 months), GTR (median PFS: 120 months) was significantly associated with better PFS (p=0.0084) and radiotherapy (p=0.7068) could not predict a better PFS (Figure 3A, B). For non-GTR group, radiotherapy was not associated with better PFS (p=0.9197) (Figure 3C).

Table 4
Cox regression model for risk factors predicting PFS

Variable	Number of patients	Recurrence (%)	Univariate Analysis		Multivariate Analysis	
			HR (95% CI)	P value	HR (95% CI)	P value
Overall	60	17 (28.3)				
Age						
Age ≤ 45	27	7 (25.9)	Reference			
Age >45	33	10 (30.3)	1.140 (0.412-3.155)	0.800		
Sex						
Female	33	10 (30.3)	Reference		Reference	
Male	27	7 (25.9)	0.600 (0.217-1.660)	0.325	1.007 (0.327-3.106)	0.990
Location						
Supratentorial	12	2 (16.7)	Reference			
Infratentorial	48	15 (31.3)	1.241 (0.268-5.744)	0.782		
GTR						
Yes	40	7 (17.5)	Reference		Reference	
No	20	10 (50)	0.285 (0.103-0.787)	0.015*	0.286 (0.092-0.887)	0.030*
Radiotherapy						
Yes	15	2 (13.3)	Reference		Reference	
No	45	15 (33.3)	1.364 (0.268-6.935)	0.708	1.115 (0.215-5.788)	0.897
*: p<0.05 was considered statistical significant.						

Illustrative Cases

Case 3

A 64-year-old female presented with a 2-year history of dizziness and headache. Neurological examination revealed no neurological dysfunction. MR scans revealed a solid mass involving right side of cerebellum with heterogeneous signals on both T1WI and T2WI (Figure 1A and B). Contrast-enhanced MRI showed significant heterogeneous enhancement of the tumor (Figure 1C). A right suboccipital paramedian approach was performed. During surgery, the tumor was gray-red with soft texture and medium blood supply. There was a clear boundary between the surrounding brain tissue and the mass. Pathological examination revealed that some fat foci are dispersed within atypical tumor cells (Figure 1G). Immunohistochemistry revealed the tumor cells were positive for Syn, NeuN, MAP2, GFAP, ATRX and negative for olig-2 and IDH1. The Ki-67 index was approximately 7.5%. A cerebellar liponeurocytoma was confirmed by pathologists. Postoperative MRI showed the tumor was completely resected (Figure 1D-F). The postoperative course was uneventful and the patient was discharged 8 days after surgery. She was uneventful for the following 57.3 months until the last telephone interview.

Case 4

A 30-year-old woman experienced a surgical resection of a mass located at lateral ventricle at other hospital 1 year ago. The tumor was not resected totally and she came to our hospital for further treatment. Neurological examination revealed weakness of left limb. MR scans revealed a cystic-solid mass involving lateral ventricle with heterogeneous signals of both T1WI and T2WI (Figure 2A, B). Contrast-enhanced MRI showed no enhancement (Figure 2C). A right frontal approach was performed and boundary between the tumor and the surrounding brain tissue was not clear. Ultimately, the patient achieved gross total resection. Pathological examination revealed a cerebellar liponeurocytoma (Figure 2G). Postoperative MRI showed no residue of the tumor (Figure 2D-F). After 29.6 months, no evidence of tumor recurrence was observed.

Discussion

Cerebellar liponeurocytoma (CLPN) is a rare tumor that is characterized by well-differentiated histological features and a favorable clinical prognosis[1, 41]. Due to the rarity, most papers focused on describing the clinical features of CLPN, but few papers discussed treatment protocol for the rare lesion. Here, we reported the largest cohort of 7 cases from our institute and reviewed 75 cases from the literature, and try to verify adverse factors for PFS.

As its name implies, cerebellar liponeurocytoma is mainly located at the cerebellum and it is uncommon for the lesion to originate within the lateral ventricle[4]. Of the pooled analysis, 24% (20/82) cases of CLPN involved lateral ventricle, this entity should be considered as solely liponeurocytoma. There is no significant gender predominance for CLPN in this study, however, for supratentorial liponeurocytoma, the male-to-female ratio was 3:1, with an obvious male predilection, for infratentorial liponeurocytoma, the male-to-female ratio was 2.4:3.8, with a female predilection. The brain tumor often affected adults and occasionally involved children, only 2 patients below 20 years old were reported previously[14, 47]. The tumors were usually solitary, but multifocal lesions were also reported[34, 54, 60, 61].

Radiologically, Gembruch et al analyzed the features of MRI and found that hypointense (55%) and isointense (27.5%) on T1-weighted images and hyperintense (88.25%) on T2-weighted images[27]. Most cases can be observed a heterogeneous contrast enhancement after administration of contrast agent[27]. The fat in CLPN can be observed on CT as a hypodense lesion. MRI imaging usually shows a hyperintense signal on T1WI and the signal often get inverted in fat-suppressed images[21]. According to previous studies, 11 C-methionine in positron emission tomography (Met-PET) showed that Met uptake values were relatively high (T_{max}/N_{ave} : 3.2, 2.7), which may help establish a preoperative diagnosis of CLPN[34, 64].

Histologically, CLPN was composed of monotonous small round tumor cells and lipomatous cells[68]. Mitosis is rare or absent. The Ki-67 or MIB-1 index in the majority of reported cases was low[49, 64, 67], which was consistent with our findings. Kakkar et al pointed out the lipomatous cells were caused by lipidization of the neurocytic cells other than true adipocytic differentiation[39]. Although most CLPNs reveal benign histological features, several cases with atypical features have been described[28, 50]. In the study, the Ki-67 index was relatively high for 5 cases ($\geq 10\%$) and no case experienced a relapse. Due to the rarity of the disease, genomic profiles of CLPNs remained uncertain. Horstmann performed DNA sequencing for 20 CLPNs and found that TP53 missense mutations were the most common genetic alterations with a 20% frequency[35]. Anghileri et al pointed that FABP4 maybe a candidate marker for CLPNs, which is significantly overexpressed in the tumor[6].

The clinical prognosis of CLPN is generally satisfactory. Among the available 66 cases (including 59 cases from the literature and 7 cases from our institute), only 4 patients died and the mortality rate was 6.1%. 3 of them died of postoperative intracranial hemorrhage or pneumonia and only one patient may die from a tumor-related event. However, recurrence of CLPN is relatively high, available 64 cases (including 57 cases from the literature and 7 cases from our institute), 17 cases (26.6%) suffered tumor recurrence. In previous studies, optimum treatment modality was considered to be totally surgical resection[21]. We performed a multivariate Cox analysis and the results revealed that GTR had significantly prolonged PFS ($p=0.030$). Kaplan-Meier analysis showed that compared to non-GTR, GTR has a 53-month longer PFS. It is noting that 9 patients suffered relatively early tumor recurrence (PFS within 67 months) and 8 (88.9%) of them accepted non-GTR previously. 8 patients suffered latterly recurrence (PFS beyond 8 years) and only 2 (25%) cases accepted non-GTR. We speculate that GTR could prevent the early recurrence of CLPN but had limited benefit on its long-term recurrence. Some studies believe that postoperative radiotherapy decrease the tumor recurrence[21, 27], but of our pooled analysis, there is no statistical difference of tumor recurrence with or without radiotherapy. Kaplan-Meier curve also showed that Radiotherapy did not significantly prolonged PFS. For the non-GTR group, radiotherapy still did not show a better PFS. Based on these facts, we recommend GTR as CLPN best treatment and close follow-up should be performed,

although GTR was achieved. However, when CLPN showed atypical histological features, radiotherapy should be advised. In recent articles, Wolf et al and Pikis et al proposed familial predisposition of the tumor with possibly an autosomal dominant mode of inheritance[55, 67]. Future studies with larger cohorts are needed to verify the findings.

Conclusion

Cerebellar liponeurocytomas (CLPNs) are very rare brain tumors. It usually occurred in adults with no gender predominance. Although they have favorable clinical prognosis, the recurrence is relatively high. GTR should be the first choice and close follow-up is necessary. Postoperative radiotherapy could not improve PFS in this study. A larger cohort is needed to verify our findings.

Declarations

Funding

This study was funded by Multicenter clinical big data study and multi-path tumorigenesis mechanisms and precision treatment research on brainstem glioma (JINGYIYAN2018-7) and Special Fund of the Pediatric Medical Coordinated Development Center of Beijing Hospitals Authority (XTYB201822).

Conflicts of interest

All authors declared that there were no conflicts of interest to be disclosed.

Availability of data and material

The datasets generated during the current study are available from the corresponding author on reasonable request.

Code availability

Not applicable

Ethics approval

This study was approved by the human research ethics committee of Beijing Tiantan Hospital. The written informed consent was obtained from the subjects.

Consent to participate

All patients signed an informed consent form

Consent for publication

Written informed consent for publication was obtained from all participants.

Authors' contributions

Writing, original draft, Conceptualization: Pengcheng Zuo

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Literature review: Zhuang Jiang, Changcun Pan, Cheng Xu

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Figures

Figure 1

Magnetic resonance imaging (MRI) shows a heterogeneous T1WI (A) and heterogeneous T2WI (B) solid mass with significant enhancement (C) located at right cerebellum. Postoperative MRI revealed the tumor was resected totally (D-F). G: Histopathological examination revealed the typical morphology of cerebellar liponeurocytoma. Some fat foci are dispersed within atypical tumor cells (H&E, ×200).

Figure 2

Magnetic resonance imaging (MRI) shows a heterogeneous T1WI (A) and heterogeneous T2WI (B) cystic-solid mass with no enhancement (C) located at lateral ventricle. Postoperative MRI revealed the tumor was resected totally (D-F). G: H&E stained section showed monotonous tumor cells with round

nuclei and clear cytoplasm with sparse lipomatous cells (H&E, ×200).

Figure 3

Kaplan-Meier survival curves showing PFS of the pooled cohort divided into GTR and Non-GTR groups (A) and RT and No RT groups (B). Survival curve analysis displaying PFS in non-GTR group between radiotherapy and no radiotherapy (C).