Pediatric Glioblastoma: A Common CNS Tumor in an Uncommon Age

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Background: *Pediatric glioblastoma still has dismal prognosis despite the treatment has been continuously developed in the last three decades.*
Objective: *To review the medical literatures regarding pediatric glioblastoma and demonstrate an illustrative case.*
Material and Method: *The author reviewed medical literatures involving pediatric glioblastoma. Inclusion criteria are the literatures published from 1993 to the present with case numbers of 15 or more and proper statistical analysis.*
Results: *Ten studies met the mentioned criteria. The overall survival of patients with pediatric glioblastoma ranges from 11 to 43 months. The prognostic factors that have statistically strongest correlation with prolong survival include gross total resection and Eastern Cooperative Oncology Group (ECOG) Performance Status Score of 0 to 1 before radiation therapy. Other statistically significant favorable prognostic factors were superficial tumor location, radiation dose higher than 50 Gray, preoperative Karnofsky Performance Status (KPS) Score ≥80 and Neurologic Function Score (NFS) of 0 to 1.*
Conclusion: *Prognosis of pediatric glioblastoma remains poor with limited overall survival rate. Current recommendation of the treatment is attempt of gross total resection followed by radiation therapy and chemotherapy.*

Keywords: *Pediatric glioblastoma, Treatment, Survival outcome, Prognostic factor*

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Glioblastoma is the most malignant form of primary central nervous system (CNS) tumor with dismal survival rates. It accounts for 14.9% of all primary CNS tumors, but the incidence is relatively rare in pediatric population. Glioblastoma comprises of approximately 2.9% of all primary CNS tumors in children and adolescence[1]. Survival period of pediatric glioblastoma was found to be longer than adult glioblastoma, median overall survival of pediatric glioblastoma ranges from 11 to 43 months, whereas that of adult glioblastoma receiving standard therapy is only 14.6 months[2-12]. The reason for this difference is possibly caused by the distinction in molecular genetics between both age groups[13]. Due to rarity of the disease, knowledge about pediatric glioblastoma in terms of clinical characteristic, natural history and standard treatment are not well established. Currently, there has been no standard treatment protocol for pediatric glioblastoma outside the brainstem. The mainstay of treatment is surgery followed by radiation therapy (RT). There is no standard consensus guideline for chemotherapeutic regimens, unlike in adult glioblastoma. Clinical trials for potential drugs and targeted therapies are still in ongoing process.

This article is a review of pediatric glioblastoma in regards to demographic data, current recommendation of treatment, outcome, and ongoing studies. Relevant studies of pediatric glioblastoma were presented. The author also revealed an illustrative case with this kind of tumor.

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Material and Method

The medical literatures involving pediatric glioblastoma, published from 1993 to the present with case numbers of 15 or more and proper statistical analysis, were included. Ten relevant studies which met the criteria were reviewed and are presented in Table 1.

Results

Demographic data

Age

Mean age of pediatric glioblastoma is ranged

Table 1. The literature review in pediatric glioblastoma

Author	n	Mean age in year (range)	M:F	Including brainstem	Location	Treatment		Median PFS (month)	Overall survival (month)	Good prognostic factor or comment
						Surgical	Adjuvant			
Artico et al, 1993[3]	27	13 (2-18)	1.4:1	N	Supratentorial 100%	GTR 25.9% STR 74.1%	RT 88.9% CMT 14.8%	N/A	1-year 74% 2-year 37%	
Sanchez-Herrera et al, 2009[4]	16	8.8 (3-15)	1.7:1	N/A	Supratentorial 81.2% Infratentorial 18.8%	GTR 31.3% STR 68.8%	RT CMT	N/A	Mean 54.97	More frequent complications in younger age
Song et al, 2010[5]	27	Median 9 (0-16)	1.1:1	N	Superficial 77.8% Deep 22.2%	GTR 44% STR 44% Biopsy 11%	RT 89% CMT 52% (TMZ 8.5%)	12	Median 43 1-year 67% 2-year 52% 5-year 40%	Superficial location (univariate), GTR (univariate, multivariate)
Perkins et al, 2011[6]	24	Median 11 (3-20)	0.8:1	Y	Supratentorial 79.2% Infratentorial 12.5% Spinal cord 8.3%	GTR 25% STR 54% Biopsy 21%	RT 100% CMT 58%	N/A	Median 13.5 1-year 57% 2-year 32%	GTR
Das et al, 2012[7]	65	13.3 (2-18)	2.6:1	N	Supratentorial 95.5% Infratentorial 4.5% Superficial 75% Deep 25%	GTR 66% STR 34%	RT 96.9% CMT 38.5% (TMZ 7.7%)	10	Median 20	Good performance status[a] and superficial location (univariate), GTR (univariate, multivariate)
Ansari et al, 2012[8]	23	15.2 (10-20)	1.55:1	Y	Supratentorial 100%	GTR 8.7% STR 39.1% Biopsy 52.2%	RT 100% CMT 43.5% (no TMZ)	9	Mean 16 Median 11 1-year 47.8% 2-year 17.4% 5-year 8.7%	RT dose >50 Gy (univariate), Good performance status[b] (univariate, multivariate)
Yang et al, 2013[9]	37	10.8 (1.5-17.3)	2.7:1	N	Superficial 70.3% Deep 29.7%	GTR 45.9% STR 35.1% Biopsy 18.9%	RT 94.6% CMT 91.9%	11.5	Median 18.7 1-year 63.9% 2-year 44.5% 5-year 17.6%	GTR
Mallick et al, 2015[10]	23	11.5 (7-9)	1.9:1	Y	Supratentorial 100%	GTR STR	RT 100% TMZ	N/A	Median 41.9 1-year 69.6% 2-year 60.9%	N/A
Nikitovic et al, 2016[11]	15	11.8 (4-17)	1.5:1	Y	Supratentorial 66.7% Infratentorial 33.3%	GTR 33.3% STR 66.7%	RT 100% CMT 73.3%	N/A	Median 13.5 Mean 28.7	GTR and good performance status[c]
Adams et al, 2016[12]	342	10 (<1-18)	1.4:1	Y, then excluded	Supratentorial 50.8% Infratentorial 22.3% Unspecified location and spinal cord 26.9%	GTR 35.4% STR 28.8% Biopsy 17.3% No surgery 17%	RT 71.6% CMT	N/A	Median 12 1-year 51.7% 2-year 28.3% 5-year 15.7%	GTR (univariate, multivariate and mPS) before and after exclusion of brainstem location

n = number of case; M = male; F = female; PFS = progression-free survival; GTR = gross total resection; STR = subtotal resection; RT = radiation therapy; CMT = chemotherapy; TMZ = temozolomide; univariate = univariate analysis; multivariate = multivariate analysis; mPS = multiple propensity score-adjusted models; performance status[a] = preoperative Karnofsky Performance Status (KPS) score $\geq$ 80; performance status[b] = Eastern Cooperative Oncology Group (ECOG) Score of 0 or 1 before radiation therapy; performance status[c] = Neurologic Function Score (NFS) of 0 or 1 before radiation therapy; Y = yes; N = No; N/A = no available data

from 8.8 to 15.2 years. Age range varies from age of less than one year to 20.

Sex

Most of the literatures reported male predominance with the male: female ranging from 1.1:1 to 2.7:1. Only a series of Perkins et al reported female predominant[6].

Tumor location

The major site is supratentorial location (50.8 to 100%). Three studies found the frontal lobe as the most common location[3,7,12], while one reported the occipital lobe[8] and another one reported the temporal lobe[10]. Infratentorial location, including the brainstem, is comprised of 0 to 33.3% of the tumors.

In terms of tumor proximity to the brain surface, 70.3 to 75% of the tumor is situated in the superficial location, while 25 to 27.9% involved the deep structures. Univariate analysis showed that superficial tumors were associated with longer survival[5,7]. However, this correlation was not found in multivariate analysis and multiple propensity score-adjusted models. Gross total resection yields longer survival consistently in observed data, univariate-, multivariate-, and multiple propensity score-adjusted models[12]. This indicates that better accessibility to superficial tumors actually plays a role in the better outcome.

Clinical presentations

The most frequent clinical presentation consists of clinical signs and symptoms related to intracranial hypertension, found in 78 to 100% of patients[3,4,7,11]. Other clinical manifestations include seizure (33.3 to 65%), neurological deficit (12.5 to 80%) and psychiatric disorder (22.2%).

Histopathological and molecular genetic features

In pediatric primary glioblastoma, their histomorphological features are found to be similar to those of adult glioblastoma. One particular histopathological subtype of glioblastoma, giant cell glioblastoma or gigantocellular glioblastoma is a rare form of glioblastoma in adult and much rarer in pediatric group. It was previously believed that giant cell glioblastoma had longer survival, but Perkins et al and Karremann M et al found no difference to the entire population of pediatric glioblastoma in terms of survival outcome[6,14].

For molecular genetic aspect, pediatric primary glioblastoma has some different features from adult primary glioblastoma. In pediatric glioblastoma, PTEN deletion and EGFR amplification are rare, whereas p53 alterations are found more frequently than in adult glioblastomas[15]. Glioblastomas with O^6-methylguanine-DNA methyltransferase (MGMT) promoter methylation correlates with better survival outcomes and better response to temozolamide[13]. More understanding about these molecular genetic features is helpful in development of individualized target therapies.

Current treatment recommendations

All literatures showed significant longer survival in patients undergoing gross total resection over subtotal resection or biopsy[12]. There was no statistical difference in survival outcome between subtotal resection and biopsy groups. The largest series, and possibly the strongest study on the extent of resection, was reported by Adams et al[12]. Retrieving the data from the national cancer registry in the United States with the sample size of 342 patients, they found that the extent of resection was a predictor of survival in observed data, univariate-, multivariate-, and multiple propensity score-adjusted models. This study is the strongest study so far because of large sample size and statistical analyses used. Even though, it is a retrospective observational study, a study design as a randomized control trial has never been done and is unlikely to be conducted in the future due to ethical issues. This study also analyzed a subgroup by excluding brainstem glioblastoma to prove a belief that brainstem glioblastoma has more dismal prognosis than that of hemispheric origin. All the results of this subgroup were consistent with the entire sample size. Even with the evidence that gross total resection yields best prognosis, due to location of the tumor, not all tumors can be totally surgically removed. There were studies showing the efficacy of using intraoperative ultrasound and MRI to maximize the extent of resection and also identify the residual tumors[16,17].

Other methods valuable for increasing extent of resection includes intraoperative neuronavigation device, diffusion tensor imaging tractography, cortical mapping and intraoperative neuromonitoring[12].

RT is the standard treatment for glioblastoma. There has been no consensus guideline for treatment of primary and recurrent tumors. Types of radiation (whole brain, two-dimensional and three-dimensional RT) do not have effect on the overall survival[6,11]. In terms of dose, Ansari et al found that longer survival

was associated with radiation dose higher than 50 Gray[8].

Likewise, there has been no standard regimen of chemotherapy for pediatric glioblastoma. In adult, temozolamide has been identified to prolong survival for a couple of months[2]. There was no study specific to the benefit(s) of temozolomide treatment in pediatric glioblastoma; but the study in pediatric high-grade glioma, which glioblastoma was the majority of the samples, showed that temozolomide did not improve outcome[9,18].

Treatment outcomes

Previous publications on pediatric glioblastoma since 1993 were reviewed. Table 1 shows details of the individual study with sample sizes more than 15 patients. Median survival time ranged from 11 to 43 months. The survival time has been not increased over decades suggesting that treatment modalities have been not much improved. Comparing to adult glioblastoma, of which overall survival time with standard treatment is 14.6 months[2], pediatric glioblastoma has better prognosis than adult glioblastoma. Adams et al showed no statistical difference in mean survival time between pediatric glioblastoma before and after excluding glioblastoma of the brainstem[12].

In the literature review, overall survival rate at 1 year was 47.8 to 74%, and at 2 years was 17.4 to 60.9%. The wide range in these overall survival rates between the different studies may be accounted for by the small sample size of each literature. Moreover, each study had a long observation period, usually in the range of 10 years; to get these sample size amounts, the treatment modalities had changed by time and were not uniform because consensus guidelines for the treatment had never been developed.

Prognostic factors of survival outcome were identified in many literatures. Univariate analysis showed that superficial location, gross total resection, good performance status (pre-operative Karnofsky performance status score $\geq$80) and radiation dose more than 50 Gray were associated with longer survival. However, in multivariate analysis, only gross total resection and good performance status (Eastern Cooperative Oncology Group score of 0 to 1 before RT) were associated with longer survival. Multiple propensity score-adjusted models confirmed the association between gross total resection and longer survival.

Regarding treatment complications, a study revealed that younger patients had complications more frequently than older groups[4].

Ongoing researches

There are potential drugs in the process of investigation for outcome benefit in glioblastoma. Bevacizumab, a monoclonal antibody targeting vascular endothelial growth factor (VEGF), has been approved to use in patients with recurrent glioblastoma. There was a report of bevacizumab used in pediatric glioblastoma as a primary treatment in 2013[19]; however, there was no statistical analysis owing to the small number of cases. To date, no literature reports its use for treating recurrent pediatric glioblastoma.

Nivolumab, an immune checkpoint inhibitor (ICI), is in phase III clinical trial for glioblastoma treatment. There was one case report of nivolumab used in pediatric glioblastoma patient which it resulted in severe cerebral edema[20].

Recurrent MET fusion genes were found in approximately 10% of pediatric glioblastoma. These genes have been identified as a potential target for the treatment[21-23]. A pediatric glioblastoma patient who had a MET-fusion-expressing tumor was treated with crizotinib, a targeted inhibitor. It resulted in tumor shrinkage, but subsequent appearance of new treatment-resistant lesions was seen[23]. Lastly, a radiosensitizer has been developed to enhance the efficacy of RT, but the study was still the in vitro phase[24].

Illustrative case

A 6-year-old girl presented with headache, vomiting and right ataxia for three months. Physical examination showed no motor weakness. Hypertonia and hyperreflexia were found on the left arm and leg with positive Babinsky sign on the left. Contrast-enhanced cranial MRI showed mixed solid and cystic mass measured 5.5x4.7x5.4 cm in at the right thalamus compressing the body and trigone of the right lateral ventricle (Fig. 1). She underwent ventriculostomy and craniotomy with tumor removal with intraoperative neurophysiologic monitoring. Postoperatively, she had normal physical examination. Immediate postoperative cranial MRI showed suspected residual tumors at the medial aspect of posterior horn of the right lateral ventricle, 2.1x0.9 cm in size, and at the splenium of corpus collosum, 1x0.9 cm in size. Final pathologic diagnosis was glioblastoma. She underwent volumetric arc therapy (VMAT) of 60 Gray. Due to financial status, temozolomide was not administered. She has currently

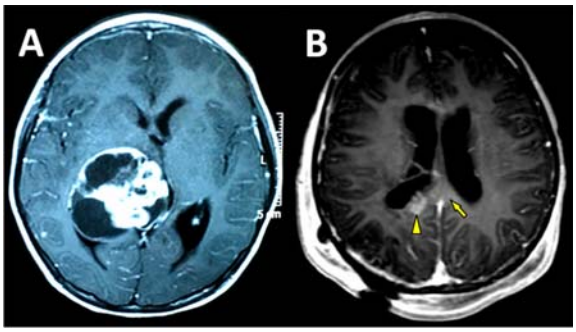

Fig. 1 (A) Contrast-enhanced cranial MRI showing (A) a solid cystic tumor at the right thalamus; (B) Postoperative MRI showing lesions suspicious of residual tumors at the medial aspect of posterior horn of the right lateral ventricle (arrowhead) and the splenium of corpus collosum (arrow).

been followed-up for 1 year postoperatively without evidence of tumor progression.

Discussion

Glioblastoma is one of the most common primary brain tumors; however, it is rarely encountered in the pediatric age group. A limited number of case series were reported in medical literature. In a review of literature, ten studies with at least 15 patients and appropriate statistical analysis were found. Most series showed male predominance[3,4,7-12]. The tumor is found in a wide variety of age ranging from neonatal period to teenage[5,12]. In terms of tumor location, pediatric primary brain tumors commonly occur in the posterior cranial fossa, but pediatric glioblastoma reported in almost all series mostly originated in the supratentorial location and infratentorial glioblastoma in children was relatively rare[3,4,6,7,8,10-12].

Survival time collected from all case series ranges from 11 to 43 months[3-12]. The illustrative case had a survival time longer than 12 months which is in this common range of survival period. Median progression free survival (PFS) was ranged from 9 to 12 months[5,7-9]. The illustrative case also had PFS at least 12 months. A longer survival was found to be correlated with gross total resection[5-7,9,11-12], good performance status before surgery and before RT[7-8,11], superficial tumor location[5] and receiving radiation dose higher than 50 Gray[8]. The illustrative patient had three of these favorable factors: good performance status before surgery and before RT, and receiving radiation dose higher than 50 Gray. Because her glioblastoma was located in a deep structure, thalamus, maximal safe resection was performed to avoid operative morbidity caused by injury of the vital neural structures. In well-equipped operating theater, intraoperative MRI is helpful to delineate maximal safe resection margin. Although gross total resection could not be achieved in this case, she received postoperative RT with total dose of 60 Gray proved to have a positive effect on survival outcome in a case series[8].

There is still no standard regimen of chemotherapy for pediatric glioblastoma. Temozolomide is an effective chemotherapeutic agent used as the standard treatment of newly diagnosed glioblastoma; however, this drug is not covered by the universal health coverage in Thailand. The illustrative case did not receive temozolomide because she used the universal health coverage for the treatment and her family could not afford the high cost of this kind of chemotherapy. Temozolomide should be in national list of essential medicines to reduce this inequity in accessibility of the standard treatment.

Some prognostic factors can be modified to be favorable factors. Doctors and other healthcare workers have a major role in providing knowledge about the disease to parents. Parents should be encouraged to bring patients to hospitals early when they have minimal symptoms and signs compatible with brain tumor or intracranial mass lesion. There is a higher chance that patients with early diagnosis would be in a good performance status before operative treatment, tumor size is still small, and the vast area of the brain is not involved by tumor. They have a better opportunity to achieve gross total resection, to have good performance status before RT, and to complete a proper radiation dose. In surgical perspective, gross total resection or better strategy so called "supratotal resection" must be done first in the treatment of glioblastoma. If gross total or supratotal resection carries a significant risk of neurologic complications, maximal safe resection should be considered. I thus encourage primary doctors and surgeons to do as much as they can to help patients meet these favorable factors to improve survival outcome in patients with pediatric glioblastoma.

Conclusion

Pediatric glioblastoma is one of the most aggressive pediatric primary brain tumors with very dismal survival outcome. Through the past three decades, the treatment guidelines remain no consensus as well as newly adapted regimens do not help much in survival improvement. Longer survival outcome is

strongly associated with gross total resection and good performance status before RT. The current recommendation includes gross total resection followed by RT, concurrent and adjuvant chemotherapy. Temozolomide does not show significant benefit in pediatric glioblastoma. In Thailand, with limited resources, what we can do is to attempt gross total resection followed in a timely manner by RT and chemotherapy administration in eligible patients. Development of new drugs and targeted therapies must be encouraged to improve survival outcome in pediatric glioblastoma.

What is already known from this topic?

Pediatric glioblastoma is a rare tumor in pediatric age group. The prognosis of pediatric glioblastoma is poor.

What this study adds?

In the literature review, mean age is 8.8 to 15.2 years with fully range from neonatal period to teenage. Male predominant is reported consistently.

Favorable prognosis is related to gross total resection, superficial location of the tumor, good performance status before surgery and before RT, and RT dose higher than 50 Gray.

Potential conflict of interest

None.

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มะเร็งไกลโอบลาสโตมาในเด็ก: เนื้องอกของระบบประสาทส่วนกลางที่พบบ่อยในอายุที่พบไม่บ่อย

อินธิรา ขัมภลิขิต

ภูมิหลัง: *มะเร็งไกลโอบลาสโตมาในเด็กยังคงมีการพยากรณ์โรคที่ไม่ดีแม้ว่าจะมีการพัฒนาการรักษาอย่างต่อเนื่องในช่วงสามทศวรรษที่ผ่านมา*

วัตถุประสงค์: *เพื่อทบทวนวรรณกรรมทางการแพทย์ที่เกี่ยวข้องกับมะเร็งไกลโอบลาสโตมาในเด็กและนำเสนอผู้ป่วยตัวอย่าง*

วัสดุและวิธีการ: *ผู้นิพนธ์ทบทวนวรรณกรรมทางการแพทย์ที่เกี่ยวข้องกับมะเร็งไกลโอบลาสโตมาในเด็ก โดยเกณฑ์การเลือกคือเป็นวรรณกรรมที่ได้รับการตีพิมพ์ตั้งแต่ปี พ.ศ. 2536 จนถึงปัจจุบันซึ่งมีจำนวนผู้ป่วยตั้งแต่ 15 รายขึ้นไปและมีการวิเคราะห์ทางสถิติที่เหมาะสม*

ผลการศึกษา: *มี 10 งานวิจัยที่เข้าได้กับเกณฑ์การเลือกข้างต้น ระยะเวลาการรอดชีวิตโดยรวมของผู้ป่วยมะเร็งไกลโอบลาสโตมาในเด็กมีระยะเวลาตั้งแต่ 11 เดือนจนถึง 43 เดือน ปัจจัยที่เกี่ยวข้องกับระยะเวลาการรอดชีวิตที่ยืนยาวซึ่งมีนัยสำคัญทางสถิติ ได้แก่ การผ่าตัดเนื้องอกออกได้หมดและมีคะแนน Eastern Cooperative Oncology Group (ECOG) Performance Status เท่ากับ 0 ถึง 1 ก่อนได้รับรังสีรักษา ปัจจัยการพยากรณ์โรคที่ดีอื่น ๆ ได้แก่ เนื้องอกอยู่ในตำแหน่งตื้น ได้รับปริมาณรังสีรักษามากกว่า 50 เกรย์ คะแนน Karnofsky Performance Status มากกว่า 80 และคะแนน Neurologic Function เท่ากับ 0 ถึง 1*

สรุป: *การพยากรณ์โรคของมะเร็งไกลโอบลาสโตมาในเด็กยังไม่ดี มีอัตราการรอดชีวิตที่จำกัด คำแนะนำสำหรับการรักษาในปัจจุบันคือพยายามผ่าตัดเนื้องอกออกให้หมดตามด้วยรังสีรักษาและเคมีบำบัด*