Risk Factors for Postoperative Hydrocephalus Following Subependymal Giant Cell Astrocytoma Resection: A Study of Under 18-Year-Old Patients in China

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Research Article

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Abstract

Background: Hydrocephalus may occur after subependymal giant cell astrocytoma (SEGA) resection. In existing literatures, SEGA almost always occurred in patients with tuberous sclerosis complex (TSC), however, many SEGA also occurred alone in our Chinese pediatric patients.

Objective: To discuss the risk factors of postoperative hydrocephalus following SEGA resection and the relationship between SEGA and TSC in Chinese children.

Materials and methods: A total of 35 children (≤18-year-old) who underwent SEGA resection were selected. From 3 months postoperatively until December 2020 all patients received telephone or clinical follow-up. Related risk factors were first screened by univariate analysis and then analyzed by multivariate logistic regression.

Results: The ratio of males to females was 3:2 and the mean age was 11.6 years. Twenty cases were associated with TSC and 15 were not. The mean maximum diameter of the SEGA for patients with and without associated TSC was 49.7mm and 30.5mm, respectively (Z=-3.293, P=0.001). Twenty-eight patients had preoperative hydrocephalus. Sixteen patients developed postoperative hydrocephalus, and amongst these, 2 did not have hydrocephalus before surgery. Multivariate analysis showed that association with TSC [odds ratio (OR), 18.81, P=0.048] and tumor resection rate (OR, 0.042, P=0.025) were independent risk factors for postoperative hydrocephalus.

Conclusion: SEGA could be associated with TSC or appear alone. The maximum diameter of SEGA associated with TSC is larger than that without TSC. Hydrocephalus is a common onset symptom and might recur following SEGA resection. Association with TSC and tumor resection rate are risk factors for postoperative hydrocephalus.

Introduction

Subependymal giant cell astrocytoma (SEGA), a tumor arising in the wall of the lateral ventricles adjacent to the foramen of Monro, accounts for about 1–2% of all pediatric brain tumors.[1, 2] It typically occurs during the first two decades of life and infrequently arises after the age of 20–25 years.[3, 4] The International Agency for Research on Cancer of World Health Organization Classification of Tumors (revised 4th edition) classified SEGA as grade I tumor in 2016.[5] SEGA is composed of different cell lineages and is not purely astrocytic in nature.[6] SEGA has been considered to be a pathognomonic finding of tuberous sclerosis complex (TSC), and its incidence in patients with TSC varies from 5–25%.[4, 7, 8] TSC is a neurocutaneous syndrome which mainly involves the central nervous system (CNS) where SEGA, subependymal nodules (SEN), and cortical tubers may be present. In 2013, the experts of an international committee defined the imaging features of SEGA as a lesion at the caudothalamic groove larger than 1 cm in any direction or subependymal lesions which showed serial growth on consecutive
imaging in any position. Because of their location and growth potential, SEGA can cause increased intracranial pressure, obstructive hydrocephalus, focal neurologic deficits, even death.

Symptomatic lesions tend to have greater morbidity, in which cases surgical resection is the preferred modality of treatment. However, surgery can have associated postoperative complications, such as intracranial infection, seizures and hydrocephalus. Many patients who develop hydrocephalus after surgery require a second operation to relieve symptoms. The previous studies reported that SEGA occurs almost exclusively in patients with TSC, however in our study cohort, nearly one-third of Chinese pediatric patients did not have associated TSC. The purpose of this study was to investigate the risk factors for hydrocephalus after SEGA resection and the relationship between SEGA and TSC in Chinese children younger than 18 years old.

Materials And Methods

Patients

We included 35 children (≤ 18 years) who underwent SEGA resection at our institutions from January 2002 to December 2020. All patients with TSC received confirmation through genetic testing. Clinical data, including age, sex, onset symptoms, radiological results, operation findings, surgical approach, postoperative complications and related treatments, were collected from the hospital database. In order to evaluate the extent of the resection and for future comparisons, all patients underwent a cranial magnetic resonance imaging (MRI) enhanced scan 24–48 hours postoperatively. From 3 months postoperatively until December 2020 all patients received follow-up, the evaluation was based on neuroimaging judgement and clinical manifestations.

Potential risk factors for hydrocephalus after resection

The potential risk factors for hydrocephalus after resection were statistically analyzed including age (< 12 or ≥ 12 years), sex (male, female), TSC (positive, negative), preoperative hydrocephalus (positive, negative), tumor location (near the foramen of Monro, far from the foramen of Monro), tumor size (< 30 or ≥ 30mm), the extent of resection [gross total resection (GTR), subtotal resection (STR)] and postoperative intracranial infection (positive, negative).

Statistical methods

Statistical analysis was performed with SPSS 25.0. Continuous variables were analyzed using the non-parametric Mann-Whitney U-test, and categorical variables were analyzed using chi-square test. Univariate analyses were performed to examine the relationship between hydrocephalus postoperatively and prognostic factors. Variables were included in a logistic regression model if their $P$ value in the univariate analysis was < .1. The odds ratio (OR) and 95% confidence interval (CI) were calculated. $P< .05$ was considered statistically significant.
Results

In total, 35 patients (21 males, 14 females) were eligible for the final analysis in the present study. The patients’ age ranged from 5 to 18 years (mean 11.6 years), the disease duration ranged from 2 weeks to 168 months (mean 27.6 months), and the follow-up period ranged from 3 to 168 months (mean 78.2 months). A total of 20 patients had associated TSC and 15 patients had no associated TSC. All data regarding age, sex, disease duration, onset symptoms, tumor location, tumor size and tumor components are presented in Table 1. Sixteen patients developed hydrocephalus after SEGA resection during the follow-up period. In order to relieve the hydrocephalus of these patients, ventriculoperitoneal shunt (VPS) was performed in 7 cases, Ommaya reservoir insertion was performed in 6 cases, ventricularostomy was performed in 2 cases, and Ommaya reservoir and ventricularostomy insertion was performed in one case. All data regarding age, sex, association with TSC, preoperative hydrocephalus, tumor location, tumor size, the extent of resection and postoperative complications are presented in Table 2. The preoperative, postoperative and follow-up images and pathological imaging of a 10-year-old SEGA patient are shown in Fig. 1a-d. Skin abnormalitis of a 12-year-old SEGA patient with TSC are shown in Fig. 1e-f.

There were no significant differences in age, sex, disease duration, onset symptoms, preoperative hydrocephalus, tumor location or tumor components between patients with and without associated TSC (Table 1). There were significant differences in tumor size (patients with associated TSC: 49.7mm vs. patients without associated TSC: 30.5mm, $P = 0.001$). Furthermore, the maximum diameter of SEGA was larger in patients with associated TSC than without associated TSC. There were no significant differences in age, sex, preoperative hydrocephalus or postoperative intracranial infection between patients with and without hydrocephalus after SEGA resection. The presence or absence of associated TSC, tumor size, tumor location and the extent of resection were included in a logistic regression model (Table 2). Logistic regression analysis showed that GTR (OR, 0.042; 95% CI, 0.003–0.670; $P = 0.025$) was a protective factor for postoperative hydrocephalus after SEGA resection, while association with TSC was a risk factor (OR, 18.814; 95% CI, 1.020-347.120; $P = 0.048$) (Table 3).

Discussion

SEGA is a clinically infrequent benign brain tumor and the incidence is slightly lower in females than in males which is consistent with our research.[13] The mean age at diagnosis was 11.6 years in our study. In existing reports,[1, 14–17] SEGA was always associate with TSC, a systemic autosomal-dominant disease, characterized by Vogt’s clinical triad of mental retardation, seizures and facial angiofibroma.[18] However, in our cohort, 20 cases of SEGA had associated TSC, and 15 cases of SEGA occurred alone. This difference from previous studies[1, 14–17] might be due to the different ethnic groups in China. We analyzed the age, sex, disease duration and other characteristics of patients with and without associated TSC and found that there were significant differences in the maximum tumor diameter between the two groups. The maximum diameter of SEGA with associated TSC was larger than that of patients without associated TSC.
SEGA tends to occur in the area of the lateral ventricle near the foramen of Monro, which easily causes hydrocephalus.[19] Associated morbidity and mortality are sometimes due to obstructive hydrocephalus rather than tumor growth.[20] Tumor resection is the main treatment method,[12] however, there could be recurrence or new discovery of hydrocephalus after resection in some patients, and severe hydrocephalus can be fatal.[6, 11, 21, 22] In our study of 35 patients, 28 had preoperative hydrocephalus, amongst these 14 had relief after resection while 14 had recurrence. Sixteen patients in our study had postoperative hydrocephalus, amongst which 2 cases were newly discovered. For the 16 patients with postoperative hydrocephalus, a second operation was carried out. All patients effectively had relief and no obvious abnormalities were found during the follow-up period.

We analyze the risk factors of postoperative hydrocephalus and found that association with TSC and extent of resection are independent risk factors. Patients with associated TSC are more likely to develop hydrocephalus after resection than patients without associated TSC. The reason might be that for patients with associated TSC, in addition to SEGA, there could have SEN or other neurostructural disorders. Compared with SEGA alone, patients with concurrent SEN or other disorders are more likely to have obstructive hydrocephalus. As for the extent of surgical resection, GTR is less prone to develop hydrocephalus after resection than STR. In general, total resection should be the goal of surgery, but for some tumors especially those with large volume and unclear boundaries with adjacent brain tissue, it may be difficult to resect the entire tumor by surgery, and there is a greater probability of hydrocephalus after surgery. For patients with these risk factors, VPS or Ommaya reservoir insertion could be considered during the first SEGA resection in order to avoid or reduce injury caused by a second operation. For patients with associated TSC, the mechanistic target of rapamycin (mTOR) inhibitors everolimus could be considered, which was approved effectively by the Food and Drug Administration in 2010.[23]

**Conclusion**

The incidence of SEGA is slightly more predominant in males than in females. The lesion tends to be located in the lateral ventricle near the foramen of Monro. SEGA could have associated TSC or occur alone. The maximum diameter of SEGA with associated TSC is larger than that without associated TSC. Hydrocephalus is a common onset symptom of SEGA, which can also occur after tumor resection. Some patients need further VPS or Ommaya reservoir insertion. Association with TSC and STR could place patients at high risk for hydrocephalus after SEGA resection. For these patients, VPS or Ommaya reservoir insertion could be considered in conjunction with SEGA resection.

**Declarations**

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**Conflicts of interest/Competing interests**
The authors have no conflicts of interest to declare that are relevant to the content of this article.

Ethics approval

This retrospective chart review study involving human participants was in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Ethics Committee of Xinhua Hospital Affiliated to Shanghai Jiaotong University School of Medicine approved this study (Approval No. XHEC-D-2020-052).

Consent to publish

The authors affirm that human research participants provided informed consent for publication of the images in Figures 1e-f.

Data availability

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

Author contributions

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by Fangjie Shen, Jia Wang, Huatao Niu and Xiaoqiang Wang. Fangjie Shen, Gang cui and Xu kang collected and analyzed the raw clinical data. The first draft of the manuscript was written by Fangjie Shen, Jia Wang and Loren Skudder-Hill, all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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References

1. Jozwiak S, Mandera M, Mlynarski W (2015) Natural History and Current Treatment Options for Subependymal Giant Cell Astrocytoma in Tuberous Sclerosis Complex. Semin Pediatr Neurol 22:274–281. doi:10.1016/j.spen.2015.10.003

2. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, Ohgaki H, Wiestler OD, Kleihues P, Ellison DW (2016) The 2016 World Health Organization Classification of
Tumors of the Central Nervous System: a summary. Acta Neuropathol 131:803–820. doi:10.1007/s00401-016-1545-1

3. Curatolo P, Bombardieri R, Jozwiak S (2008) Tuberous sclerosis. Lancet 372:657–668. doi:10.1016/S0140-6736(08)61279-9

4. Kothare SV, Singh K, Chalifoux JR, Staley BA, Weiner HL, Menzer K, Devinsky O (2014) Severity of manifestations in tuberous sclerosis complex in relation to genotype. Epilepsia 55:1025–1029. doi:10.1111/epi.12680

5. David N. Louis HO, Otmar D, Wiestler WK, Cavenee (eds) (2016) WHO Classification of Tumours of the Central Nervous System (Revised 4th edition). Lyon, IARC

6. Sharma MC, Ralte AM, Gaekwad S, Santosh V, Shankar SK, Sarkar C (2004) Subependymal giant cell astrocytoma—a clinicopathological study of 23 cases with special emphasis on histogenesis. Pathol Oncol Res 10:219–224. doi:10.1007/BF03033764

7. Adriaensen ME, Schaefer-Prokop CM, Stijnen T, Duyndam DA, Zonnenberg BA, Prokop M (2009) Prevalence of subependymal giant cell tumors in patients with tuberous sclerosis and a review of the literature. Eur J Neurol 16:691–696. doi:10.1111/j.1468-1331.2009.02567.x

8. Kingswood JC, d'Augeres GB, Belousova E, Ferreira JC, Carter T, Castellana R, Cottin V, Curatolo P, Dahlin M, de Vries PJ, Feucht M, Fladrowski C, Gislimberti G, Hertzberg C, Jozwiak S, Lawson JA, Macaya A, Nabbout R, O'Callaghan F, Benedik MP, Qin J, Marques R, Sander V, Sauter M, Takahashi Y, Touraine R, Youroukos S, Zonnenberg B, Jansen AC, consortium T, investigators T (2017) TuberOus SClerosis registry to increase disease Awareness (TOSCA) - baseline data on 2093 patients. Orphanet J Rare Dis 12: 2 doi:10.1186/s13023-016-0553-5

9. Roth J, Roach ES, Bartels U, Jozwiak S, Koenig MK, Weiner HL, Franz DN, Wang HZ (2013) Subependymal giant cell astrocytoma: diagnosis, screening, and treatment. Recommendations from the International Tuberous Sclerosis Complex Consensus Conference 2012. Pediatr Neurol 49: 439–444 doi:10.1016/j.pediatrneurol.2013.08.017

10. Crino PB, Nathanson KL, Henske EP (2006) The tuberous sclerosis complex. N Engl J Med 355:1345–1356. doi:10.1056/NEJMra055323

11. de Ribaupierre S, Dorfmuller G, Bulteau C, Fohlen M, Pinard JM, Chiron C, Delalande O (2007) Subependymal giant-cell astrocytomas in pediatric tuberous sclerosis disease: when should we operate? Neurosurgery 60: 83–89; discussion 89–90 doi:10.1227/01.NEU.0000249216.19591.5D

12. Krueger DA, Northrup H, International Tuberous Sclerosis Complex Consensus G (2013) Tuberous sclerosis complex surveillance and management: recommendations of the 2012 International Tuberous Sclerosis Complex Consensus Conference. Pediatr Neurol 49: 255–265 doi:10.1016/j.pediatrneurol.2013.08.002

13. Nguyen HS, Doan NB, Gelsomino M, Shabani S, Awad AJ, Best B, Kaushal M, Mortazavi MM (2018) Subependymal Giant Cell Astrocytoma: A Surveillance, Epidemiology, and End Results Program-Based Analysis from 2004 to 2013. World Neurosurg 118:e263–e268. doi:10.1016/j.wneu.2018.06.169
14. Braffman BH, Bilaniuk LT, Naidich TP, Altman NR, Post MJ, Quencer RM, Zimmerman RA, Brody BA (1992) MR imaging of tuberous sclerosis: pathogenesis of this phakomatosis, use of gadopentetate dimeglumine, and literature review. Radiology 183:227–238. doi:10.1148/radiology.183.1.1549677

15. Shepherd CW, Gomez MR, Lie JT, Crowson CS (1991) Causes of death in patients with tuberous sclerosis. Mayo Clin Proc 66:792–796. doi:10.1016/s0025-6196(12)61196-3

16. Nabbout R, Santos M, Rolland Y, Delalande O, Dulac O, Chiron C (1999) Early diagnosis of subependymal giant cell astrocytoma in children with tuberous sclerosis. J Neurol Neurosurg Psychiatry 66:370–375. doi:10.1136/jnnp.66.3.370

17. Jung TY, Kim YH, Jung S, Baek HJ, Lee KH (2015) The clinical characteristics of subependymal giant cell astrocytoma: five cases. Brain Tumor Res Treat 3:44–47. doi:10.14791/brtt.2015.3.1.44

18. Clarke MJ, Foy AB, Wetjen N, Raffel C (2006) Imaging characteristics and growth of subependymal giant cell astrocytomas. Neurosurg Focus 20:E5. doi:10.3171/foc.2006.20.1.6

19. Jansen AC, Belousova E, Benedik MP, Carter T, Cottin V, Curatolo P, Dahlin M, D'Amato L, d'Augeres GB, de Vries PJ, Ferreira JC, Feucht M, Fladrowski C, Hertzberg C, Joziwiaik S, Lawson JA, Macaya A, Marques R, Nabbout R, O'Callaghan F, Qin J, Sander V, Sauter M, Shah S, Takahashi Y, Touraine R, Youroukos S, Zonnenberg B, Kingswood JC (2019) Clinical Characteristics of Subependymal Giant Cell Astrocytoma in Tuberous Sclerosis Complex. Front Neurol 10:705. doi:10.3389/fneur.2019.00705

20. Fohlen M, Ferrand-Sorbets S, Delalande O, Dorfmuller G (2018) Surgery for subependymal giant cell astrocytomas in children with tuberous sclerosis complex. Childs Nerv Syst 34:1511–1519. doi:10.1007/s00381-018-3826-6

21. Kumar R, Singh V (2004) Subependymal giant cell astrocytoma: a report of five cases. Neurosurg Rev 27:274–280. doi:10.1007/s10143-004-0339-4

22. Cuccia V, Zuccaro G, Sosa F, Monges J, Lubienieky F, Taratuto AL (2003) Subependymal giant cell astrocytoma in children with tuberous sclerosis. Childs Nerv Syst 19:232–243. doi:10.1007/s00381-002-0700-2

23. Chan DL, Calder T, Lawson JA, Mowat D, Kennedy SE (2018) The natural history of subependymal giant cell astrocytomas in tuberous sclerosis complex: a review. Rev Neurosci 29:295–301. doi:10.1515/revneuro-2017-0027

Tables

Table 1.

Univariate analyses of 35 children with or without TSC.
| Variables                        | Total (n=35) | TSC (n=20) | No TSC (n=15) | P value |
|---------------------------------|--------------|------------|---------------|---------|
| Age, years\(^a\)               | 11.6±4.0     | 11.4±4.0   | 11.3±4.2      | 0.987   |
| Sex\(^b\)                       |              |            |               |         |
| Male                            | 21           | 12 (60.0)  | 9 (60.0)      | 0.999   |
| Female                          | 14           | 8 (40.0)   | 6 (40.0)      |         |
| Disease duration, months\(^a\)  | 3.0 (0.5, 19.0) | 3.0 (1, 72.0) | 1.0 (0.5, 12.0) | 0.243   |
| Onset symptom, number of cases\(^b\) |              |            |               |         |
| Headache                        | 23 (65.7)    | 13 (65.0)  | 10 (66.7)     | 0.700   |
| Seizure                         | 11 (31.4)    | 7 (35.0)   | 4 (26.6)      |         |
| Unsteady walking                | 1 (2.9)      | 0 (0)      | 1 (6.7)       |         |
| Hydrocephalus\(^b\)            | 28 (80.0)    | 17 (85.0)  | 11 (73.3)     | 0.430   |
| Tumor size\(^a\)                | 41.4±19.4    | 49.7±19.0  | 30.5±14.1     | 0.001   |
| Tumor location\(^b\)           |              |            |               |         |
| LV near the FM                  | 23 (65.7)    | 12 (60.0)  | 11 (73.3)     | 0.281   |
| LV far from the FM              | 11 (31.4)    | 8 (40.0)   | 3 (20.0)      |         |
| Medulla                         | 1 (2.9)      | 0 (0)      | 1 (6.7)       |         |
| Cyst\(^b\)                      | 5 (14.3)     | 4 (20.0)   | 1 (6.7)       | 0.365   |
| Calcification\(^b\)             | 12 (34.3)    | 7 (35.0)   | 5 (33.3)      | 0.999   |

Data are presented as mean ± standard deviation or median (interquartile range) or n (%).

FM, foramen of Monro
LV, lateral ventricle
TSC, tuberous sclerosis complex.
\(^a\)non-parametric Mann-Whitney U-test; \(^b\)chi-square test.

Table 2.
Risk factors for hydrocephalus after resection according to chi-square test.
| Factors                        | Hydrocephalus after resection (n=16) | No hydrocephalus after resection (n=19) | P value |
|-------------------------------|-------------------------------------|----------------------------------------|---------|
| Age (years)                   |                                     |                                        |         |
| <12                           | 8 (50.0)                            | 9 (47.4)                               | 0.999   |
| ≥12                           | 8 (50.0)                            | 10 (52.6)                              |         |
| Sex                           |                                     |                                        |         |
| Male                          | 10 (62.5)                           | 11 (57.9)                              | 0.999   |
| Female                        | 6 (37.5)                            | 8 (42.1)                               |         |
| TSC                           |                                     |                                        |         |
| Yes                           | 13 (81.3)                           | 7 (36.8)                               | 0.016   |
| No                            | 3 (18.8)                            | 12 (63.2)                              |         |
| Preoperative hydrocephalus    |                                     |                                        |         |
| Yes                           | 14 (87.5)                           | 14 (73.7)                              | 0.415   |
| No                            | 2 (12.5)                            | 5 (26.3)                               |         |
| Tumor location                |                                     |                                        |         |
| near the FM                   | 8 (50.0)                            | 15 (78.9)                              | 0.090   |
| far from the FM               | 8 (50.0)                            | 4 (21.1)                               |         |
| Tumor size (mm)               |                                     |                                        |         |
| <30                           | 2 (12.5)                            | 8 (42.1)                               | 0.071   |
| ≥30                           | 14 (87.5)                           | 11 (57.9)                              |         |
| The extent of resection       |                                     |                                        |         |
| GTR                           | 9 (56.3)                            | 17 (89.5)                              | 0.050   |
| STR                           | 7 (43.8)                            | 2 (10.5)                               |         |
| Postoperative intracranial infection |                 |                                        |         |
| Yes                           | 6 (37.5)                            | 7 (36.8)                               | 0.999   |
| No                            | 10 (62.5)                           | 12 (63.2)                              |         |

Data are presented as n (%).

FM, foramen of Monro.
GTR, gross total resection.

STR, subtotal resection.

TSC, tuberous sclerosis complex.

Table 3.

Risk factors for hydrocephalus after resection according to multivariate logistic regression.

|                | OR (95%CI)       | P value |
|----------------|------------------|---------|
| TSC            | 18.814 (1.020-347.120) | 0.048   |
| Tumor location | 5.595 (0.793-39.477)   | 0.084   |
| Tumor size     | 0.886 (0.043-18.266)  | 0.938   |
| Extent of resection | 0.042 (0.003-0.670) | 0.025   |

CI, confidence interval

OR, odds ratio.

TSC, tuberous sclerosis complex.

**Figures**

**Figure 1**

a-d: T1 enhanced sequence of brain MRI and pathological imaging in a 10-year-old girl. e-f: Skin lesions in a 12-year-old girl with associated TSC. a: Preoperative T1 enhancement showed a mass located in the left lateral ventricle near the foramen of Monro and inhomogeneous enhancement after enhancement. b: Postoperative T1 enhancement showed that no obvious abnormal enhancement was observed in the operative area after tumor resection. c: MRI showed hydrocephalus 2 months after surgery. d: The tumor consisted of polygonal cells and ganglionic-like cells with fascicles (Hematoxylin and eosin stain, ×400 magnification). e: Shagreen patch could be seen on left back, hypopigmented macule could be seen on right back. f: Angiofibromas could be seen on the face.