Published in final edited form as:
World Neurosurg. 2012 July ; 78(1-2): 137–144. doi:10.1016/j.wneu.2011.09.012.

Surgical Treatment of Brain Tumors in Infants Less than 6 months of Age and Literature Review

Shih-Shan Lang, MD[1,2], **Lauren A. Beslow, MD**[3], **Brandon Gabel, MD**[1], **Alex R. Judkins, MD**[4], **Michael J. Fisher, MD**[5,6], **Leslie N. Sutton, MD**[1,2], **Phillip B. Storm, MD**[1,2], and **Gregory G. Heuer, MD PhD**[1,2]

[1]Department of Neurosurgery, University of Pennsylvania Medical Center

[2]Division of Neurosurgery, Children's Hospital of Philadelphia

[3]Division of Neurology, Children's Hospital of Philadelphia

[4]Department of Pathology & Laboratory Medicine at Children's Hospital Los Angeles

[5]Division of Oncology, Children's Hospital of Philadelphia

[6]Department of Pediatrics, University of Pennsylvania

Abstract

Object—Brain tumors are rare in infants under 6-months of age. These tumors can be challenging to treat surgically. We analyzed a modern series of patients treated by a multidisciplinary team at a tertiary care center and performed a literature review of this unique population.

Methods—Retrospective clinical data was collected for patients surgically treated for intracranial mass lesions at The Children's Hospital of Philadelphia from 1998 to 2007. Dermoid cysts and other skull-based lesions were excluded from the analysis.

Results—Sixteen patients under 6-months of age underwent surgery for primary intracranial mass lesions. The median age of the patients at surgery was 5.2 months (range 1.4 to 6 months of age). Children most often presented with a bulging fontanelle, hydrocephalus, or macrocephaly (7 patients). Vomiting was seen in 5 patients, cranial nerve palsies in 1 patient, and seizures in 3 patients.

All patients had tumor resections and post-operatively were monitored in the intensive care unit. The final pathology consisted of atypical teratoid/rhabdoid tumor (3 cases), primitive neuroectodermal tumor/medulloblastoma (3 cases), choroid plexus papilloma (2 cases), astrocytoma (2 cases), ganglioglioma (2 cases), desmoplastic infantile ganglioglioma (2), glioblastoma multiforme (1), and choroid plexus carcinoma (1).

Two intra-operative deaths occurred. Of the surviving 14, a gross total resection was achieved in 4. Adjuvant therapy was determined by a multidisciplinary team composed of neuro-oncology, neurosurgery, and radiation oncology. Seven patients were treated with chemotherapy, 1 patient

Correspondence: Shih-Shan Lang, MD, Department of Neurosurgery, University of Pennsylvania, 3400 Spruce Street, 3rd Floor Silverstein Pavilion, Philadelphia, PA 19104, Phone: 215-662-3487, Fax: 215-349-5534, shihshan.lang@uphs.upenn.edu.

None of the authors has any disclosures or conflicts of interest.

had proton beam therapy. Five-year overall survival was 45%. The eight surviving patients had neurological sequelae, and developmental outcome was variable.

Conclusions—Brain tumors are uncommon in children under 6-months of age. Patients present with a variety of tumor pathologies. Children who survive have neurological sequelae. More studies are necessary to understand the impact that different treatment options, tumor pathology, and tumor location have on neurological outcome.

Keywords

brain tumor; surgery; neonates; congenital

Introduction

Brain tumors are rare in infants less than 6 months of age. Although brain tumors in children have been well characterized, there are relatively few studies that have looked specifically at children less than 6 months of age.

Treatment of very young children with malignant brain tumors requires an interdisciplinary approach[36]. Surgery remains a vital component of most treatment algorithms[28,40,48,50]. In order to minimize the harmful effects of radiation, chemotherapy has become the critical component of adjuvant therapy in the neonatal age group[14,16,20,22,28]. Chemotherapy is also used as a time “bridge” in order to delay radiation therapy, especially in younger children.

The treatment algorithms for childhood brain tumors are well defined for older children but are not as well defined for infants less than 6 months of age[40]. Therefore, a more thorough examination of the treatment modalities used, and the associated outcomes, will help establish the best methods for managing these patients. Additionally, characterizing the pathology and frequency of the various tumor types in this patient population is paramount to defining and developing the best treatment options[47].

Characterizing the types of tumors, and the treatment options that provide the best results may improve the chances of meaningful neurological and developmental recovery. This study illustrates the experience of a large tertiary care children’s hospital in treating infants less than 6 months of age with brain tumors. Additionally, since brain tumors in this age group are rare, we performed a literature review of published cases.

Clinical Materials and Methods

Study design and subjects

A neurosurgical database of brain tumor patients who were treated surgically was queried to identify all patients less than 6 months of age who underwent resection of primary intracranial mass lesions between 1998 and 2007 at The Children’s Hospital of Philadelphia (CHOP). With institutional review board approval, retrospective analysis of these patients was performed.

Clinical data

Data including patient demographics, clinical presentation, and treatment were abstracted from inpatient hospital records, neurosurgical outpatient clinic charts, and neuro-oncology outpatient clinic charts. Extent of resection was categorized based on the immediate (within one day) post-operative MRI. Categories were gross total resection (no residual disease), subtotal resection (50–99% reduction in tumor size), and biopsy (less than 50% reduction in tumor size). The Kaplan-Meier estimate of survival was calculated using STATA 11.1

(STATA Corporation, College Station, TX). Survival time was calculated from the date of the first neurosurgical intervention. Neurological outcomes were retrieved from neuro-oncology and/or neurosurgery clinic charts.

Literature Review

PubMed was searched for key words including infant, neonatal, and brain tumor (as well as specific brain tumor types). References of included papers were examined to find additional papers. A large literature review was used to identify potential cases that were missed in the original PubMed Search (Isaacs). The majority of the relevant publications reviewed patients up to 3 years of age. Only studies in which subjects less than 6 month of age could be identified clearly were included in our review.

Results

Patient characteristics and presentation

From 1998 to 2007, 16 patients 6 months old and younger with pathologically confirmed malignant and benign intracranial brain tumors were identified. Five (31%) were male and 11 (69%) were female. The median age at time of surgical treatment was 5.2 months (range 1.4 to 6.0 months), and only 2 patients were less than 3 months of age at the time of surgery. The most common presenting clinical signs were bulging fontanelle, hydrocephalus, or macrocephaly, occurring in 7 patients. Vomiting was seen in 5 patients, irritability or lethargy in 5 patients, seizures in 3 patients, and cranial nerve palsies in 1 patient. One patient (#5) was noted to have hydrocephalus on prenatal ultrasound (Table 1).

Pre-treatment imaging

Eleven tumors were supratentorial, and 5 were infratentorial. Specific tumor locations are presented in Table 1. Figure 1 and Figure 2 show examples of pre-treatment imaging.

Treatment

The patients were evaluated and treated by an interdisciplinary team as clinically indicated including pediatric neurosurgeons, neuroradiologists, neuropathologists, neurologists, neuro-oncologists, and radiation oncologists. All patients underwent surgical resection of their tumors with subsequent histopathological evaluation. Post-operatively all patients were monitored in the intensive care unit. Four patients (25%) had gross total surgical resection, nine patients (56%) had a subtotal surgical resection, and one patient (6%) had a biopsy. Two patients (13%) did not have a post-operative MRI because of intraoperative death. Seven patients (44%) received post-operative chemotherapy. One patient (6%) underwent proton beam therapy at another facility.

Pathology

Table 1 shows the pathological features of the sixteen patients' tumors.

Survival and neurological outcomes

Follow-up data was available in all patients. The median follow-up time was 0.9 years (interquartile range 0.3 years to 5.5 years). The median survival among the subjects was 1.4 years. Using the Kaplan-Meier method, the 1-year, 2-year, and 5-year survival were 53%, 45%, 45%, respectively (Figure 3). There were 8 deaths that occurred at a median of 6.4 months of age (interquartile range 5.9 to 15.5 months) and a median of 3.9 months from surgical resection (interquartile range 0.2 to 10.7 months). Two infants died intraoperatively, one of whom presented with herniation and was operated upon emergently. The other 6 deaths were a result of tumor progression. Of the 8 surviving patients, all are in continuous

remission (as demonstrated by yearly MRIs) at a median age of 5.9 years (interquartile range 1.6–8 years). Of the four patients in whom a gross total resection was achieved, two continue to be tumor-free; the other two patients (both with atypical rhabdoid teratoid tumors) died of progression of recurrent tumor (Table 1). All 8 surviving patients had neurological sequelae. Deficits and their severity varied, ranging from mild hemiparesis to severe motor deficits with cognitive and behavioral problems (Table 1).

Literature review

One hundred twenty-three papers were reviewed and 44 papers that were suitable for the current study were identified. These included 29 case reports or series and 15 retrospective studies. The number of patients included in each reviewed paper ranged from a single patient in a case report to 250 patients in a literature review[24,25]. These 44 papers yielded 468 cases of intracranial tumors in children 6 months of age and less[1,3–5,8–10,12,13,15,17–19,21,26,27,29–31,33–35,37,38,40–46,51–55,57–59,61,62,65–67]. There was significant variability in brain tumor histologies (Table 2). Teratoma was the most common tumor reported in the existing literature and was mainly found in neonates within a few weeks of life; up to 83.7% of these patients died. The next most common tumor types were choroid plexus tumors and astrocytomas. The estimated survival of the 445 subjects who had follow-up data available was 30.8% (Table 2).

Discussion

Location of tumors in infants less than 6 months does not appear to follow the trends seen in older pediatric populations. In our series, nearly 70% of tumors were located supratentorially, a finding consistent with prior reports[55]. This contrasts with older pediatric patients in whom infratentorial tumors are more common[7,11,22–25,32,39,40,56]. As with older children, the histopathological diagnoses varied widely; however, there was a higher incidence of atypical teratoid rhabdoid tumors and choroid plexus tumors.

Our patients had a 5-year survival of 45%, compared with an estimated survival of 30.8% in the literature review. However, follow-up time was not available for all the subjects in the literature review, limiting our ability to calculate a 5-year survival for the patients represented in the literature and thus to compare the survival in the literature to that of our patients. Additionally, in contrast to prior studies[55] our study had no subjects with teratoma, a tumor type with high mortality. The absence of teratoma patients may explain the higher survival of our patient population compared with the literature. However, since our selection of patients utilized a surgical database, it is possible that infants with teratoma may have been missed if no surgical treatment was indicated or elected due to poor prognosis, thereby inflating the survival estimate. Additionally, our series had 5 patients with low-grade tumors which have the highest survival of the infantile brain tumors. Strengths of this study include its relatively long follow-up compared with historical data. In addition, given that all patients presented since 1998, it represents a relatively modern surgical series.

Developmental and neurological outcomes in very young children with tumors are variable. All surviving children in our series had residual neurological or developmental deficits at follow-up. However, the degree of disability varied widely between patients. Post-operative and long-term quality of life is a topic seldom discussed in the neurosurgical literature. However, neurological sequelae are important. Compared to other pediatric tumor patients and even to adult brain tumor patients, the pediatric brain tumor population faces significantly more devastating neurological, psychosocial, and economic effects[63]. Further characterization of the neuro-cognitive deficits of this population as the children enter school, utilizing formal neuro-psychological testing, will be crucial to characterizing the scope of issues facing survivors of infant brain tumors.

The primary mode of treatment for infantile brain tumors remains surgery, although the prognosis for many of these tumors remains poor. Importantly, in our experience it can be difficult to predict the tumor type based on the preoperative imaging for some of these young patients, necessitating surgical treatment to establish the diagnosis and in some cases to provide definitive treatment. The use of adjuvant chemotherapy and its effects on outcome need to be explored further, especially since radiation therapy is not usually an option in infants[16]. Re-operating on stable residual masses has also been questioned by some authors, citing that a “wait and see” method may be better than subjecting the child to a second operation[6]. It is our practice to determine the role for reoperation based on the tumor histology and the neurological status of the child. As more data for this patient population becomes available, the characterization and best treatment strategies will become further refined. Given the many advancements and improvements in surgical technique, radiation, and chemotherapy options, the population of pediatric brain tumor survivors continues to grow. The advent and increasing availability of proton beam radiation, with the goal of increasing survival while minimizing neurological repercussions, may allow for the use of radiotherapy in patients younger than was previously believed to be safe. A thorough understanding of the residual neurological deficits and quality of life in survivors is required in order to improve the care of these patients. More research is being dedicated to treatment-related effects, such as the long-term impact of focal proton beam radiation on the growing brain and body[2,16,20,49,60,64]. Larger series will be necessary to correlate outcome with tumor type, extent of resection, and/or response to adjuvant therapies.

References

1. Alexiou GA, Stefanaki K, Sfakianos G, Prodromou N. Desmoplastic infantile ganglioglioma: a report of 2 cases and a review of the literature. Pediatr Neurosurg. 2008; 44:422–425. [PubMed: 18703892]
2. Allen JC. What infants can teach us about brain tumors. N Engl J Med. 1993; 328:1780–1781. [PubMed: 8497288]
3. Araki K, Aori T, Takahashi JA, Nozaki K, Nagata I, Kikuchi H, Yokoyama M, Hattori H, Akiyama Y, Kubota Y, Yokomizo H. [A case report of choroid plexus carcinoma]. No Shinkei Geka. 1997; 25:853–857. [PubMed: 9300457]
4. Arico M, Raiteri E, Bossi G, Giordana MT, Corbella F, Locatelli D, Pezzotta S. Choroid plexus carcinoma: report of one case with favourable response to treatment. Med Pediatr Oncol. 1994; 22:274–278. [PubMed: 8107660]
5. Arnstein LH, Boldrey E, Naffziger HC. A case report and survey of brain tumors during the neonatal period. J Neurosurg. 1951; 8:315–319. [PubMed: 14841542]
6. Benesch M, Eder HG, Sovinz P, Raith J, Lackner H, Moser A, Urban C. Residual or recurrent cerebellar low-grade glioma in children after tumor resection: is re-treatment needed? A single center experience from 1983 to 2003. Pediatr Neurosurg. 2006; 42:159–164. [PubMed: 16636617]
7. Blaney, SMH-KD.; Poussaint, TY.; Santi, M.; Gibertson, R.; Parsons, D.; Pollack, I., editors. Principles and Practice of Pediatric Oncology, ed 6th. Philadelphia: Lippincott Williams & Wilkins; 2011.
8. Borch K, Jacobsen T, Olsen JH, Hirsch F, Hertz H. Neonatal cancer in Denmark 1943–1985. Pediatr Hematol Oncol. 1992; 9:209–216. [PubMed: 1524999]
9. Boyd MC, Steinbok P. Choroid plexus tumors: problems in diagnosis and management. J Neurosurg. 1987; 66:800–805. [PubMed: 3572508]
10. Carstensen H, Juhler M, Bogeskov L, Laursen H. A report of nine newborns with congenital brain tumours. Childs Nerv Syst. 2006; 22:1427–1431. [PubMed: 16804715]
11. CBTRUS: Central Brain Tumor Registry of the United States. Primary brain tumors in the United States statistical report 2000–2004, in, 2007–2008.

12. Chadduck WM, Gollin SM, Gray BA, Norris JS, Araoz CA, Tryka AF. Gliosarcoma with chromosome abnormalities in a neonate exposed to heptachlor. Neurosurgery. 1987; 21:557–559. [PubMed: 3683793]

13. Chung SK, Wang KC, Nam DH, Cho BK. Brain tumor in the first year of life: a single institute study. Journal of Korean Medical Science. 1998; 3:65–70. [PubMed: 9539322]

14. Cohen BH, Packer RJ, Siegel KR, Rorke LB, D'Angio G, Sutton LN, Bruce DA, Schut L. Brain tumors in children under 2 years: treatment, survival and long-term prognosis. Pediatric Neurosurgery. 1993; 19:171–179. [PubMed: 8329301]

15. de Leon-Bojorge B, Rueda-Franco F, Anaya-Jara M. Central nervous system atypical teratoid rhabdoid tumor: experience at the National Institute of Pediatrics, Mexico City. Childs Nerv Syst. 2008; 24:307–312. [PubMed: 17876589]

16. Duffner PK, Horowitz ME, Krischer JP, Friedman HS, Burger PC, Cohen ME, Sanford RA, Mulhern RK, James HE, Freeman CR, et al. Postoperative chemotherapy and delayed radiation in children less than three years of age with malignant brain tumors. N Engl J Med. 1993; 328:1725–1731. [PubMed: 8388548]

17. Fenton LZ, Foreman NK. Atypical teratoid/rhabdoid tumor of the central nervous system in children: an atypical series and review. Pediatr Radiol. 2003; 33:554–558. [PubMed: 12759791]

18. Galassi E, Godano U, Cavallo M, Donati R, Nasi MT. Intracranial tumors during the 1st year of life. Childs Nerv Syst. 1989; 5:288–298. [PubMed: 2804999]

19. Gianella-Borradori A, Zeltzer PM, Bodey B, Nelson M, Britton H, Marlin A. Choroid plexus tumors in childhood. Response to chemotherapy, and immunophenotypic profile using a panel of monoclonal antibodies. Cancer. 1992; 69:809–816. [PubMed: 1370394]

20. Gottardo NG, Gajjar A. Chemotherapy for malignant brain tumors of childhood. J Child Neurol. 2008; 23:1149–1159. [PubMed: 18952581]

21. Hahn JS, Bejar R, Gladson CL. Neonatal subependymal giant cell astrocytoma associated with tuberous sclerosis: MRI, CT, and ultrasound correlation. Neurology. 1991; 41:124–128. [PubMed: 1985277]

22. Hargrave DR, Zacharoulis S. S: Pediatric CNS tumors: current treatment and future directions. Expert Rev Neurother. 2007; 7:1029–1042. [PubMed: 17678498]

23. Heuer GG, Jackson EM, Magge SN, Storm PB. Surgical management of pediatric brain tumors. Expert Rev Anticancer Ther. 2007; 7:S61–S68. [PubMed: 18076320]

24. Isaacs H Jr. I. Perinatal brain tumors: a review of 250 cases. Pediatr Neurol. 2002; 27:249–261. [PubMed: 12435562]

25. Isaacs H Jr. II. Perinatal brain tumors: a review of 250 cases. Pediatr Neurol. 2002; 27:333–342. [PubMed: 12504200]

26. Itoh Y, Kowada M, Mineura K, Kojima H. Congenital glioblastoma of the cerebellum with cytofluorometric deoxyribonucleic acid analysis. Surg Neurol. 1987; 27:163–167. [PubMed: 3810444]

27. Jung WH, Choi S, Oh KK, Chi JG. Congenital glioblastoma multiforme--report of an autopsy case. J Korean Med Sci. 1990; 5:225–231. [PubMed: 1966036]

28. Knab B, Connell PP. Radiotherapy for pediatric brain tumors: when and how. Expert Rev Anticancer Ther. 2007; 7:S69–S77. [PubMed: 18076312]

29. Kumabe T, Tominaga T, Kondo T, Yoshimoto T, Kayama T. Intraoperative radiation therapy and chemotherapy for huge choroid plexus carcinoma in an infant--case report. Neurol Med Chir (Tokyo). 1996; 36:179–184. [PubMed: 8869156]

30. Kwinta P, Kwiatkowski S, Tomasik T, Grudzien A, Korab-Chrzanowska E, Adamek D, et al. [Congenital brain tumor in neonate--case report and review of literature]. Przeglad Lekarski. 2005; 62:1302–1307. [PubMed: 16512626]

31. Lalitha VS, Rubinstein LJ. Reactive glioma in intracranial sarcoma: a form of mixed sarcoma and glioma ("sarcoglioma"): report of eight cases. Cancer. 1979; 43:246–257. [PubMed: 216476]

32. Larouche V, Huang A, Bartels U, Bouffet E. Tumors of the central nervous system in the first year of life. Pediatr Blood Cancer. 2007; 49:1074–1082. [PubMed: 17943961]

33. Lasky JL, Choi EJ, Johnston S, Yong WH, Lazareff J, Moore T. Congenital brain tumors: case series and review of the literature. J Pediatr Hematol Oncol. 2008; 30:326–331. [PubMed: 18391706]
34. Mapstone TB, Warf BC. Intracranial tumor in infants: characteristics, management, and outcome of a contemporary series. Neurosurgery. 1991; 28:343–348. [PubMed: 2011214]
35. Marsh JS. Supratentorial glioblastoma multiforme in infancy; report of three cases occurring before the age of one. Bull Los Angel Neuro Soc. 1956; 21:30–36. [PubMed: 13329555]
36. Mazewski CM, Hudgins RJ. Neonatal brain tumors: a review. Semin Perinatol. 1999; 23:286–298. [PubMed: 10475542]
37. Medhkour A, Traul D, Husain M. Neonatal subependymal giant cell astrocytoma. Pediatr Neurosurg. 2002; 36:271–274. [PubMed: 12053047]
38. Mehrotra N, Shamji MF, Vassilyadi M, Ventureyra EC. Intracranial tumors in first year of life: the CHEO experience. Childs Nerv Syst. 2009; 25:1563–1569. [PubMed: 19551387]
39. Merchant TE, Pollack IF, Loeffler JS. Brain tumors across the age spectrum: biology, therapy, and late effects. Semin Radiat Oncol. 2010; 20:58–66. [PubMed: 19959032]
40. Nejat F, El Khashab M, Rutka JT. Initial management of childhood brain tumors: neurosurgical considerations. J Child Neurol. 2008; 23:1136–1148. [PubMed: 18952580]
41. Newbould MJ, Kelsey AM, Arango JC, Ironside JW, Birch J. The choroid plexus carcinomas of childhood: histopathology, immunocytochemistry and clinicopathological correlations. Histopathology. 1995; 26:137–143. [PubMed: 7737660]
42. Packer RJ, Perilongo G, Johnson D, Sutton LN, Vezina G, Zimmerman RA, Ryan J, Reaman G, Schut L. Choroid plexus carcinoma of childhood. Cancer. 1992; 69:580–585. [PubMed: 1728390]
43. Papadakis N, Millan J, Grady DF, Segerberg LH. Medulloblastoma of the neonatal period and early infancy. Report of two cases. J Neurosurg. 1971; 34:88–91. [PubMed: 5312860]
44. Parkes SE, Muir KR, Southern L, Cameron AH, Darbyshire PJ, Stevens MCG. Neonatal Tumors. A Thirty-Year Population-Based Study. Medical and Pediatric Oncology. 1994; 22:309–317. [PubMed: 8127254]
45. Pascual-Castroviejo I, Villarejo F, Perez-Higueras A, Morales C, Pascual-Pascual SI. Childhood choroid plexus neoplasms. A study of 14 cases less than 2 years old. Eur J Pediatr. 1983; 140:51–56. [PubMed: 6873113]
46. Raimondi AJ, Tomita T. Brain tumors during the first year of life. Childs Brain. 1983; 10:193–207. [PubMed: 6603338]
47. Reddy AT. Advances in biology and treatment of childhood brain tumors. Curr Neurol Neurosci Rep. 2001; 1:137–143. [PubMed: 11898509]
48. Rivera-Luna R, Medina-Sanson A, Leal-Leal C, Pantoja-Guillen F, Zapata-Tarres M, Cardenas-Cardos R, Barrera-Gómez R, Rueda-Franco F. Brain tumors in children under 1 year of age: emphasis on the relationship of prognostic factors. Childs Nerv Syst. 2003; 19:311–314. [PubMed: 12732940]
49. Rivera-Luna R, Zapata-Tarres M, Medina-Sanson A, Lopez-Aguilar E, Niembro-Zuniga A, Amador Zarco J, Marhx-Bracho A, Rueda-Franco F, Bornstein-Quevedo L. Longterm survival in children under 3 years of age with low-grade astrocytoma. Childs Nerv Syst. 2007; 23:543–547. [PubMed: 17226033]
50. Rutka JT, Kuo JS. Pediatric surgical neuro-oncology: current best care practices and strategies. J Neurooncol. 2004; 69:139–150. [PubMed: 15527086]
51. Sabet LM. Congenital glioblastoma multiforme associated with congestive heart failure. Arch Pathol Lab Med. 1982; 106:31–34. [PubMed: 6274272]
52. Sacrez R, Juif JG, Friederich E. [Cerebral tumor in infant.]. Arch Fr Pediatr. 1954; 11:261–263. [PubMed: 13159411]
53. Sakamoto K, Kobayashi N, Ohtsubo H, Tanaka Y. Intracranial tumors in the first year of life. Childs Nerv Syst. 1986; 2:126–129. [PubMed: 3779667]
54. Sato O, Tamura A, Sano K. Brain tumors of early infants. Childs Brain. 1975; 1:121–125. [PubMed: 1183254]

55. Serowka K, Chiu Y, Gonzalez I, Gilles F, McComb G, Krieger M, et al. Central nervous system (CNS) tumors in the first six months of life: the Children's Hospital Los Angeles experience, 1979–2005. Pediatr Hematol Oncol. 27:90–102. [PubMed: 20201690]
56. Severino M, Schwartz ES, Thurnher MM, Rydland J, Nikas I, Rossi A. Congenital tumors of the central nervous system. Neuroradiology. 52:531–548. [PubMed: 20428859]
57. Shamji MF, Vassilyadi M, Lam CH, Montes JL, Farmer JP. Congenital tumors of the central nervous system: the MCH experience. Pediatr Neurosurg. 2009; 45:368–374. [PubMed: 19907201]
58. Shimamura N, Asano K, Ogane K, Yagihashi A, Ohkuma H, Suzuki S. A case of definitely congenital glioblastoma manifested by intratumoral hemorrhage. Childs Nerv Syst. 2003; 19:778–781. [PubMed: 13680296]
59. Singhal RM, Jain M, Tanwar RK, Raman NV. Congenital glioblastoma. Indian J Pediatr. 1994; 61:733–736. [PubMed: 7721383]
60. Skowronska-Gardas A. Evaluation of radiotherapy for pediatric CNS tumors. Expert Rev Neurother. 2003; 3:491–500. [PubMed: 19810933]
61. Takaku A, Kodama N, Ohara H, Hori S. Brain tumor in newborn babies. Childs Brain. 1978; 4:365–375. [PubMed: 308000]
62. Thiele RL, Dimmick MJ. Intracranial malignant neoplasm with onset before birth. J Pediatr. 1951; 39:611–615. [PubMed: 14889406]
63. Turner CD, Rey-Casserly C, Liptak CC, Chordas C. Late effects of therapy for pediatric brain tumor survivors. J Child Neurol. 2009; 24:1455–1463. [PubMed: 19841433]
64. Ward C, Phipps K, de Sousa C, Butler S, Gumley D. Treatment factors associated with outcomes in children less than 3 years of age with CNS tumours. Childs Nerv Syst. 2009; 25:663–668. [PubMed: 19247674]
65. Xue H, Horwitz JR, Smith MB, Lally KP, Black CT, Cangir A, Takahashi H, Andrassy RJ. Malignant solid tumors in neonates: a 40-year review. J Pediatr Surg. 1995; 30:543–545. [PubMed: 7595829]
66. Yang C-P, Hung I-J, Jaing T-H, Shih L-Y, Chang W-H. Cancer in infants: a review of 82 cases. Pediatric Hematology & Oncology. 2005; 22:463–481. [PubMed: 16169813]
67. Young HK, Johnston H. Intracranial tumors in infants. J Child Neurol. 2004; 19:424–430. [PubMed: 15446390]

Abbreviations

R	right
L	left
B	bilateral
PNET	primitive neuroectodermal tumor

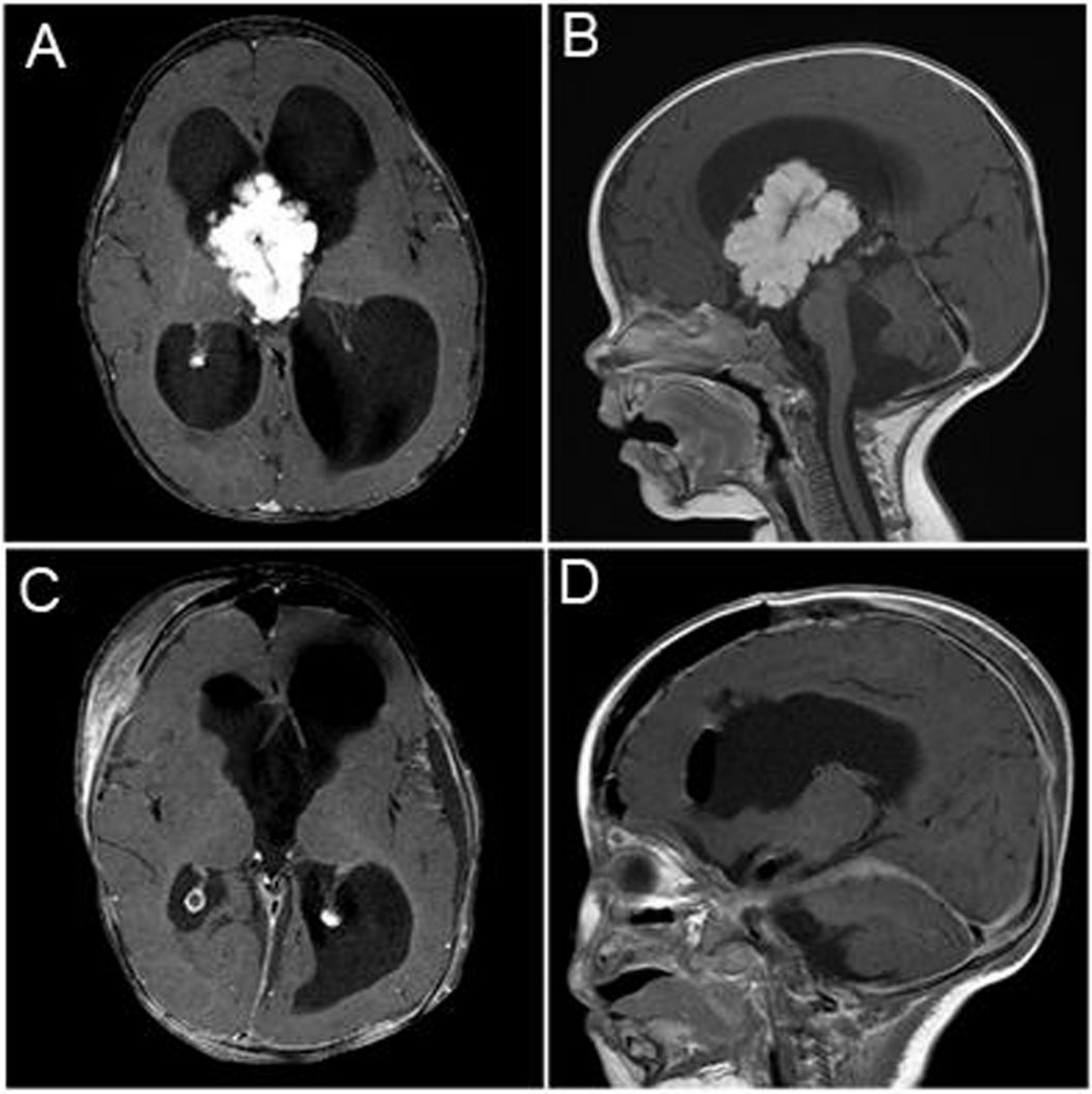

Figure 1.
Patient 1 was a 5 month-old girl that presented with increased head size and neck pain and stiffness. Axial (A) and sagittal (B) post-gadolinium T1 MR images demonstrated a large enhancing intraventricular mass with significant hydrocephalus. The patient underwent a craniotomy and resection. Post-operative axial (C) and coronal (D) post-gadolinium T1 imaging demonstrated gross total resection of the mass. On pathology the patient was found to have a choroid plexus papilloma.

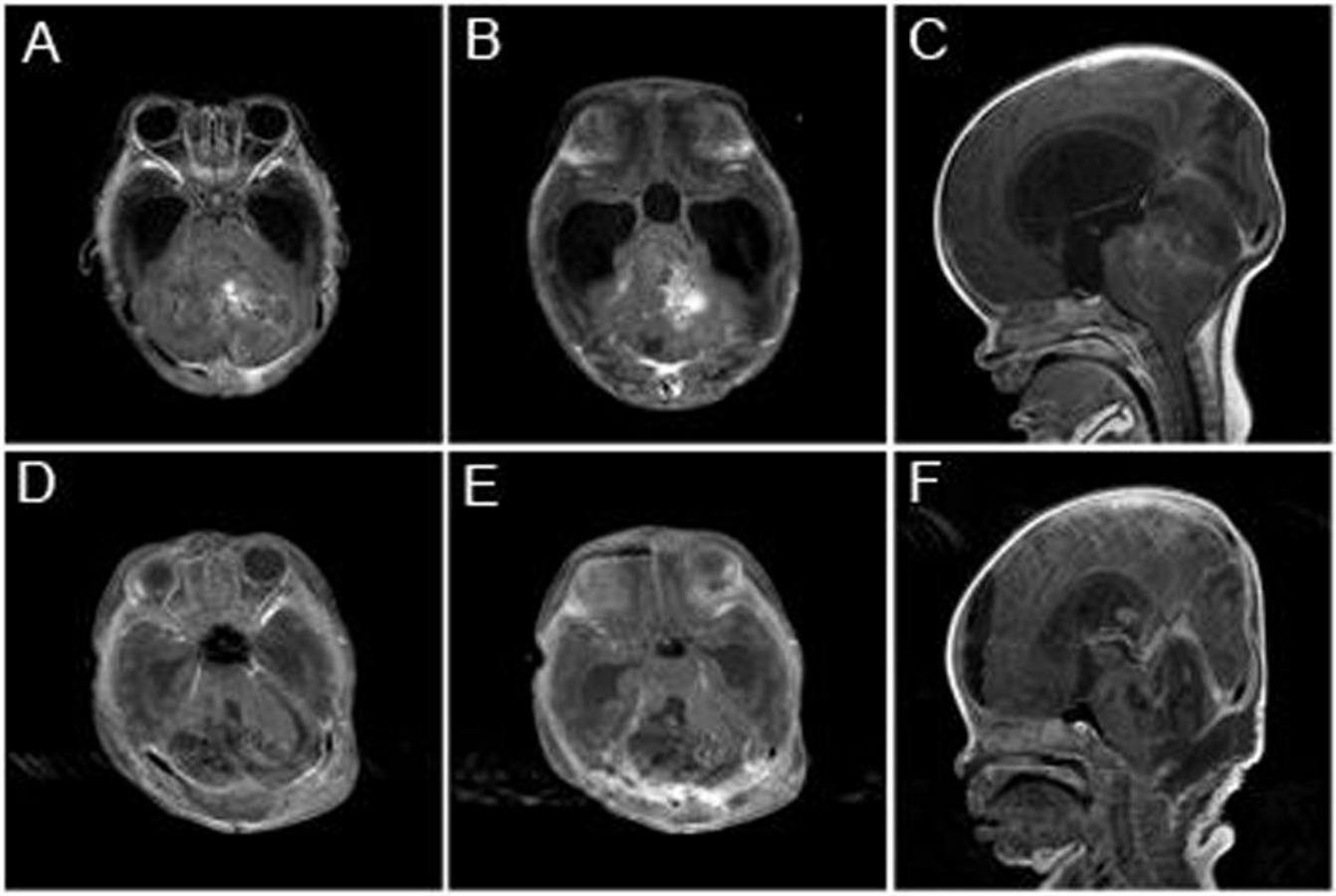

Figure 2.
Patient 16 was a 1 month-old girl that presented with increased head size and fussiness. Axial (A and B) and sagittal (C) post-gadolinium T1 MR images a demonstrated a large enhancing posterior fossa mass with significant hydrocephalus. The patient underwent a suboccipital craniotomy and resection. Post-operative axial (D and E) and sagittal (F) post-gadolinium T1 images demonstrated resection of the mass. On pathology the patient was found have a medulloblastoma with myoblastic differentiation.

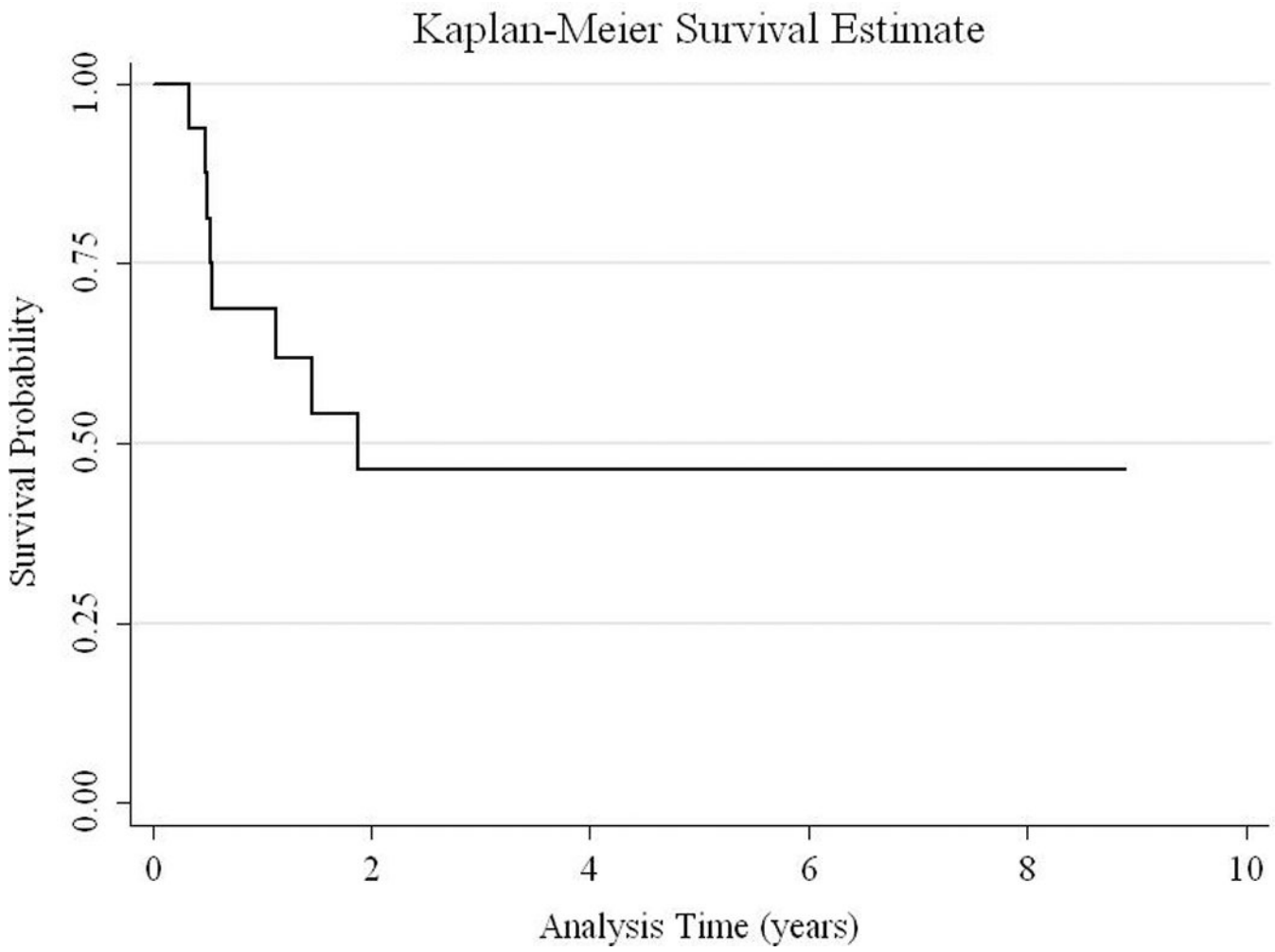

Figure 3.
Kaplan-Meier Survival Curve

Table 1

Clinical Information on Subjects

Patient	Age at Surgery (months)	Presenting Symptoms and Signs	Tumor Location	Diagnosis	Extent of Resection	Adjuvant therapy	Age at Death or Last Follow-up (years)	Outcome
1	5.6	Irritability, frontal bossing	3rd ventricle with extension into lateral ventricles	Choroid plexus papilloma	GTR	None	0.9	Difficulty cruising and standing
2	3.8	Not available	Superior cerebellar and pineal region	Atypical teratoid/rhabdoid tumor	GTR	None	0.5	Deceased
3	5.6	Seizures, bulging fontanelle	R temporal lobe	Desmoplastic infantile ganglioglioma	STR (>90%)	VP shunt	7.5	Partial R cranial nerve III palsy, L hemiparesis, aggressive behavior and learning difficulties
4	5.7	Vomiting, irritability	L lateral ventricle	Choroid plexus carcinoma	STR (>90%)	Chemotherapy	1.9	Deceased
5	2.1	Hydrocephalus on fetal ultrasound	Hypothalamic/suprasellar region	Ganglioglioma	STR (>70%)	B VP shunt Chemotherapy	4.8	Precocious puberty, seizures
6	5.2	Hydrocephalus, limited gaze, increased tone	L parietal/occipital lobe	Anaplastic astrocytoma	STR (>70%)	VP shunt Chemotherapy	8.4	Seizures, spastic gait, B adduction palsies, severe cognitive and developmental delay
7	5.9	Vomiting, irritability, hydrocephalus	L lateral ventricle	Choroid plexus papilloma	Intraoperative death	None	0.5	Deceased (intraoperative death)
8	6	Vomiting, large fontanelle	Pineal region	Atypical teratoid/rhabdoid tumor	STR (>90%)	VP shunt Chemotherapy	1.5	Deceased
9	3.5	Not available	Pineal region	PNET	STR (>70%)	None	1.1	Deceased
10	4.0	Not available	B cerebellum	Anaplastic ganglioglioma	STR (>90%)	None	8.9	Dysarthric speech, nystagmus, significant learning problems (requires classroom aide)
11	5.6	Vomiting, multiple cranial nerve deficits	R cerebellar-pontine angle	Atypical teratoid/rhabdoid tumor	Biopsy	None	0.5	Deceased
12	3.9	Lethargy, strabismus, herniation	R cerebral hemisphere	Glioblastoma multiforme	Intraoperative death	None	0.3	Deceased, (intra operative death, herniation prior to resection)
13	5.7	Not available	R hippocampal region extending to lateral basal ganglia and thalamus	Pilocytic astrocytoma	STR (>70%)	Chemotherapy	6.9	L spastic hemiparesis
14	5.2	Not available	Cerebellar vermis	Medulloblastoma	STR (>90%)	VP shunt Chemotherapy Radiation (proton)	1.2	Does not crawl, cruise or stand, few spoken words, little developmental progress

Patient	Age at Surgery (months)	Presenting Symptoms and Signs	Tumor Location	Diagnosis	Extent of Resection	Adjuvant therapy	Age at Death or Last Follow-up (years)	Outcome
15	4.4	Partial seizures with staring, apnea, cyanosis	R temporal, occipital, and posterior parietal lobes	Fibrillary astrocytoma	GTR *	None	2.1	Seizure free, mild L hemiparesis, no behavioral problems
16	1.4	Irritability, bulging fontanelle	B cerebellum	PNET (medullo-myoblastoma)	GTR	VP shunt	0.5	Deceased

* After initial surgery, post-operative day 1 MRI revealed residual enhancement. The patient underwent additional surgery within weeks and post-operative MRI demonstrated gross total resection.

GTR, gross total resection; STR, subtotal resection; VP, ventriculoperitoneal; R, right; L, left; B, bilateral

Table 2

Literature Review Tumor Types and Survival

Tumor Type	Number (%) *	Living	Dead	Percent Survival §
Astroblastoma	2 (0.4%)	1	1	50
Astrocytoma (most grades not specified)	85 (18.2%)	34	48 3 additional lost to follow-up	40
Atypical teratoid rhabdoid tumor	11 (2.4%)	0	11	0
Craniopharyngioma	20 (4.3%)	4	16	20
Choroid plexus papilloma	38 (8.1%)	33	4 1 additional lost to follow-up	87
Choroid plexus carcinoma	24 (5.1%)	10	14	42
Desmoplastic infantile ganglioglioma	2 (0.4%)	2	0	100
Ependymoma	16 (3.4%)	2	13 1 additional lost to follow-up	12.5
Ependymoblastoma	4 (0.9%)	0	4	0
Ganglioglioma	8 (1.7%)	8	0	100
Gangliocytoma	3 (0.6%)	3	0	100
Glioma (most grades not specified)	6 (1.3%)	3	3	50
Glioblastoma multiforme	23 (4.9%)	3	20	13
Gliosarcoma	3 (0.6%)	1	1 1 additional lost to follow-up	33
Hemangioblastoma	2 (0.4%)	1	1	50
Medulloblastoma	22 (4.7%)	2	17 3 additional lost to follow-up	9
Medulloepithelioma	4 (0.9%)	0	4	0
Meningioma	8 (1.7%)	5	3	62.5
Meningeal sarcoma	8 (1.7%)	2	6	25
Oligodendroglioma	3 (0.6%)	1	2	33
Oligoastrocytoma	1 (0.2%)	1	0	100
Pineoblastoma	3 (0.6%)	0	2 1 additional lost to follow-up	0
Primitive neuroectodermal tumor	44 (9.4%)	6	36 2 additional lost to follow-up	13.6
Sarcoma	3 (0.6%)	0	2 1 additional lost to follow-up	0
Subependymal giant cell astrocytoma	2 (0.4%)	1	1	50
Spongioblastoma	4 (0.9%)	0	4	0
Teratoma	98 (20.9%)	16	78 4 additional lost to follow-up	16.3
Miscellaneous **	21 (4.5%)	5	10 6 additional lost to follow-up	23.8
TOTAL	468 (100%)	144 alive	301 dead 23 lost to follow-up	30.8

* Percent of total cases

** 9 choroid plexus tumors not otherwise specified, 1 atypical choroid plexus papilloma, 1 middle fossa neuroblastoma,1 gliofibroma, 1 malignant embryonic tumor, 1 plexus tumor, 1 oligodendrocytoma, 1 midbrain cystic blastoma, 1 melanotic prognoma, 1 hemangioendothelioma, 3 not otherwise specified

§ Patients lost to follow-up were not included in the calculation of percentage survival.