J Neurosurg Pediatrics 1:152–155, 2008

Intraventricular astroblastoma

Case report

Luca Denaro, M.D.,[1,2] Marina Gardiman, M.D.,[3] Milena Calderone, M.D.,[4] Marta Rossetto, M.D.,[1] Pietro Ciccarino, M.D.,[1] Felice Giangaspero, M.D.,[5] Giorgio Perilongo, M.D.,[6] and Domenico d'Avella, M.D.[1]

[1]Department of Neuroscience, Neurosurgery, Departments of [3]Pathology and [6]Pediatrics, and [4]Neuroradiology Service, University of Padua; [2]Department of Neurosurgery, Catholic University, Rome; and [5]Department of Experimental Medicine, University of Rome La Sapienza, IRCCS Neuromed, Pozzilli, Italy

✓Astroblastoma is a rare primary brain neoplasm that accounts for 0.45–2.8% of brain gliomas. Intraventricular localization is extremely rare. The authors report a case of well-differentiated completely intraventricular astroblastoma in a 6-year-old girl and review the relevant literature. Their patient presented with a 5-week history of progressive nausea and vomiting. Magnetic resonance (MR) imaging revealed a large, well-demarcated, solid-cystic mass in the left temporooccipital ventricular horn. Macroscopic radical resection of the tumor was performed via the superior temporal sulcus. The postoperative course was uneventful and no adjuvant therapy was administered after surgery. No recurrence was detected at 9-months follow-up.

Gross-total resection has the greatest impact on patient survival. In differentiated tumors, recurrence is usually local, and adjuvant therapy is recommended after repeated resection for the treatment of recurrence. In patients harboring anaplastic astroblastoma, gross-total resection and adjuvant therapy after the initial surgery seems to be the best choice. It is important to distinguish astroblastoma from ependymoma in clinical practice because of the differences in therapeutic approaches. *(DOI: 10.3171/PED/2008/1/2/152)*

Key Words • astroblastoma • intraventricular tumor • pediatric neurosurgery

Astroblastoma is a rare primary brain neoplasm that accounts for 0.45–2.8% of brain gliomas.[3] It is usually a circumscribed, well-demarcated solid or cystic tumor with a characteristic perivascular orientation of epithelioid neoplastic cells.[1] Our review of the literature showed that since the lesion was first described by Bailey and Cushing in 1924, 140 cases of astroblastoma have been reported.[1,3,6–16,18,19,23,24,26] These tumors are usually located in the cerebral hemispheres but have also been described in the cerebellum, brainstem, corpus callosum, optic nerve, cauda equina, and hypothalamus.[18,26] In only 4 cases were the lesions located in the ventricular system.[3,18] We report a new case of a well-differentiated completely intraventricular astroblastoma.

Case Report

History and Presentation. This 6-year-old girl presented with a 5-week history of progressive headache and vomiting.

Neuroimaging Findings. Computed tomography of the patient's brain showed a solid, slightly hyperintense, microcystic intraventricular lesion with punctuate calcifications. Considerable shift of the midline structures was present, but perilesional edema was scarce (Fig. 1). An MR imaging study revealed a large well-demarcated solid-cystic mass in the left temporooccipital ventricular horn (Fig. 2). The solid portion appeared as hypointense to white matter on T1-weighted images and hypointense on T2-weighted images, with evidence of cystic components. After gadolinium injection, the lesion showed a heterogeneous enhancement of the solid portion and rim enhancement of cysts walls (Fig. 2). Relatively little peritumoral edema was present.

Operation. A left temporoparietal craniotomy was performed. After the dura was opened, the cerebral cortex appeared tight. The tumor, which was approached via the superior temporal sulcus, was found to be a soft, reddish intraventricular mass located immediately below the cortex. The lesion was completely intraventricular with well-circumscribed margins and no evidence of infiltration or local invasion into the ependyma. A macroscopic radical resection of the tumor was performed with the use of microsurgical technique with bipolar diathermy and suction.

Abbreviations used in this paper: EMA = endothelial membrane antigen; GFAP = glial fibrillary acidic protein; MR = magnetic resonance.

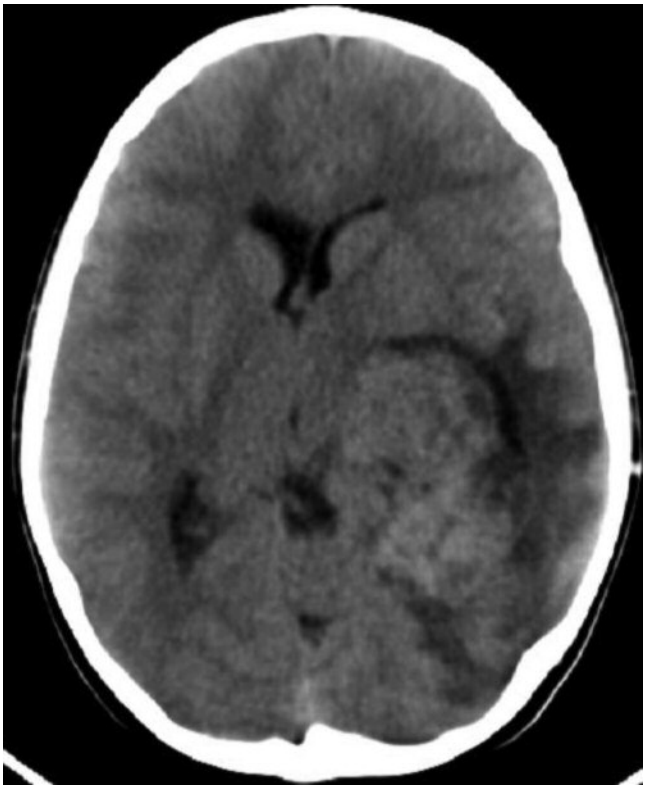

FIG. 1. Preoperative computed tomography showing a solid, slightly hyperintense, microcystic intraventricular lesion.

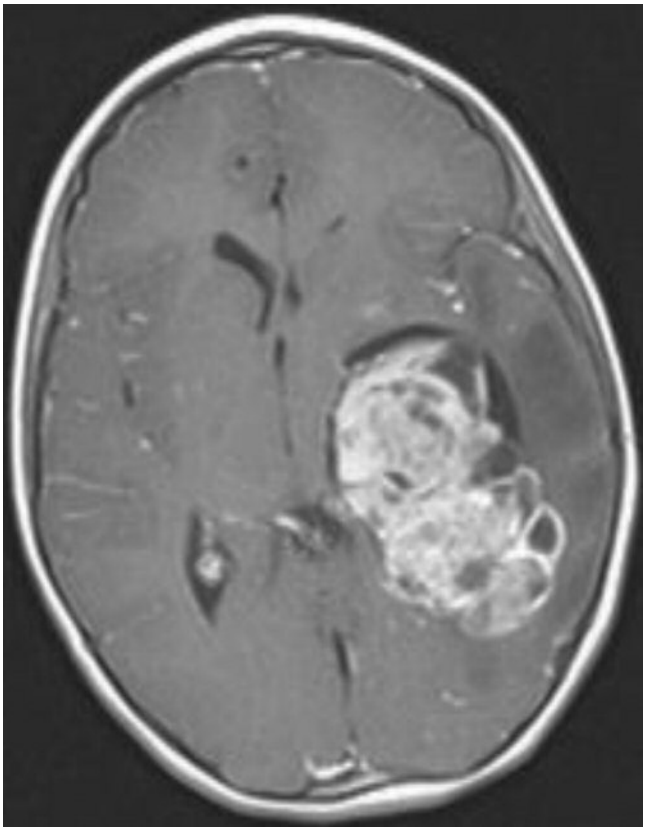

FIG. 2. Preoperative axial postcontrast T1-weighted MR image showing a large, demarcated solid-cystic mass (soap-bubble appearance) in the left temporooccipital ventricular horn.

Histopathological Findings. Histological examination revealed papillary areas with tumor cells clustered around the blood vessels forming perivascular pseudorosettes. Cytoplasmic processes were thick and short; the nuclei were located away from vessels and were oval and slightly irregular with coarse chromatin nodes. Vessels showed conspicuous hyaline thickening. The space between pseudorosettes was rarefied and occupied by spindle cells (Fig. 3). Solid and compact areas without papillary aspect were present, with the club-shaped or fusiform cells characteristic of an astrocytoma. No cellular atypia or vascular endothelial proliferation was found. There were small foci of necrosis and a brisk mitotic activity (MIB-1 index 14%). Immunohistochemical studies showed intense staining for GFAP, S100 protein, and EMA (Fig. 4).

Postoperative Course. The patient's postoperative course was uneventful. No adjuvant therapy was administrated after surgery. An MR imaging study obtained at 9 months' follow-up showed complete excision of the lesion (Fig 5). In T1-weighted images, both with and without contrast enhancement, a minimal hyperintensity in the posterior margin of surgical site was present, but it was considered to be without pathological significance.

Discussion

Astroblastoma is a rare glial tumor of uncertain origin. In the most recent World Health Organization classification of central nervous system tumors, astroblastoma is listed as a glial neoplasm of uncertain origin.[1,4] Some authors have proposed that it might originate from embryonic precursor cells transitional between astrocytes and ependymal cells.[21] Other authors have observed that the tumor cells recapitulate the structure of ependymal tanycytes, precursor cells that are normally found along the ependymal lining of the embryonic brain, and have proposed these cells as the origin of astroblastomas,[16,22] even though the lesions usually have no rela-

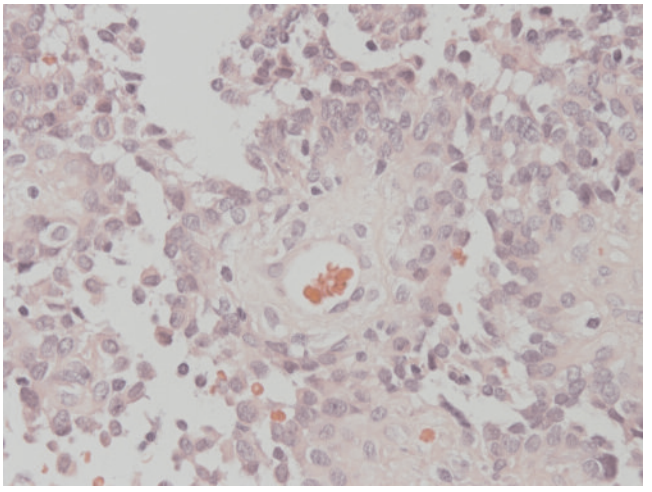

FIG. 3. Photomicrograph of a section of the resected lesion illustrating the thick and short cytoplasmic processes and epithelioid appearance of the tumor cells. H & E, original magnification × 320.

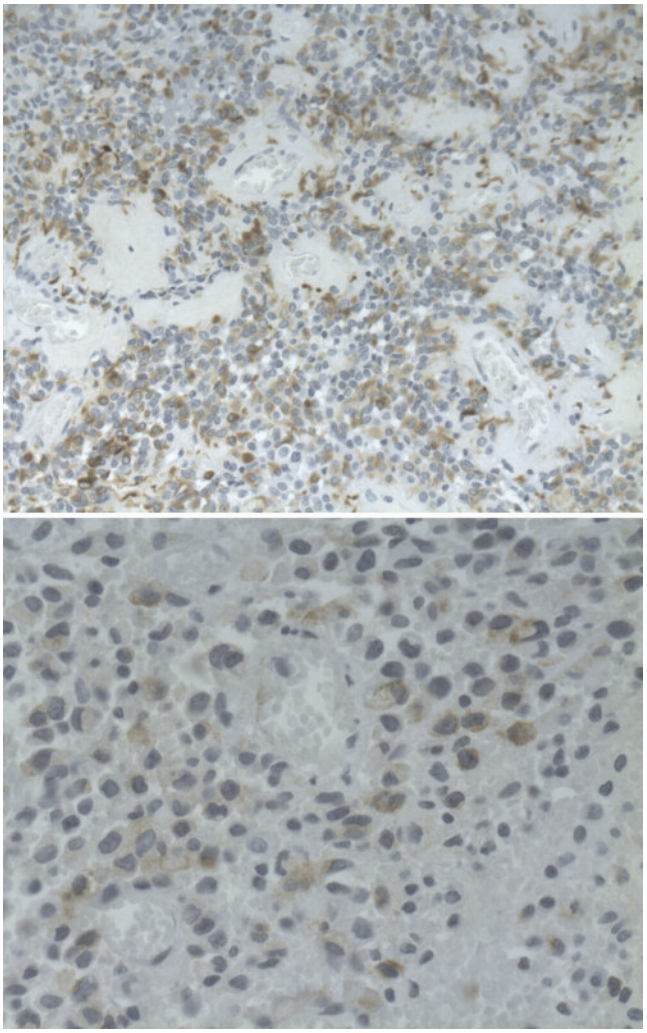

FIG. 4. Photomicrographs showing the results of immunohistochemical studies. The lesion stained intensely for GFAP *(upper)* and EMA *(lower)*. Original magnifications × 20 *(upper)* and 40 *(lower)*.

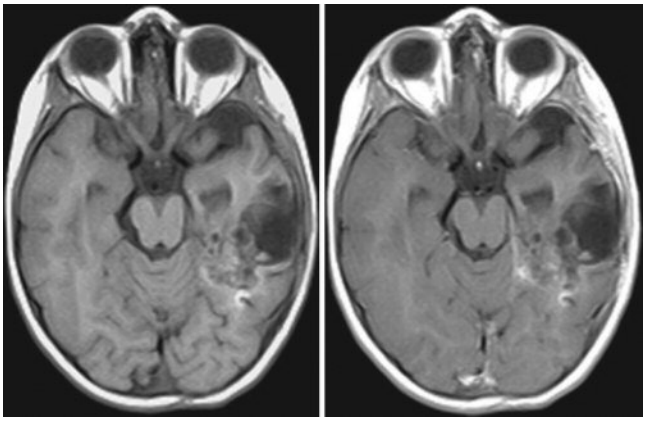

FIG. 5. Axial T1-weighted MR images obtained before *(left)* and after *(right)* gadolinium administration, showing complete excision of the lesion at 9 months after surgery. An area of minimal hyperintensity in the posterior margin of the surgical site is present in both the images but has no pathological significance.

tion to the ventricle. Histologically, astroblastoma has characteristic perivascular pseudorosettes with short, thick cytoplasmic processes that show blunt-ended foot plates and are attached to the basal lamina of blood vessels, frequently with hyalinization.[1,4,5] Two histological types have been recognized: 1) Differentiated astroblastoma is characterized by uniform perivascular arrangement of pseudorosettes, low to moderate numbers of mitotic figures, minimal cellular atypia, minimal to no vascular endothelial proliferation, and sclerosis of the vascular walls. 2) Anaplastic astroblastomas show cytological atypias, compact cellularity, perivascular cells with high mitotic rates, and hypertrophy of the vascular endothelium.[1,3,4] The other lesion that will most often have to be considered in the differential diagnosis is ependymoma. In contrast to astroblastoma, ependymoma is characterized by regular nuclei with delicate chromatin, thick perivascular cytoplasmic processes (broad and foot-plate in astroblastoma, radiating and tapering processes in ependymoma) and compact cellular intervascular architecture. Special caution should be taken in examination because astroblastoma focal features may be found in anaplastic astrocytoma or glioblastoma.

Astroblastomas are more often seen in older children and young adults than in older patients; these lesions tend to occur more often in female patients[3,26] and to have a supratentorial location. Patients present with nonspecific symptoms such as seizures, headaches, signs of increased intracranial pressure, and focal neurological deficit.[25,26] Hemorrhage has been described in only a few cases.[1,16,26] On MR images astroblastomas are generally well demarcated, with solid and cystic components, and have a "soap-bubble" appearance. This bubble-like appearance is probably due to the angioarchitecture of the mass causing signal voids.[18,20] The lesions usually appear hypointense to white matter on T1-weighted images and hyperintense on T2-weighted images. They show heterogeneous enhancement and frequent rim enhancement after gadolinium injection. Some degree of calcification is present. Relatively little peritumoral edema is reported, which may be related to the lack of local tumor infiltration into the surrounding brain tissue.[2,3]

Gross-total resection has the greatest impact on patient survival. Adjuvant treatment is recommended after resection for patients with anaplastic astroblastoma; in contrast, in differentiated tumors, recurrence is usually local, and adjuvant therapy is recommended only in cases of recurrent disease and is administered after repeated resection.[4,18,25,26]

Intraventricular astroblastomas are extremely rare. In our review of the literature we found that only 4 cases of intraventricular astroblastomas had been reported.[3,18] Two of the 4 patients were male. Two of the patients were 5 years old, 1 was 3, and 1 was 32. Their mean age, at the time of diagnosis, was 11 years. The tumor was located in the posterior third ventricle in 1 case, in the fourth ventricle in another, and in the lateral ventricles in 2 cases. Information about the histological type of the 2 masses located in the lateral ventricle are not available; the other 2 were well-differentiated lesions. Resection was attempted in all patients. One patient was lost to follow-up. Three patients underwent gross-total resection and radiotherapy after the initial operation. They had a mean event-free period of 63 months and a mean follow-up of 189 months (range 24–90 months).

The case we report involved a well-differentiated astroblastoma located in the left temporooccipital ventricular horn. Complete surgical resection was achieved as in the cases previously reported but no adjuvant therapy was given. At 9 months' follow-up there were no signs of recurrence.

Increased awareness of such a clinicopathological entity will probably result in an increase in the number of astroblastomas encountered by neurosurgeons. It is important to be aware of potential pitfalls in the neuropathological diagnosis and to differentiate astroblastoma from ependymoma, because of the different treatment protocols. Patients harboring an ependymoma are treated with surgery followed by radiotherapy regardless of tumor grade.[17,21]

Conclusions

Intraventricular astroblastomas are very rare. They are generally not infiltrative and are well circumscribed. Gross-total resection is the recommended therapy. The role of radiotherapy in these cases remains controversial. The rarity of the intraventricular localization seems to contradict the theory of a tanycyte origin in astroblastoma.

Acknowledgment

We thank Antonio Valerio Gerunda for technical assistance in preparing the figures.

References

1. Alaraj A, Chan M, Oh S, Michales E, Valyi-Nagy T, Hersonsky T: Astroblastoma presenting with intracerebral hemorrhage misdiagnosed as dural arteriovenous fistula: review of a rare entity. **Surg Neurol 67:**308–313, 2007
2. Baka JJ, Patel SC, Roebuck JR, Hearshen DO: Predominantly extraaxial astroblastoma: imaging and proton MR spectroscopy features. **AJNR Am J Neuroradiol 14:**946–950, 1993
3. Bell JW, Osborn AG, Salzam KL, Blaser SI, Jones BV, Chin SS: Neuroradiologic characteristics of astroblastoma. **Neuroradiology 49:**203–209, 2007
4. Bonnin JM, Rubinstein LJ: Astroblastomas: a pathological study of 23 tumors, with a postoperative follow-up in 13 patients. **Neurosurgery 25:**6–13, 1989
5. Brat DJ, Hirose Y, Cohen KJ, Feuerstein BG, Burger PC: Astroblastoma: clinicopathologic features and chromosomal abnormalities defined by comparative genomic hybridization. **Brain Pathol 10:**342–352, 2000
6. Cabrera-Zubizarreta A, Catón B, Martínez de Guereñu B, Larena-Iturbe JA, Ontañón JM, Catalán-Urribarrena G: [Low grade astroblastoma: pathological and magnetic resonance findings.] **Rev Neurol 34:**936–939, 2002 (Span)
7. Caroli E, Salvati M, Esposito V, Orlando ER, Giangaspero F: Cerebral astroblastoma. **Acta Neurochir (Wien) 146:**629–632, 2004
8. Han YM, Kim JT, Chung DS, Park JS: A case of astroblastoma. **J Korean Neurosurg Soc 40:**373–376, 2006
9. Hoang G, Sima AA, Rozdilsky B: Astroblastoma revisited: a report of three cases. **Acta Neuropathol (Berl) 70:**10–16, 1986
10. Huhn SL, Yung Y, Cheshier S, Harsh G, Ailles L, Weissman I, et al: Identification of phenotypic neural stem cells in a pediatric astroblastoma. **J Neurosurg 103 (5 Suppl):**446–450, 2005
11. Husain AN, Leestma JE: Cerebral astroblastoma: immunohistochemical and ultrastructural features. Case report. **J Neurosurg 64:**657–661, 1986
12. Jay V, Edwards V, Squire J, Rutka J: Astroblastoma: report of case with ultrastructural, cell kinetic, and cytogenetic analysis. **Pediatr Pathol 13:**323–332, 1993
13. Kaji M, Takeshima H, Nakazato Y, Kuratsu J: Low-grade astroblastoma recurring with extensive invasion. **Neurol Med Chir (Tokyo) 46:**450–454, 2006
14. Kim BS, Kothbauer K, Jallo G: Brainstem astroblastoma. **Pediatr Neurosurg 40:**145–146, 2004
15. Kim DS, Park SY, Lee SP: Astroblastoma: a case report. **J Korean Med Sci 19:**772–776, 2004
16. Kubota T, Sato K, Arishima H, Takeuchi H, Kitai R, Nakagawa T: Astroblastoma: immunohistochemical and ultrastructural study of distinctive epithelial and probable tanycytic differentiation. **Neuropathology 26:**72–81, 2006
17. McLaughlin MP, Marcus RB Jr, Buatti JM, McCollough WM, Mickele JP, Kedar A, et al: Ependymoma: results, prognostic factors and treatment recommendations. **Int J Radiat Oncol Biol Phys 40:**845–850, 1998
18. Navarro R, Reitman AJ, de León GA, Goldman S, Marymont M, Tomita T: Astroblastoma in childhood: pathological and clinical analysis. **Childs Nerv Syst 21:**211–220, 2005
19. Pizer BL, Moss T, Oakhill A, Webb D, Coakham HB: Congenital astroblastoma: an immunohistochemical study. Case report. **J Neurosurg 83:**550–555, 1995
20. Port JD, Bral DJ, Burger PC, Pomper MG: Astroblastoma: radiologic-pathologic correlation and distinction from ependymoma. **AJNR Am J Radiol 23:**243–247, 2002
21. Robertson PL, Zelter PM, Boyett JM, Rorke LB, Allen JC, Geyer JR, et al: Survival and prognostic factors following radiation therapy and chemotherapy for ependymomas in children: a report of the Children's Cancer Group. **J Neurosurg 88:**695–703, 1998
22. Rubinstein LJ, Herman MM: The astroblastoma and its possible cytogenic relationship to the tanycyte. An electron microscopic, immunohistochemical, tissue- and organ-culture study. **Acta Neuropathol (Berl) 78:**472–483, 1989
23. Sener RN: Astroblastoma: diffusion MRI, and proton MR spectroscopy. **Comput Med Imaging Graph 26:**187–191, 2002
24. Shuangshoti S, Mitphraphan W, Kanvisetsri S, Griffiths L, Navalitloha Y, Pornthanakasem W, et al: Astroblastoma: report of a case with microsatellite analysis. **Neuropathology 20:**228–232, 2000
25. Thiessen B, Finlay J, Kulkarni R, Rosenblum MK: Astroblastoma: does histology predict biologic behavior? **J Neurooncol 40:** 59–65, 1998
26. Tumialán LM, Brat DJ, Fountain AJ, Barrow DL: An astroblastoma mimicking a cavernous malformation: case report. **Neurosurgery 60:**E569–E570, 2007

Manuscript submitted May 23, 2007.
Accepted October 17, 2007.

Address correspondence to: Luca Denaro, M.D., Department of Neurosurgery, Catholic University, Largo A. Gemelli 8, Roma, Italy. email: lucadenaro@hotmail.com.