A Rare Pediatric Tumor: Supratentorial High-Grade Astroblastoma Presenting as a huge Mass

Deniz Sürmeli[1] Boran Urfalı[2] Tümay Özgür[1]

[1] Department of Pathology, Faculty of Medicine, Hatay Mustafa Kemal University, Hatay, Turkey
[2] Department of Neurosurgery, Faculty of Medicine, Mustafa Kemal University, Hatay, Turkey

Address for correspondence Tümay Özgür, MD, Department of Pathology, Faculty of Medicine, Hatay Mustafa Kemal University, Hatay, Turkey (e-mail: ozgurtumay@yahoo.com).

Indian J Neurosurg 2024;13:265–270.

Abstract

Background Astroblastoma is a rare neuroepithelial tumor of unknown origin, usually seen in children and young adults. It is usually localized to the cerebral hemisphere. Computed tomography and magnetic resonance imaging show a well-demarcated, contrast-enhancing mass with a cystic area. Characteristic histological findings are perivascular pseudorosette formation and frequent vascular hyalinization. The presented case is a 3.7-month-old female patient diagnosed with high-grade astroblastoma.

Case Presentation We report the case of a 3.7-year-old female patient admitted to the neurosurgery clinic with strabismus for 25 days. Magnetic resonance imaging revealed a contrast-enhancing mass that contained cystic and necrotic areas. The tumor mass has been totally resected and histological examination combined with immunohistochemical study confirmed the diagnosis of high-grade astroblastoma.

Keywords
- astroblastoma
- brain
- neuroepithelial

Introduction

Astroblastoma is a rare neuroepithelial tumor of unknown origin and accounts for 0.45 to 2.8% of all neuroglial tumors, almost seen in children and young adults. It is usually localized to the cerebral hemispheres. Computed tomography (CT) and magnetic resonance imaging (MRI) show a well-demarcated, contrast-enhancing mass with cystic areas.[1,2] Characteristic histological findings are perivascular pseudorosette formation and frequent vascular hyalinization. Perivascular pseudorosettes in astroblastoma have short and thick cytoplasmic processes.[3]

Case Report

Our case was a 3.7-year-old female patient admitted to the neurosurgery clinic with strabismus for 25 days. Magnetic resonance imaging (MRI) revealed a contrast-enhancing mass containing solid and cystic areas that compressed the surrounding brain tissue, the third ventricle, and the left lateral ventricle (►**Fig. 1a, 1b**). The tumor was totally resected. Evaluation of routine hematoxylin–eosin (H&E) sections revealed proliferation of polar cells having abundant eosinophilic cytoplasm and eccentrically located nuclei with short stout cytoplasmic processes surrounded by blood vessels and composing of perivascular pseudorosettes (►**Fig. 2a, 2b**). Vascular and stromal hyalinization accompanied the pseudorosette formations. There were also cystic degenerative changes and foam cell infiltration. Besides these findings, rhabdoid differentiation of tumor cells was observed. There were foci of pseudopalisating necrosis and atypical mitosis. In the immunohistochemical study, tumor cells were diffuse and positive for GFAP and vimentin, focally positive for NSE, S-100, and CD56. There was also focal EMA positivity in the pseudorosettes. Ki67 proliferative activity was determined as 30 to 40%. The case was evaluated as "high-grade astroblastoma" with histomorphological and immunohistochemical findings.

article published online
February 23, 2023

DOI https://doi.org/10.1055/s-0042-1744245.
ISSN 2277-954X.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

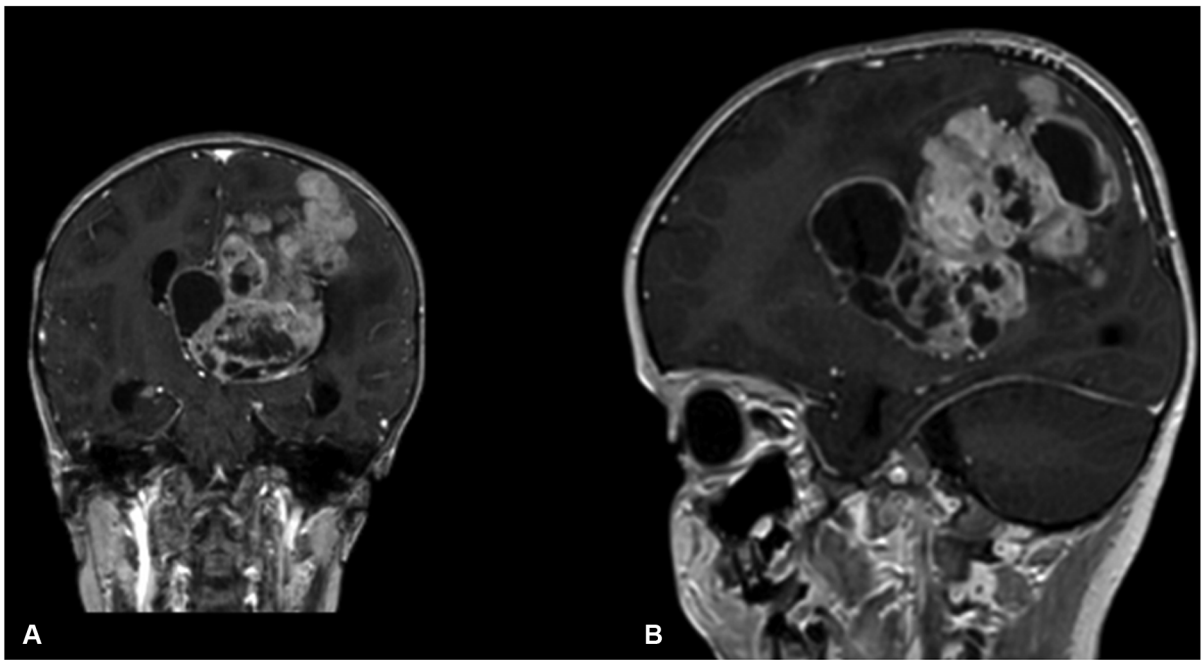

Fig. 1 Radiologic findings revealed on MRI T1-weighted contrast-enhanced coronal and sagittal images and T2-weighted axial image showing a well-circumscribed solid and cystic lesion with little perilesional edema. The solid and cystic lesion was hypo- to hyper-intense to gray matter on T1- weighted contrast-enhanced image (**A, B**).

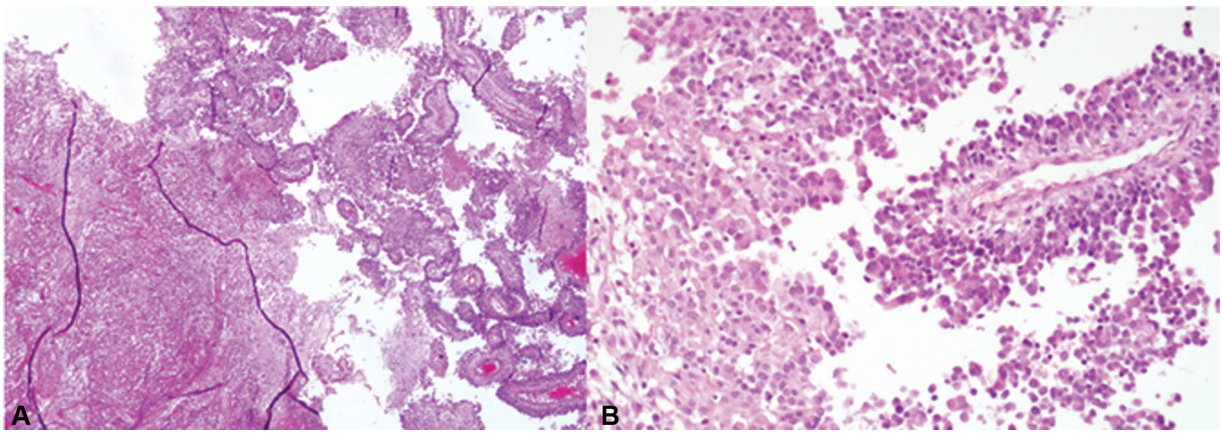

Fig. 2 Histopathologic examination; The tumor was composed of polar cells, having eosinophilic cytoplasm and eccentrically placed nuclei with short stout cytoplasmic processes anchored to the blood vessels (**A, B**; H&E ×40, 100).

The patient was followed up in the pediatric intensive care unit, she had been consulted to the pediatric oncology clinic for adjuvant therapy protocols. No residual tumoral tissue was observed in the postoperative contrast-enhanced brain MRI (►**Fig. 3**).

Discussion

Astroblastomas are currently classified as "other neuroepithelial tumors" according to the World Health Organization (WHO 2016) and this tumor is rare.[4] There are a few cases of astroblastoma in the literature (►**Table 1**). Astroblastomas are more common in the two age ranges; prominent peak between the ages of 5 and 10 years, the second peak between the ages of 21 and 30 years.[2] Clinical signs and symptoms depend on the location and size of the tumor. Vomiting, headache, seizures, and focal neurological deficits are the most common symptoms.[5]

Astroblastoma shows radiologically characteristic features. This tumor usually occurs as a well-limited, superficial mass. Astroblastoma is typically seen in the cerebral hemispheres, but corpus callosum, cerebellum, brain stem, optic nerve, and cauda equina are other locations.[2] The tumor is typically a heterogeneous, contrast-enhancing, well-circumscribed mass with a solid cystic component with little vasogenic edema. The solid part gives a bubbly appearance.[3] As reported in the

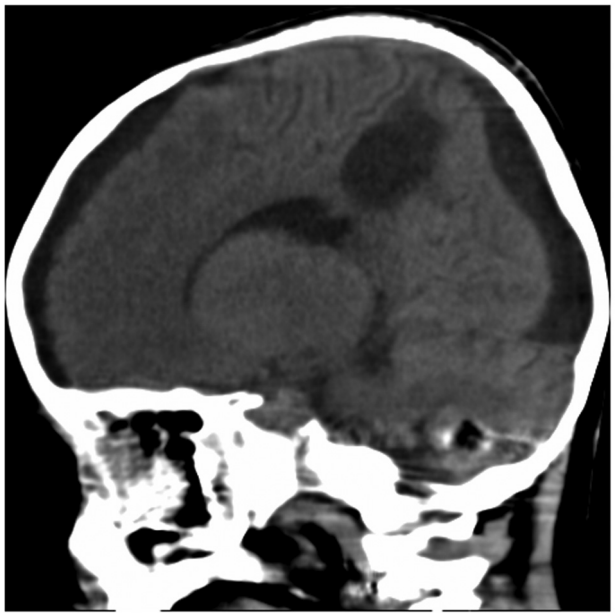

Fig. 3 No residual tumoral tissue was observed in the postoperative contrast-enhanced brain MRI.

literature, this case also emerged as a contrast-enhancing solid mass containing a cystic component.

Differential diagnoses of astroblastoma are anaplastic astrocytoma, glioblastoma, and ependymoma. Histologically, these are composed of a monotonous population of spindle-shaped cells with coarse chromatin. These cells make pseudorosettes around blood vessels. Glioblastoma and anaplastic astrocytoma also form pseudorosettes, but in these tumors, it is focal, while in astroblastoma, these are distributed all over the tumor tissue.[6]

Histologically, hallmark features of astroblastomas are the presence of perivascular pseudorosettes and prominent perivascular hyalinization. These histological features are similar to ependymoma.[7] The presence of pseudorosettes with shorter thick cytoplasmic processes and the hyalinized vessels separate the astroblastoma from the ependymoma.[8]

These tumors are immunopositive with GFAP EMA pancytokeratin vimentin and S100. They are negative for *IDH1/2* and *TP53* mutations. Vimentin and S100 are more characteristic of astrocytic origin.[6] In our case, there were pseudorosettes surrounded by short thick cytoplasmic

Table 1 Reviewed patients with astroblastoma (epidemiological and grading characteristics)

NUMBER	REFERENCE	AGE/GENDER	LOCATION	GRADE
1.	Port et al,2002;23:243–7.	30; M 42;F 24;F 5;F 3;F 15;F	FL TL Supratentorial Corpus collosum	LG LG LG HG HG HG
2.	Sugita Y, Terasaki M, Shigemori M, et al Astroblastoma with unusual signet-ring-like cell components: a case report and literature review. Neuropathology. 2002;22:200–5.	33;F	FL	LG
3.	Cabrera-Zubizarreta A, Catón B, Martínez de Guereñu B, et al Low grade astroblastoma: pathological findings and on magnetic resonance. Rev Neurol. 2002;34:936–9.	18;F	FL	LG
4.	Kim DS, Park SY, Lee SP. Astroblastoma: A case report. J Korean Med Sci. 2004;19:772–6.	15;F	FL	LG
5.	Caroli E, Salvati M, Esposiro V, et al Cerebral astroblastoma. Acta Neurochir. 2004;146:629–33.	30;M	TL	HG
6.	Kaji M, Takeshima H, Nakazato Y, et al Low-grade astroblastoma recurring with extensive invasion–case-report. Neurol Med Chir. 2006;46:450–4.	17;M	Frontal operculum	LG
7.	Lau PP, Thomas TM, Lui PC, et al 'Low-grade' astroblastoma with rapid recurrence: a case report. Pathology. 2006;38:78–80.	21;F	Parietal	LG
8.	Miranda P, Lobato RD, Cabello A, Gómez PA, Martínez de Aragón A. Complete surgical resection of high-grade astroblastoma with long time survival: case report and review of the literature. Neurocirugia (Astur). 2006;17[1]:60–3	42;F	FL	
9.	Hata N, Shono T, Yoshimoto K, et al An astroblastoma case associated with loss of hetreozygosty on chromosome 9p. J Neuro-Oncol. 2006;80:69–73.	16;F	PL	LG

(*Continued*)

Table 1 (*Continued*)

NUMBER	REFERENCE	AGE/GENDER	LOCATION	GRADE
10.	Kubota T, Sato K, Arishima H, et al Astroblastoma: immunohistochemical and ultrastructural study of distinctive epithelial and probable tanycytic differentiation. Neuropathology. 2006;26:72–81.	8;F	FPL	HG
11.	Alaraj A, Chan M, Oh S, Michals E, Valyi-Nagy T, Hersonsky T. Astroblastoma presenting with intracerebral hemorrhage misdiagnosed as dural arteriovenous fistula: review of a rare entity. Surg Neurol. 2007;67:308–13.	33;M	TL	HG
12.	Eom K-S, Kim JM, Kim T-Y. A Cerebral Astroblastoma Mimicking an Extra-axial Neoplasm. J Korean Neurosurg Soc. 2008;43:205–8.	20;F	TL	HG?
13.	Fathi AR, Novoa E, El-Koussy M, et al Astroblastoma with rhabdoid features and favorable long-term outcome: report of a case with a 12-year follow-up. Pathol Res Pract. 2008;204:344–51.	53;M	PL	LG
14.	Unal E, Koksal Y, Vajtai I, Toy H, Kocaogullar Y, Paksoy Y. Astroblastoma in a child. Childs Nerv Syst. 2008;24:165–8.	4;M	FPL	HG
15.	Salvati M, D'Elia A, Brogna C, et al Cerebral astroblastoma: analysis of six cases and critical review of treatment options. J Neuro-Oncol. 2009;93:369 78.	30;M 27;F 39;F 43;F 33;M 50;F	TL POL TL FL Rolandic area OL	HG LG LG LG HG HG
16.	Kemerdere R, Dashti R, Ulu MO, et al Supratentorial high grade astroblastoma: report of two cases and review of the literature. Turk Neurosurg. 2009;19:149–53.	6;F 7;F	FPL PL	HG HG
17.	Mastrangelo S, Lauriola L, Coccia P, et al Two cases of pediatric high-grade astroblastoma with different clinical behavior. Tumori. 2010;96:160–3.	21;F 12;F	FPL TL	HG HG
18.	Bergkåsa M, Sundstrøm S, Gulati S, Torp SH. Astroblastoma – a case report of a rare neuroepithelial tumor with complete remission after chemotherapy. Clin Neuropathol. 2011;30[6]:301–6.	50;F	FL	HG
19.	Bhattacharjee S, Pulligopu AK, Uppin MS, et al Astroblastoma with bone invasion. Asian J Neurosurg. 2011;6:113–5.	4;F	POL	HG
20.	Agarwal V, Mally R, Palande DA, Velho V. Cerebral astroblastoma: A case report and review of literature. Asian J Neurosurg. 2012;7:98–100.	12;F	PL	LG
21.	Khosla D, Yadav BS, Kumar R, Agrawal P, Patel NKFD, Sharma SC. Pediatric Astroblastoma: A Rare Case with a Review of the Literature. Pediatr Neurosurg. 2012;48:122–5.	11;F	FPL	HG
22.	Nasit JG, Trivedi P. Recurrent low-grade astroblastoma with signet ring-like cells and high proliferative index. Fetal Pediatr Pathol. 2013;32:284–92.	10;F	FPL	LG
23.	De la Garma VH, Arcipreste AA, Vazquez FP, Aguilar RR, Castruita UO, Guerra RM. High-grade astroblastoma in a child: Report of one case and review of literature. Surg Neurol Int. 2014;5:111.	9;F	FPL	HG
24.	Janz C, Buhl R. Astroblastoma: Report of two cases with unexpected clinical behavior and review of the literature. Clin Neurol Neurosurg. 2014;125:114–24.	16;F 24;F	POL TL	LG with transition toHG HG

Table 1 (*Continued*)

NUMBER	REFERENCE	AGE/GENDER	LOCATION	GRADE
25.	Singh DK, Singh N, Singh R, Husain N. Cerebral astroblastoma: A radiopathological diagnosis. J Pediatr Neurosci. 2014;9:45–7.	12;F	PL	LG
26.	Yao K, Wu B, Xi M, Duan Z, Wang J, Qi X. Distant dissemination of mixed low-grade astroblastoma-arteriovenous malformation after initial operation: a case report. Int J Clin Exp Pathol. 2015;8[6]:7450–6.	36;M	OTL	LG
27.	Narayan S, Kapoor A, Singhal MK, Jakhar SL, Bagri PK, Rajput PS, et al Astroblastoma of cerebrum: A rare case report and review of literature. J Can Res Ther. 2015;11:667.	16;M	OTL	LG
28.	Singla N, Dhandapani SS, Kapoor A, Chatterjee D, Vashishta Yeo RK. Hemorrhage in astroblastoma: An unusual manifestation of an extremely rare entity. J Clin Neurosci. 2016;25:147–50.	30;F 11;M	FL FPL	LG HG
29.	Yuzawa S, Nishihara H, Tanino M, Kimura T, Moriya J, Kamoshima Y, Nagashima K, Tanaka S. A case of cerebral astroblastoma with rhabdoid features: a cytological, histological, and immunohistochemical study. Brain Tumor Pathol. 2016;33[1]:63–70.	18;F		LG
30.	Yeo JJY, Low YYS, Putti TC, Koh KMR. Adult intraventricular astroblastoma. Singap Med J. 2016;57[1]:53–4.	35;M	Lateral ventricle	LG
31.	Hammas et al.[2]	8;F	OTL	LG
32.	Bhalerao, Sagar, Rajnish Nagarkar, and Aditya Adhav. "A case report of high-grade astroblastoma in a young adult." CNS oncology 8.1 (2019): CNS29.	18;F	FPL	HG
33.	Sarper, Binnaz, et al. "Malignant Astroblastoma." Balkan medical journal 37.4 (2020): 224.	23;M		HG
34.	Dey et al[6]	38;F	TL	HG
35.	Our case	3;F	Lateral ventricle	HG

Abbreviations: F, female; FL, frontal lobe; FPL, frontoparietal lobe; HG, high grade; LG, low grade; M, male; OL, occipital lobe; OTL, occipitotemporal lobe; PL, parietal lobe; POL, parieto-occipital lobe; TL, temporal lobe.

processes. In these pseudorosettes, widespread positive staining was observed with GFAP, S100, and vimentin, while focal positive staining was observed in tumor cells with EMA. Rhabdoid differentiation of tumor cells in our case was an interesting finding like the case of Yuzawe et al, which raised the question of atypical rhabdoid/teratoid tumor in differential diagnosis but INI-1 immunostaining was non-specifically positive and remained apart.[9] Bonnin and Rubinstein divided astroblastomas into two groups as low-grade (well-differentiated) and high-grade (anaplastic). Those with a perivascular pattern, low cellular atypia, low-moderate mitotic activity, and pronounced sclerosis of the vascular walls were evaluated as low-grade astroblastoma. High-grade astroblastoma is more aggressive, such as high cellularity, cytological atypia, a high mitotic rate, pseudopalisading necrosis, and non-sclerosis of vascular walls, and microvascular hyperplasia.[10] In previous reports, Ki67 proliferation activity was 0.5 to 8% in low-grade astroblastoma and 4 to 20% in high-grade astroblastoma.[9] This case also had pseudopalisating necrosis and atypical mitosis. Also, Ki67 proliferation activity was 30 to 40%. Therefore, we reported this case as a high-grade astroblastoma.

The standard treatment of astroblastomas is complete resection.[10] There was also no evidence of tumor recurrence for 2 months after resection in our case.

In conclusion, astroblastoma is a rare primary brain tumor and should be remembered in the differential diagnosis of tumors with ependymal morphology at supratentorial intra-axial localization in children and young adults.

Conflict of Interest
None declared.

References

1 Port JD, Brat DJ, Burger PC, Pomper MG. Astroblastoma: radiologic-pathologic correlation and distinction from ependymoma. Am J Neuroradiol 2002;23(02):243–247
2 Hammas N, Senhaji N, Alaoui Lamrani MY, et al. Astroblastoma–a rare and challenging tumor: a case report and review of the literature. J Med Case Reports 2018;12(102):1–10

3 Barakat MI, Ammar MG, Salama HM, Abuhashem S. Astroblastoma: case report and review of literature. Turk Neurosurg 2016;26(05):790–794
4 Aldape KD, Rosenblum MK. Astroblastoma. In: Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, eds. WHO Classification of Tumors of the Central Nervous System, 4th ed. Lyon: IARC; 2007:88–89
5 Agarwal V, Mally R, Palande DA, Velho V. Cerebral astroblastoma: a case report and review of literature. Asian J Neurosurg 2012;7 (02):98–100
6 Dey B, Dutta S, Saurabh A, Raphael V, Khonglah Y. Cerebral astroblastoma: a rare tumor. Cureus 2021;13(07):e16323
7 Burger P, Scheithauer B. Tumors of the Central Nervous System. Atlas of Tumor pathology, 3rd series, fascicle 10 Washington, DC: Armed Forces Institute of Pathology; 1994:146–148
8 Eom KS, Kim JM, Kim TY. A cerebral astroblastoma mimicking an extra-axial neoplasm. J Korean Neurosurg Soc 2008;43(04):205–208
9 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 2006;26(01):72–81
10 Bonnin JM, Rubinstein LJ. Astroblastomas: a pathological study of 23 tumors, with a postoperative follow-up in 13 patients. Neurosurgery 1989;25(01):6–13