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Case Reports & Case Series

Angiocentric glioma: Literature review and first case in Brazil

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A B S T R A C T

Background: Angiocentric glioma (AG) was recently added in the World Health Organization (WHO) classification of tumors (2007) after two case reports published in 2005 as neoplasm of the central nervous system (CNS). AG predominantly affects children and young adults, and very often patients presenting with difficult to control epilepsy. Since its initial description, AG has been considered a rare entity with fewer than 100 cases reported hitherto. We report the first case of AG in Brazil, to our knowledge, in a male patient presenting refractory seizures, which neuroimaging results evidenced compatible lesion.

Case description: Male patient, 23-year-old, with a history of long-standing headache and episodes of tonic-clonic seizures. His clinical evaluation and physical examination were normal. According to the MRI were evidenced tumefactive lesions next to the pars opercularis of the inferior frontal gyrus and part of the corresponding subcentral gyrus. The patient was submitted to total resection of the lesion. Patient was kept awaken with intraoperative monitoring and using left frontotemporal approach. Three days after surgery the patient was discharged, without neurological deficits and no seizure in the postoperative period. The anatomical pathology exam was compatible with AG.

Conclusions: Angiocentric Glioma is a rare neoplasm mostly present in patients with difficult to control epilepsies. Tumoral resection provides broad control of symptoms, besides reducing recurrence and the progression of the disease, leading to an extremely favorable prognosis.

1. Introduction

Angiocentric glioma (AG) was recently added in the World Health Organization (WHO) classification of tumors (2007) after two case reports published in 2005 as central nervous system (CNS) neoplasm [1–3]. At first, this entity was classified as “other neuroepithelial tumors” [4], along with astroblastomas and chordoid tumors of the third ventricle. Later, in 2016, it was reclassified as “Other gliomas” [5], grade I, due to its benign behavior and the possibility of surgical resection [6]. AG predominantly affects children and young adults, and very often in patients presenting with difficult to control epilepsy [1,6,7]. The average age of initial diagnosis is 16 years with male predominance [8–10]. Since Wang et al. [2], with its initial description, AG has been considered a rare entity with fewer than 100 cases reported hitherto. We report the first case of AG in Brazil, to our knowledge, in a male patient, 23 year-old, presenting refractory seizures, which neuroimaging results evidenced compatible lesion.

2. Case report

Male patient, 23-year-old, with a history of long-standing headache and episodes of tonic-clonic seizures since January 2017. His clinical evaluation and physical examination were normal. According to the brain magnetic resonance imaging (MRI) were evidenced tumefactive lesions next to the pars opercularis of the inferior frontal gyrus and part of the corresponding subcentral gyrus. Also, T1 weighted sequences with hypersignal next to the cortico- subcortical aspect and suppression on FLAIR sequence of hypersignal on T2 weighted sequences (Fig. 1).

The magnetic resonance spectroscopy (MRS) evidenced N-Acetylaspartate peak reduction (denoting loss of neuronal population/viability) and discreet increase in the peak of choline, suggesting a possible extreme turnover of the membrane cells. In addition, the perfusion study showed reduced relative cerebral blood volume (rCBV) in relation to the contralateral normal appearing white matter (Fig. 2).

During the investigation, the patient underwent functional MRI of the brain, which showed strong activation of the Broca's area next to the pars opercularis of the left inferior frontal gyrus, with correspondence

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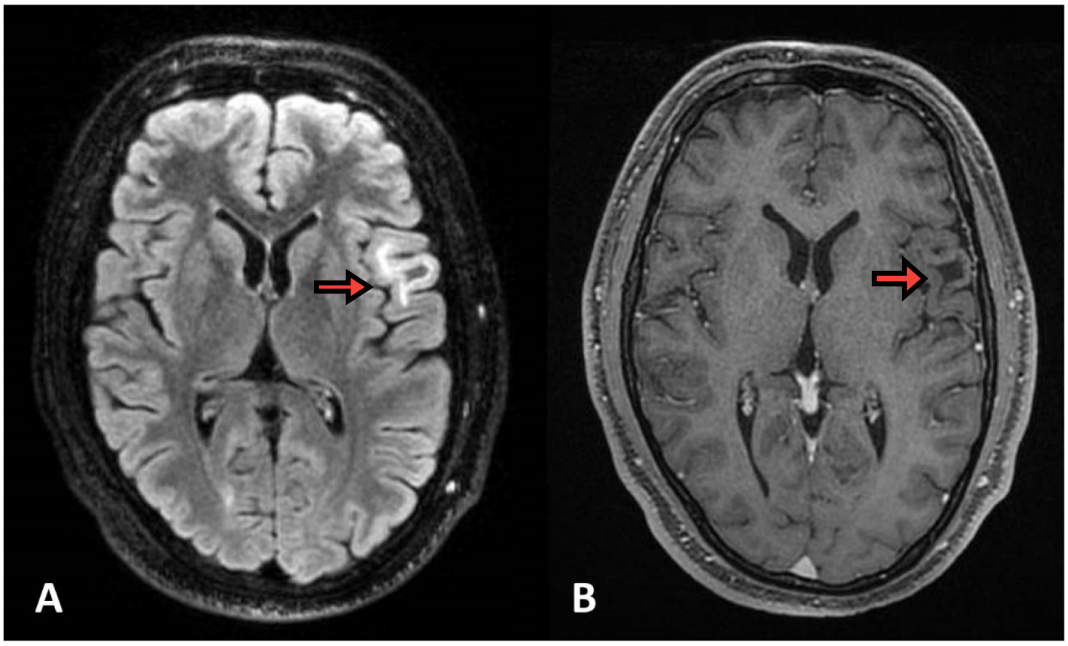

Fig. 1. Expansive lesion to the pars opercularis with suppression on FLAIR and T1WI sequence without paramagnetic contrast enhancement.

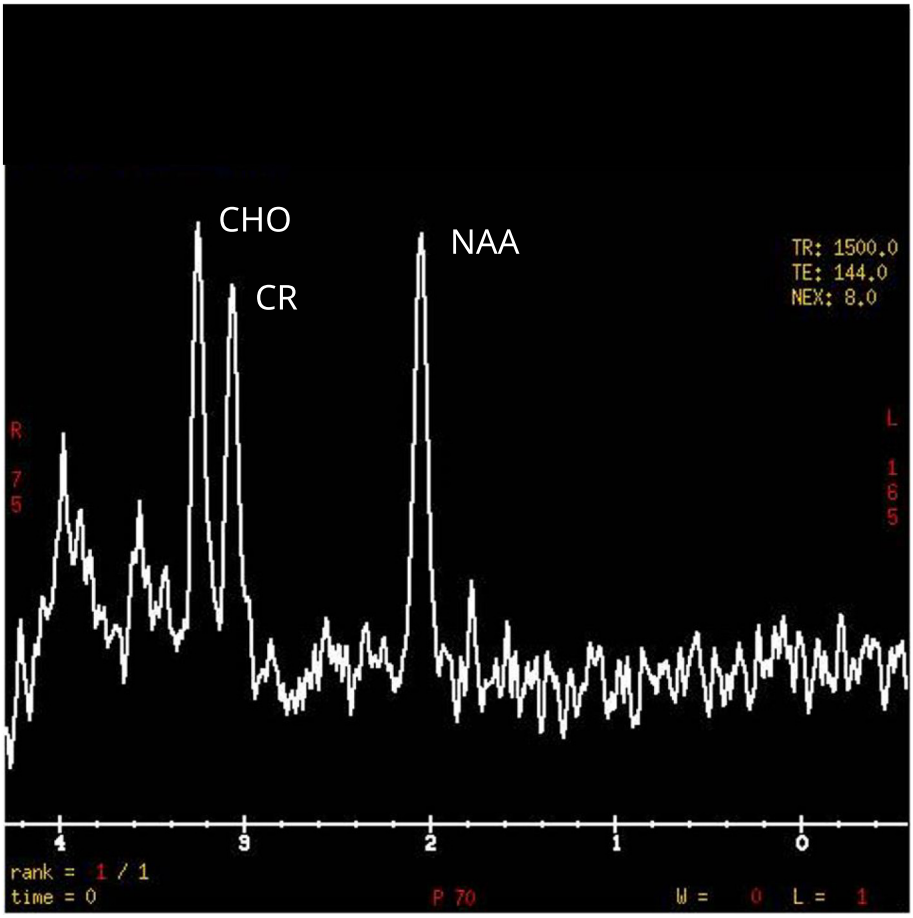

Fig. 2. N-Acetylaspartate peak reduction and choline peak.

in the contralateral area. Activations of the Wernicke's area were also observed along the posterior aspect of the left superior temporal gyrus, with small contralateral correlation. Therefore, the possibility of lateralization of language to the left is enhanced with a probable contralateral cognitive reserve (Fig. 3).

The patient was submitted to total resection of the lesion. Patient was kept awaken with intraoperative monitoring and using left fronto-temporal approach. Three days after surgery the patient was

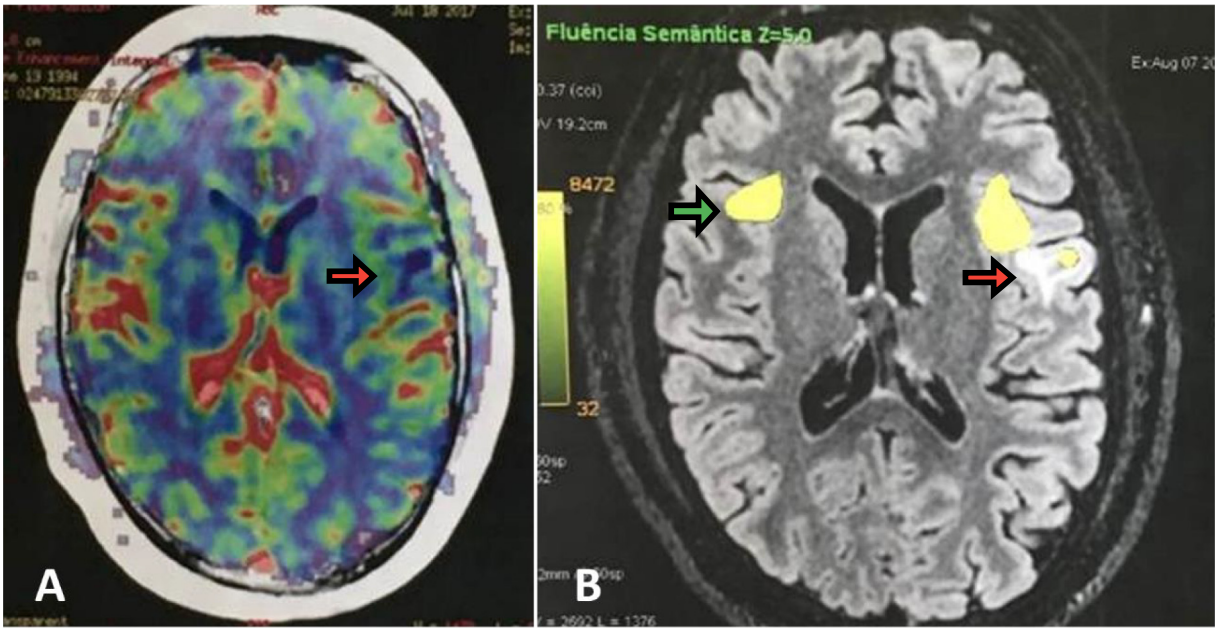

Fig. 3. A. Perfusion study with reduced relative cerebral blood volume; B. fMRI showing activation in the Broca's area of the left inferior frontal gyrus, with correspondence in the contralateral area.

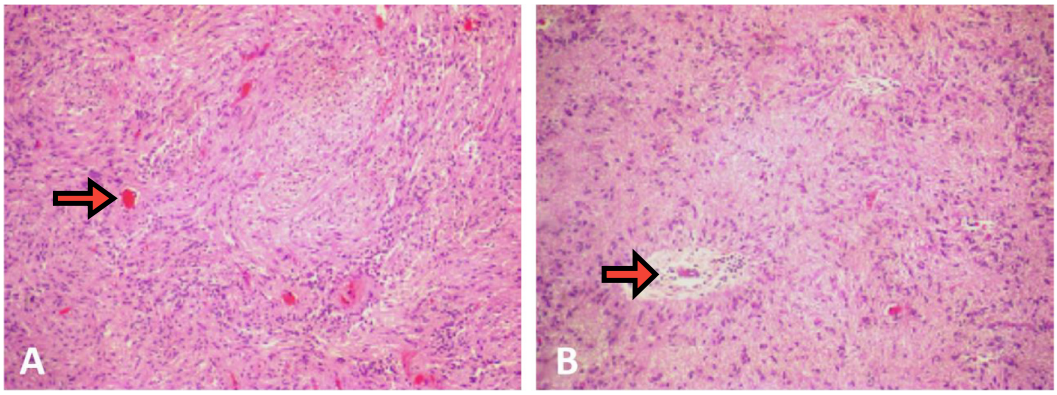

Fig. 4. Hematoxylin and eosin staining. Spindle tumor cells in sparse perivascular and multidirectional pseudorosettes. Original magnification x200.

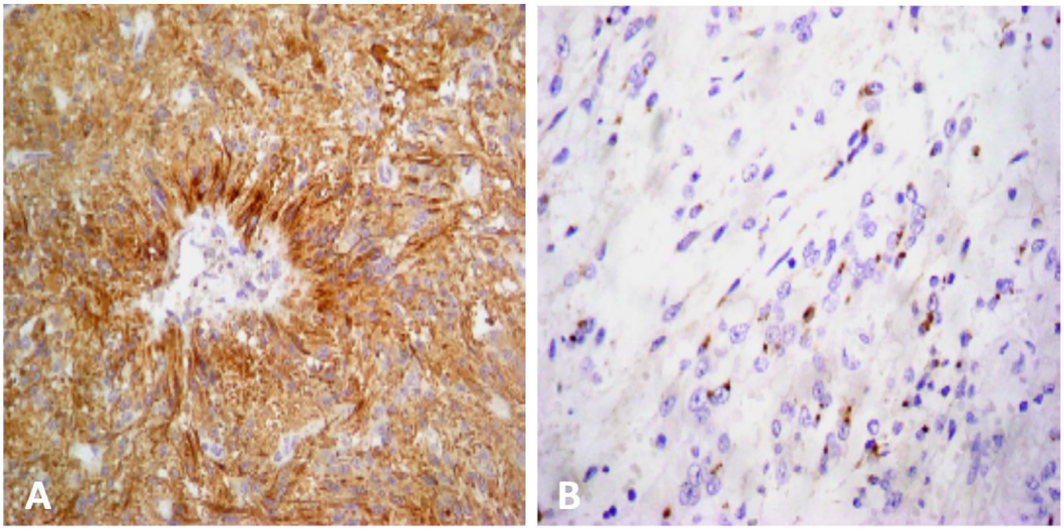

Fig. 5. A. Positive glial fibrillary acid protein (GFAP). B. Positive epithelial membrane antigen (EMA), "dot" pattern. Original magnification ×400.

discharged, without neurological deficits and no seizure in the post-operative period. The anatomical pathology exam was compatible with AG (Figs. 4, 5).

3. Discussion

This is the first case of AG reported in Brazil, which is clinically presented as difficult to control epilepsy, and described as typical in this type of lesion as well as in low grade injuries [8]. Regarding to the epidemiology, these lesions occur predominantly in children and young adults, who tend to present the main clinical aspect of drug-resistant epilepsies [1,6,7]. The average age of initial diagnosis is 16 years and there is some prevalence in males (1.5:1) [8–10]. After the initial description by Wang et al. [2], it is still considered a rare entity, having been reported fewer than 100 cases until the moment.

In this case reported, the epidemiological aspects were according to the literature, young adult male with a history of seizures that are difficult to be controlled. The initial description of this tumor reports slow growth, in the vast majority supratentorial located, mainly in the temporal lobe, and more specifically in the hippocampal, frontal and parietal region [1,6,7]. Ampie et al. [8] demonstrated that according to the location, the most common is the temporal lobe (39%), followed by frontal lobe (15%) and parietal (30%), and the less common is the thalamus (1%). Until this moment only 3 cases of brainstem lesions were reported [6,8]. Our patient presented a lesion located in the frontal lobe, more specifically in the pars opercularis of the inferior frontal gyrus and in part of the correspondent subcentral gyrus. Superficial lesions are more prone to epileptogenic events when compared to deeper tumors [8]. According to Ampie et al. [8], 86% of the patients presented seizures, followed by headache (8%), and these same symptoms were presented by our case. Other clinical presentations reported in the literature include ataxia, otalgia, dizziness, paresis and visual changes [8,9,11].

From the radiological point of view, they are similar to low-grade gliomas.

[1,7]. On MRI, lesions are infiltrative, well-defined, hypointense on T1WI and hyperintense on T2WI and FLAIR sequences, without paramagnetic contrast enhancement [10,12]. Some cases present increased ribbon-like signal on T1WI or cysts [10]. During neuroimaging examination of our patient was found cortico- subcortical hypersignal on T1WI and suppression on FLAIR sequence, unlike most of the cases in the literature, besides the increased signal on T2WI. Although scarce, the literature reports some cases with MRS patterns similar to those found in our case, with peak of the choline and reduction of N- acetylaspartate [13].

AG are histologically known as bipolar monomorphic population of elongated spindle-shaped, tending to assume a perivascular growth in a pattern known as “pseudo-rosette”, similar to ependymomas and astroblastomas [1,9]. Also, they show immunoreactivity to the glial fibrillary acid protein (GFAP), S100 protein and vimentin, but not to the neuronal antigens [1,6]. AG is positive to the epithelial membrane antigen (EMA), dot-like with microlumen appearance [9]. Usually, the vascular proliferation and necrosis do not to occur in this histological type, as well as the mitosis, which are rare or even absent. This low proliferative rate, confirmed by the rates of reaction to antigen Ki-67 ranging from < 1 to 5% [2,3,14], reinforces the benign aspect of the tumor. However, some studies reported cases with high rate of proliferation of tumor cells, recurrence, mixed histology and even transformation to Anaplastic Astrocytoma (WHO grade III) demonstrating the malignant potential of AG [9,15]. The immunohistochemical findings of our patient were according to the literature, with positive GFAP, S100 and EMA with “dot” pattern, and < 1% Ki67 positive.

The cornerstone of the low-grade glioma treatment is surgical resection [11]. The procedure tends to be curative, usually resulting in satisfactory postoperative period with the regression of epileptogenic events [8,15]. The subtotal resection, although contributing to the control of seizures, shows greater recurrence rate or tumor progression according to the literature [9], and ideally, total resection – as our case – is preferred. Cases in which adjuvant radiotherapy becomes necessary are rare [10]. Outcome is mostly excellent, with highly favorable prognosis [11,12].

4. Conclusion

Angiocentric Glioma is a rare neoplasm. Similar to low-grade lesions, it is mostly present in patients with difficult to control epilepsies, and in the differential diagnosis of neoplasia, especially in children and young adults, it should be considered. This slow-growing tumor, usually is benign with excellent curative rate through surgical treatment. Tumoral resection provides broad control of symptoms, besides reducing recurrence and the progression of the disease, leading to an extremely favorable prognosis. However, long-term monitoring is necessary to ensure the indolent behavior of the tumor, as most reports showed so far.

Support

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References

[1] S.F. Shakur, M.J. McGirt, M.W. Johnson, P.C. Burger, E. Ahn, B.S. Carson, et al., Angiocentric glioma: a case series, J. Neurosurg. Pediatr. 3 (3) (2009 Mar) 197–202, https://doi.org/10.3171/2008.11.PEDS0858.

[2] M. Wang, T. Tihan, A.M. Rojiani, S.R. Bodhireddy, R.A. Prayson, J.J. Iacuone, et al., Monomorphous angiocentric glioma: a distinctive epileptogenic neoplasm with features of infiltrating astrocytoma and ependymoma, J. Neuropathol. Exp. Neurol. 64 (10) (2005) 875–881 Oct.

[3] A. Lellouch-Tubiana, N. Boddaert, M. Bourgeois, M. Fohlen, A. Jouvet, O. Delalande, et al., Angiocentric neuroepithelial tumor (ANET): a new epilepsy-related clinicopathological entity with distinctive MRI, Brain Pathol. 15 (4) (2005 Oct) 281–286, https://doi.org/10.1111/j.1750-3639.2005.tb00112.x.

[4] D.N. Louis, H. Ohgaki, O.D. Wiestler, W.K. Cavenee, P.C. Burger, A. Jouvet, et al., The 2007 WHO classification of tumours of the central nervous system, Acta Neuropathol. 114 (2) (2007) 97–109, https://doi.org/10.1007/s00401-007-0243-4 Aug.

[5] D.N. Louis, A. Perry, G. Reifenberger, A. von Deimling, D. Figarella-Branger, W.K. Cavenee, et al., The 2016 World Health Organization classification of tumors of the central nervous system: a summary, Acta Neuropathol. 131 (6) (2016) 803–820, https://doi.org/10.1007/s00401-016-1545-1 Jun.

[6] K.J. Weaver, L.M. Crawford, J.A. Bennett, M.L. Rivera-Zengotita, D.W. Pincus, Brainstem angiocentric glioma: report of 2 cases, J. Neurosurg. Pediatr. 20 (4) (2017) 347–351, https://doi.org/10.3171/2017.5.PEDS16402 Oct.

[7] D. Alexandru, B. Haghighi, M.G. Muhonen, The treatment of angiocentric glioma: case report and literature review, Perm J 17 (1) (2013 Winter) e100–e102, https://doi.org/10.7812/TPP/12-060.

[8] L. Ampie, W. Choy, J.D. DiDomenico, J.B. Lamano, C.K. Williams, K. Kesavabhotla, et al., Clinical attributes and surgical outcomes of angiocentric gliomas, J. Clin. Neurosci. 28 (2016) 117–122, https://doi.org/10.1016/j.jocn.2015.11.015 Jun.

[9] J.A. McCracken, M.F. Gonzales, P.M. Phal, K.J. Drummond, Angiocentric glioma transformed into anaplastic ependymoma: review of the evidence for malignant potential, J. Clin. Neurosci. 34 (2016) 47–52, https://doi.org/10.1016/j.jocn.2016.08.012 Dec.

[10] R. Soffietti, R. Rudà, D. Reardon, Rare glial tumors, Handb. Clin. Neurol. 134 (2016) 399–415, https://doi.org/10.1016/B978-0-12-802997-8.00024-4.

[11] Gonzalez-Quarante LH, Fernández Carballal C, Agarwal V, Vargas Lopez AJ, Gil de Sagredo Del Corral OL, Sola Vendrell E. Angiocentric glioma in an elderly patient: case report and review of the literature. World Neurosurg 2017 Jan;97:755.e5- 755.e10. doi: https://doi.org/10.1016/j.wneu.2016.10.034.

[12] D. Adamek, G.P. Siwek, A.A. Chrobak, I. Herman-Sucharska, S. Kwiatkowski, R. Morga, et al., Angiocentric glioma from a perspective of A-B-C classification of epilepsy associated tumors, Folia Neuropathol. 54 (1) (2016) 40–49, https://doi.org/10.5114/fn.2016.58914.

[13] M.T. Whitehead, G. Vezina, MR spectroscopic profile of an angiocentric glioma, Anticancer Res. 35 (11) (2015 Nov) 6267–6270.

[14] M. Preusser, A. Hoischen, K. Novak, T. Czech, D. Prayer, J.A. Hainfellner, et al., Angiocentric glioma: report of clinico-pathologic and genetic findings in 8 cases, Am. J. Surg. Pathol. 31 (11) (2007) 1709–1718, https://doi.org/10.1097/PAS.0b013e31804a7ebb Nov.

[15] S. Pokharel, J.R. Parker, J.C. Parker Jr., S. Coventry, C.B. Stevenson, K.K. Moeller, Angiocentric glioma with high proliferative index: case report and review of the literature, Ann. Clin. Lab. Sci. 41 (3) (2011 Summer) 257–261.