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Case Study

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ASTROBLASTOMA IN A YOUNG FEMALE PATIENT: ABOUT A CASE

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ABSTRACT

Astroblastoma is an uncommon glial tumor predominantly diagnosed in children and young adults, with female preponderance. It shares almost the same clinical and radiologic features with the other glial neoplasm. Histologically, astroblastomas can be graded as either a low-grade (Well differenciated astroblastoma) or high-grade (Anaplastic/ malignant) variant. Due to its rarity, the treatment is not codified. Upfront surgery however play a major role in the management of these tumors, and the prognosis is sometimes closely related to the completeness of the resection. The role of adjuvant therapy is still subject of controversies particularly in the low-grade subset. We report a case of 36-year-old female patient with a low-grade astroblastoma who underwent incomplete resection followed by adjuvant radiotherapy with a good 3 years local control.

KEYWORDS: Low-grade astroblastoma, surgery, adjuvant radiotherapy.

INTRODUCTION

First described by Bailey et al. in 1926, astroblastoma is a rare glial-origin tumor that affects predominantly children and young adults. It accounts for 0.45 to 2.8% of all neuroglial tumors.[1] There is no specificity in clinical behaviour and radiological appearance compared with the other glial tumors. Histologically, astroblastoma is characterised by astroblastic pseudorosettes and perivascular hyalinisation. According to WHO 2016 classification of gliomas, astroblastoma, chordoid glioma of the third ventricle and angiocentric glioma belong to the "other gliomas" group. While angiocentric gliomas are grade I, chordoid gliomas II, there were insufficient data available for unequivocal grading of astroblastomas. Morphologically, they were classified into prognostically favorable 'low-grade' and unfavourable 'high-grade' groups.

Owing to its rarity, information available on both diagnosis and management is provided predominantly and sparsely by clinical case series reports. The largest reported series of astroblastoma patients in the literature is those of Ahmed and al. who collected data of 239 patients using the SEER database.[2] However, the findings of this report do not provide strict therapeutic guidance. Gross total resection is the treatment of choice, but the role of adjuvant chemotherapy and/or radiation therapy is not yet well established. While high-grade astroblastoma could benefit from adjuvant radiation therapy, its role in their counterpart low-grade is controversial and unclear.

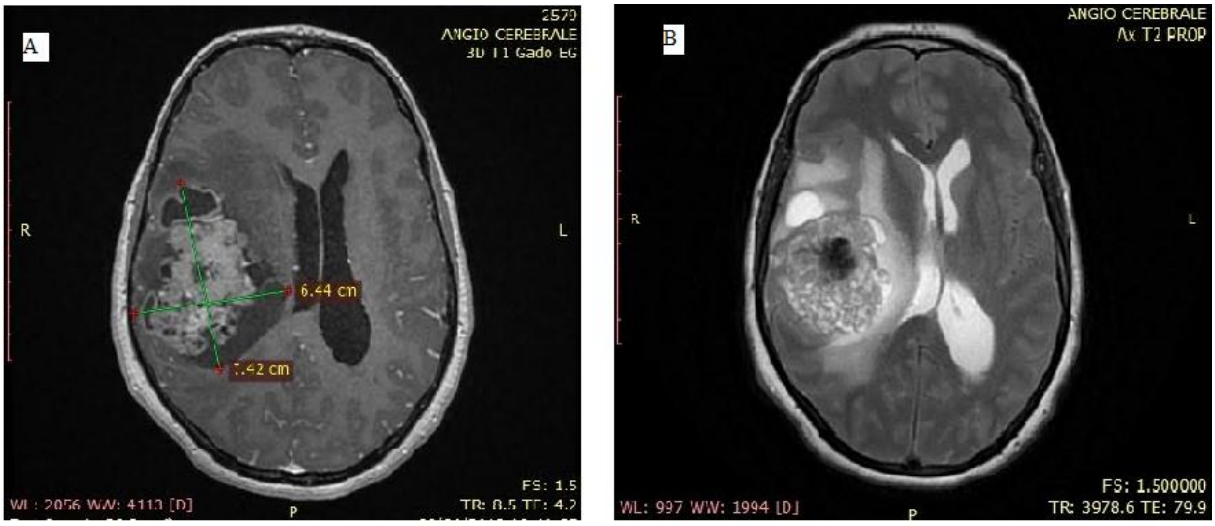
We report in this study a case of low-grade astroblastoma in an adult-young female patient treated with adjuvant radiotherapy after partial surgical resection.

Case presentation

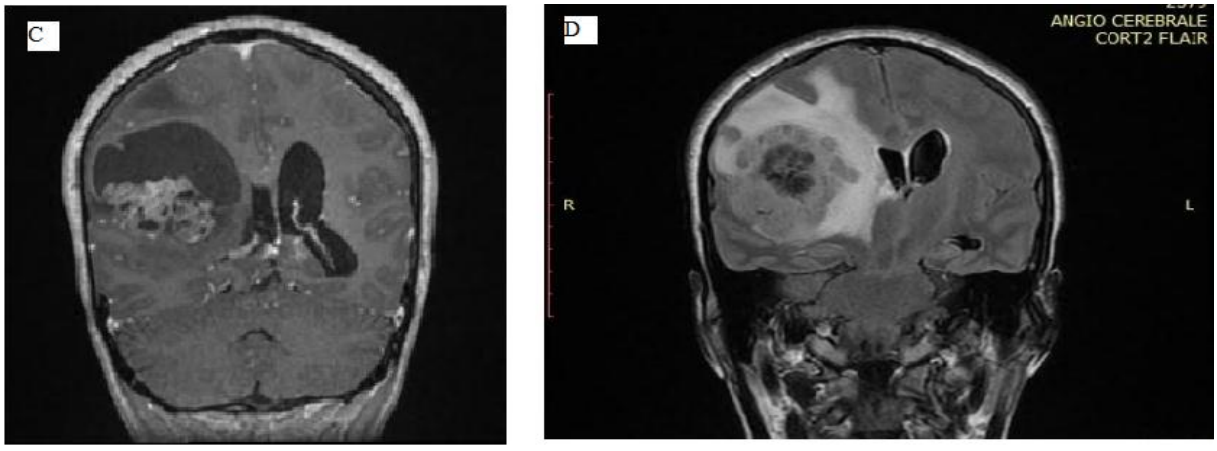
She is a 36-year-old patient who presented with a 1-year history of epileptic seizure, headache, decreased visual acuity of right eye, right hemiplegia, and sudden aphasia in postpartum.

Cranial magnetic resonance imaging (MRI) angiography revealed a right, tempo-parietal, well-demarcated large mass with double cystic and solid component.

The solid component raise intensely and heterogeneously after the injection of gadolinium. This process measures 60x80mm. A range of surrounding cerebral oedema was noted. There was a mass effect on the right lateral ventricle, which is collapsed and pushes back the median structures on the left (Figure 1).



Axial T1-weighted (A) and T2-weighted images (B)



Coronal T1-weighted (C) and T2-weighted images (D)

Figure 1: MRI-angiography.

The ophthalmologic examination carried out noted a visual acuity rated at 1/10 in the right eye and 10/10 in the left eye with a fundus which notes a bilateral optic atrophy more pronounced in the right eye.

She underwent a large surgical excision. Pathologic report of resection specimen revealed the presence of high density tumor proliferation. Tumor cells were quite monomorphic, globular or endowed with an extension. The mitosis count is estimated at 2 mitoses/10 fields (x40 magnification). These cells were arranged in perivascular rosettes with a hyalinized fibrous aspect of the vessel wall. The stroma is, by place, largely remodelled by a dense hyalinized fibrosis. There was no necrosis or capillary proliferation.

On immunohistochemistry (IHC) staining, cells were fully positive for glial fibrillary acid protein (GFAP), anti-vimentin, epithelial membrane antigen (EMA) patchy antibodies and focally positive for PS100 and anti-Olig2 antibodies. Anti-Ki67 antibody was estimated at 10% on a hot spot.

A control MRI found the presence of a temporo-parietal right cortical-subcortical process poorly limited in heterogeneous T2 signal measuring 38x50x32 mm.

An adjuvant radiotherapy was decided. The treatment plan was performed with intensity modulated radiotherapy (IMRT). Patient was simulated in supine position, head in neutral position. Immobilization was made with a 3 points thermoplastic mask. We performed a contrast-enhanced CT-scan from vertex to C7-T1 level with 3mm slights thickness.

MRI scan with T1 contrast-enhanced, T2-weighted and T2-flair sequences in the same position was also performed. Dataset of this MRI scan was fused with CT-scan images data for target volume and organ at risk delineation.

The Gross Tumor Volume (GTV) included resection cavity and the residual enhancing tumor on contrast-enhanced T1 with without peripheral edema.

The Clinical Target Volume (CTV) was defined as GTV included the GTV plus 10 mm geographic margin with respect to anatomic barriers. The CTV margins were reduced near critic organ. A 0,5 cm margin in all directions was added to the CTV to obtain the planning target volume (PTV).

The total dose of radiotherapy was 54 Gy in conventional fractionation (2 Gy per fraction/ day, 5 days per week). Organs at risk were normal brain, hippocampus, contralateral temporal lobe, optic chiasm, optic nerves, brain stem, lens, and retina. Dose constraints to OAR were set according to the RECORAD.

Evolution after 36 months is marked by a lesion stability on MRI, persistence of hemiplegia and right eye blindness with complete regression of convulsive seizures.

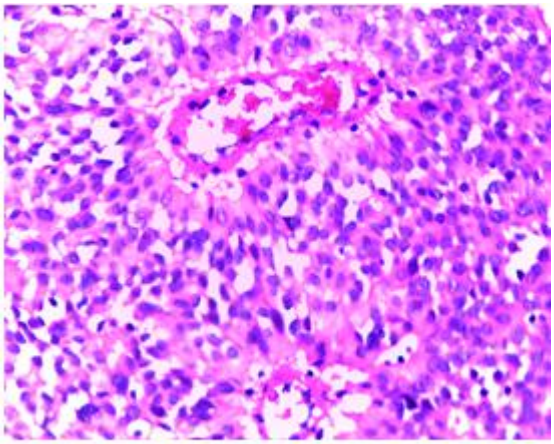


Figure 2 : Magnification X 40

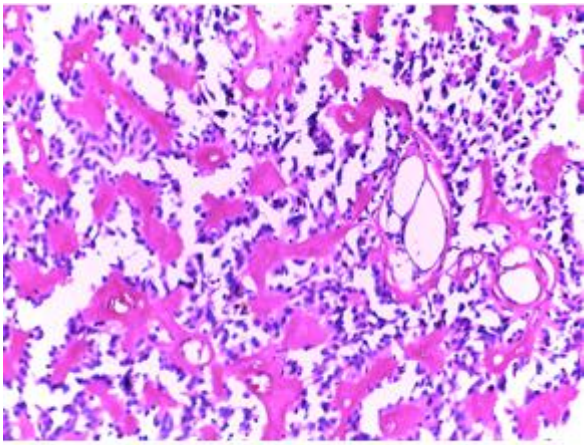


Figure 3 : Magnification X 20

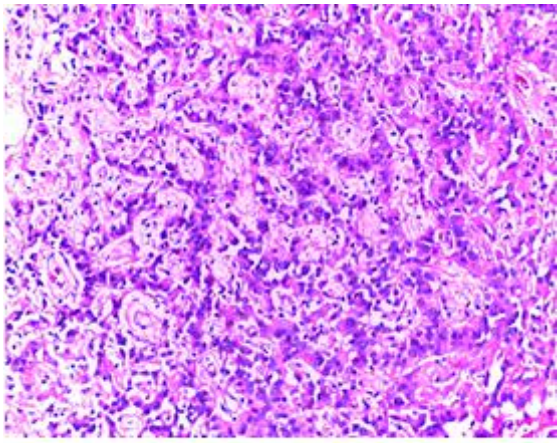


Figure 4 : Magnification X 20

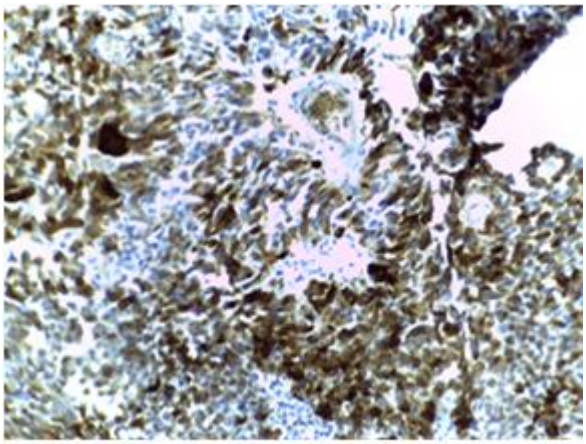


Figure 5 : Immunohistochemistry GFAP

Tumor proliferation forming pseudorosettes around the vessels (Figure 2) and hyalinized fibrous vessel walls (Figure 3 and 4). GFAP confirms the glial nature of this tumor (Figure 5).

DISCUSSION

We reported a case of low-grade astroblastoma in a young-adult female managed by subtotal surgical resection followed by post-operative radiotherapy that resulted in a lesion stability after 24 months of follow-up.

Astroblastomas are very rare glial tumors mostly diagnosed in children and young-adult as in our case. The studies performed to date show a striking female preponderance.[3] There are uncertainties in histogenesis, classification, treatment and prognosis of theses tumours. They develop typically in the cerebral hemispheres, affecting most likely the frontal lobe followed by parietal and temporal lobes.[3] As notified in our patient, clinical manifestations are usually related to the raise of intracranial pressure, and include headache, nausea or vomiting, seizures, and visual disturbance that depend on tumor size and location.

On histologic examination, astroblastoma is characterized by the presence of perivascular pseudorosettes and prominent hyalinization. They exhibit characteristic epithelioid cells with cytoplasmic processes having blunt-ended foot plates attached to the basal lamina of blood vessels.[5-7] Another feature of diagnostic importance is lack of fibrillary background.[7] On immunohistochemistry, they are positive for GFAP,

S100 and Vimentin and frequently with focal expression of EMA. WHO 2016 Classification of Tumors of CNS describes malignant astroblastoma as having focal or multiple foci of high cellularity, anaplasia, increased mitotic activity (>5 mitoses per HPF), elevated proliferative index (typically >10%), microvascular proliferation and necrosis.[8-9]

One histological aspect that is relevant and have a therapeutic and prognostic impact is the tumor grade. Tumors are graded either as low-grade or as high-grade variant based on some histological features. High-grade tumors show focal or multifocal regions of high cellularity, anaplastic nuclear features, high mitotic rates, vascular proliferation, and necrosis with pseudo palisading. They have shorter postoperative survival times. Low-grade tumors include astroblastomas with uniform perivascular arrangement of pseudorosettes, low to moderate numbers of mitotic figures, minimal cellular atypia, minimal to no vascular endothelial proliferation, and predominant sclerosis of the vascular walls. They are generally indolent and associated with a more favorable prognosis after surgical resections. Thiessen et al. discussed the predictive value of histopathological subtyping based on a series of seven patients. All four patients with high-grade tumors received surgery, radio- and chemotherapy. Three of them died. The three low-grade patients survived recurrence free after surgery alone. However, there are some reports on early recurrences in low-grade cases.[9-11]

The review of the literature performed by Mangano[10] found nosignificant difference in overall survival of low-grade patients who underwent surgery versus surgery and adjuvant therapy, thus indicating that surgery and close follow-up might be a good treatment strategy for low-grade astroblastoma.

On MRI the solid part of the tumor is hypointense to grey matter on T1-weighted imaging, isointense on T2-weighted imaging, has a soap-bubble appearance and shows heterogeneous contrast enhancement.[11] In relation to its large size, there is relatively little peritumoral T2 hyperintensity indicating perhaps a lack of infiltration of surrounding areas. Occasionally T2 hyperintensity due to edema may be seen in lesions with acute hemorrhage. Similar appearing lesions on imaging include anaplastic astrocytoma and glioblastoma, but central necrosis and peritumoral T2 hyperintensity help in differentiation. Anaplastic oligodendroglioma and juvenile pilocytic astrocytoma may also have a similar appearance but calcification and a mural nodule aid in differentiation, respectively.[11,12]

Due to the rarity of the lesion and limited cases reported in the literature, the natural course and response to therapy remains poorly defined.[11] Surgery is the treatment of choice for this disease and is associated with increased overall survival.[2] Gross total resection is achievable for the majority of astroblastomas (>80% of cases) and is associated with improved clinical outcomes compared to subtotal resection.[13] When patients have undergone STR, adjuvant radiotherapy has been administered to most patients, but the benefit of this approach lakes and is unknown.[13] Some studies mention surgical resection followed by close observation only for low grade lesions and surgery followed by radiotherapy for high grade lesions.[10,11,14] Others advocate radiotherapy for low as well as for high-grade lesions. In our patient, a gross total resection was not achieved, thus a post-operative radiotherapy has been administered. In review of the literature of Janz and Buhl, 5 out of 8 patients had a recurrence after GTR without further therapy. All of them displayed a marked oedema on initial MRI. They therefore suggested that this subgroup of low-grade patients should undergo postoperative radiotherapy. Based on these observations (marked oedema and subtotal resection) the adjuvant radiotherapy was warrant in our patient.

CONCLUSION

Astroblastoma is a very rare primary brain tumor. Progresses in imaging, histology and immunohistochemistry have permitted accurate diagnosis. Prognosis of low-grade tumor remain unclear, as some authors have reported early recurrences. Total surgery resection, the mainstay of the treatment, can be mostly sufficient for them. However, adjuvant radiotherapy could provide a good tumor control in case of incomplete resection.

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