



Case Report

# Thirteen-year long-term follow-up in a rare case of anaplastic astroblastoma: What makes the difference?

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## ABSTRACT

**Background:** Astroblastomas are uncommon neuroepithelial tumors of the central nervous system with a distinct, yet, controversial radiological, histological, and molecular profile. Debatable differences between low- and high-grade astroblastoma have been reported in the medical literature; indeed, despite the increasing relevance of molecular genetic profiling in the realm of astroblastoma, its application is still in its early stages. As a result, the diagnostic criteria for astroblastoma remain undecided with yet no real consensus on the most ideal management.

**Case Description:** This report describes a case of astroblastoma diagnosed 13 years ago in a young woman who despite six episodes of recurrence, transformation, and progression was able to retain a performance status of 0 by World Health Organization standard, throughout.

**Conclusion:** This report discusses the clinical, radiological, histological features, and management of this rare tumor with an extraordinarily long survival, with an aim to strengthen the literature on management options. To the best of our knowledge, this is the longest surviving case of anaplastic astroblastoma reported in the available medical literature.

**Keywords:** Astroblastoma, Histomolecular diagnosis, Long-term follow up, Neuro-oncology, Prognosis

## INTRODUCTION

Astroblastomas are scarce and controversial primary neuroepithelial tumors of the central nervous system (CNS) occurring in the cerebral hemispheres of children and young adults with an incidence from 0.45% to 2.8% of all neuroglial tumors.<sup>[1,13,22]</sup> Historically, astroblastomas were first described by Bailey and Cushing in 1926 followed by a further characterization in 1930 by Bailey and Bucy.<sup>[4,5]</sup> With a median age at diagnosis of 35 years, alongside a median overall survival (OS) of 55 months, astroblastoma remains a devastating diagnosis to receive.<sup>[2]</sup> Rarely, certain cases have been reported to survive longer than 10 years.<sup>[2]</sup> Clinically, these tumors present due to symptoms associated with the sequelae of raised intracranial pressure, such as persistent headache, vomiting, and focal neurological deficit.<sup>[1,2,24]</sup> Due to the extreme rarity of the entity, there is currently no standard protocol for patients undergoing treatment for astroblastoma, with surgical interventions and adjuvant therapy being decided on by senior members of the multidisciplinary team.

As a result of a paucity of evidence on management, both low- and high-grade astroblastomas remain associated with a high degree of mortality. Moreover, with astroblastoma grading being

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a contentious issue due to the specific definition criteria, the OS of each grade is infrequently reviewed. In general terms, low-grade tumors have an OS in the region of years, whereas, high-grade or anaplastic tumors have been associated with a particularly poor prognosis, with the majority of cases not surviving beyond 1 year.<sup>[12]</sup> The longest surviving case of anaplastic astroblastoma that the authors could locate in the available medical literature was a case reported by Bergkåsa *et al.* in which a 50-year-old female was diagnosed with a brain neoplasm following a seizure, subsequently revealed to be anaplastic astroblastoma; she was in complete remission 7 years after surgery and more than 6 years after completing chemotherapy.<sup>[8]</sup> If the authors are correct, this would make our patient the longest ever reported case of anaplastic astroblastoma in the available medical literature.

### CASE PRESENTATION

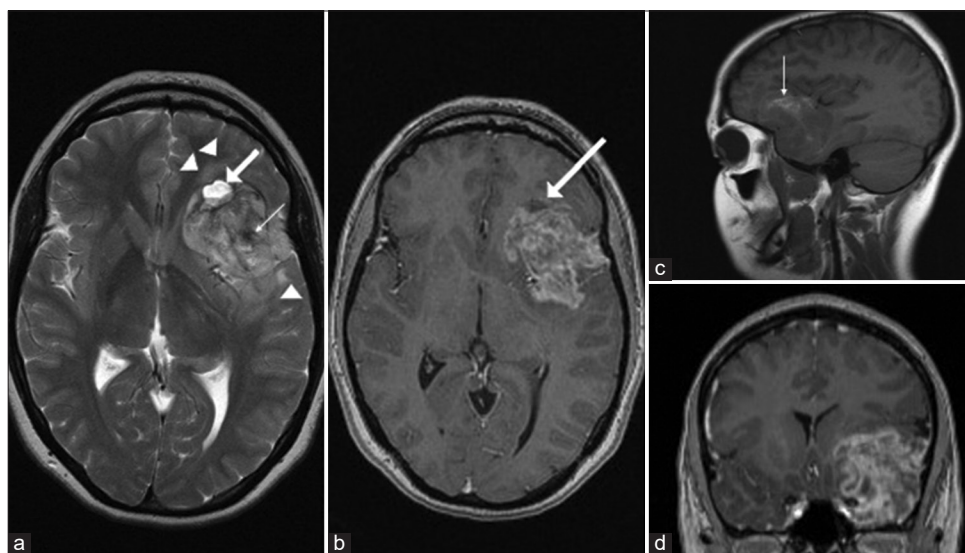
The authors present the case of a 40-year-old female who initially presented 13 years previously (October 2008) at the age of 27 years. The patient was 6-month postpartum complaining of progressive left-sided headaches. Initial magnetic resonance imaging (MRI) on T1-weighted series demonstrated a 5.5 cm × 3.9 cm × 4.8 cm left frontotemporal peripherally located mass with solid and cystic components with evidence for internal hemorrhage but relatively little surrounding parenchymal T2 hyperintensity [Figure 1].

Following radiological diagnosis, initial craniotomy and resection were performed (October 2008), with gross total resection (GTR) being satisfactorily achieved. Indeed, the

postoperative MRI demonstrated postsurgical gliotic changes with focal atrophy in the left temporal lobe with no evidence of residual tumor [Figure 2]. The postoperative period was unremarkable with the patient being weaned off steroids quickly, with no residual neurological deficit and rapid recovery. Subsequent histological analysis deemed the tumor to be an astroblastoma, based on morphological criteria of the then current 4<sup>th</sup> edition of the World Health Organization (WHO) classification of tumors of the CNS, with typical features such as perivascular rosettes [Figure 3a], hyalinization of the vascular wall [Figure 3b], and positive immunostaining for glial fibrillary acid protein [Figure 3c]. It is important to remind the reader that the WHO grading was not formally applicable to astroblastomas at that time and that molecular analysis was not available to the laboratory then; yet, features suggestive of a high-grade anaplastic tumor such as conspicuous mitotic activity, cytological atypia, architectural disorganization, and pseudopalisading necrosis were not apparent.

Initial postoperative management consisted of close surveillance with 3 monthly MR imaging, with clinical follow-up thereafter. At initial follow-up in May 2009, the patient was clinically well with the MRI demonstrating postoperative changes only with no visible residual or recurrent tumor.

At 26-month postdiagnosis (December 2010), the corresponding MRI revealed two lesions in the left temporal MRI revealed two lesions in the left temporal surgical site. One anteriorly, in close proximity to the optic nerve, with the second more superior and posterior, located within the Sylvian fissure. At this stage, re-do craniotomy with excision of the recurrent tumor was decided on. Ultimately,



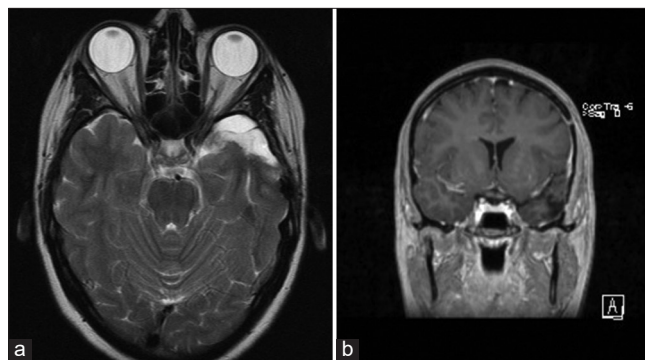
**Figure 1:** (October 2008): axial T2-weighted (a) and sagittal T1-weighted (b) images show a supratentorial peripherally located left frontotemporal lobulated fairly well-defined heterogeneous mass having cystic (thick arrows) and solid components with evidence of hemorrhage (thin arrows), and relatively little adjacent peritumoral parenchymal T2 hyperintensity (arrow heads). Axial (c) and coronal (d) postcontrast T1-weighted images reveal heterogeneous enhancement of the tumor with rim enhancement of the cystic components (arrows).

GTR was again achieved, with histological analysis revealing frequent mitoses and more cytological atypia, suggesting the presence of a high-grade astroblastoma.

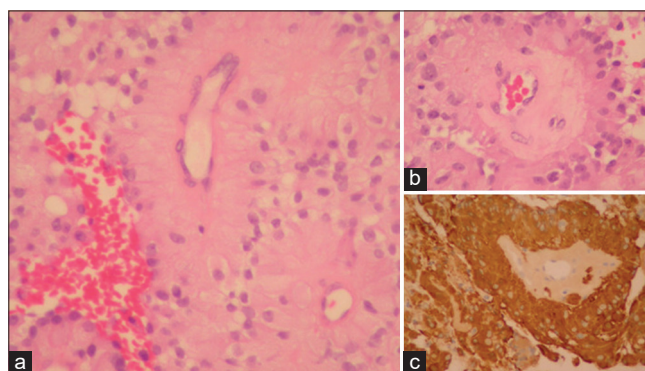
Adjunct treatment was discussed at this stage; considering the rapid timeline of recurrence and potential high-grade transformation, “glioblastoma-targeted” treatment was offered. The patient underwent concurrent radical chemoradiotherapy (60 Gy in 30 fractions along with 75 mg/m<sup>2</sup> temozolomide daily) given from February to March 2011. On completion, she was started on cyclic single-agent temozolomide (cycle 1: 150 mg/m<sup>2</sup> daily for 5 days; cycles 2–6: 200 mg/m<sup>2</sup> daily for 5 days). Unfortunately, due to excessive suppression of the bone marrow despite of subsequent dose reduction, cyclic chemotherapy had to be discontinued after just two cycles (May–June 2011).

During routine MR surveillance in January 2014, an asymptomatic relapse was identified, and the patient was treated with Gamma Knife radiosurgery. Despite of the latter, the tumor recurred in August 2015 and the patient was reoperated with craniotomy and excision; the pathology reports confirmed the recurrence, with tumor cells present in the scar tissue at the site of the previous Gamma Knife surgery. This process of recurrence occurred again in September 2016 [Figure 4] and May 2018, with the patient being treated with re-do craniotomy followed by 4 cycles of procarbazine (75%), lomustine, and vincristine (PCV) with granulocyte-colony-stimulating factor (G-CSF) cover (lipegfilgrastim) due to hematotoxicity (July–December 2018). In January 2020, she experienced a further relapse, this time managed only with surgery due to previous low tolerance to chemotherapy; as expected, the tumor progressed further in July 2020 [Figure 5]. No further neurosurgery was deemed appropriate at this stage, due to the tumor’s close proximity to the left middle cerebral artery with the inherent risk of cerebrovascular accident-related morbidity and intraoperative mortality. As such, after careful consideration, she underwent 8 cycles of reduced temozolomide (50%) from October 2020 to May 2021; G-CSF cover was not required as she tolerated treatment well. Gladly, the MRI follow-ups of December 2020–April 2021 showed satisfactory response to treatment, with decreased contrast enhancement and cerebral blood volume (CBV) at the site of the lesion. Although the surveillance MRI of July 2021 [Figure 6] showed a small contrast enhancement in the medial aspect of the left temporal lobe, there was overall less tumor volume compared to the previous studies [Figure 5]; the nature of the enhancing lesion remains unclear as the most recent MRI (September 2021) showed minimal changes compared to the previous study of July.

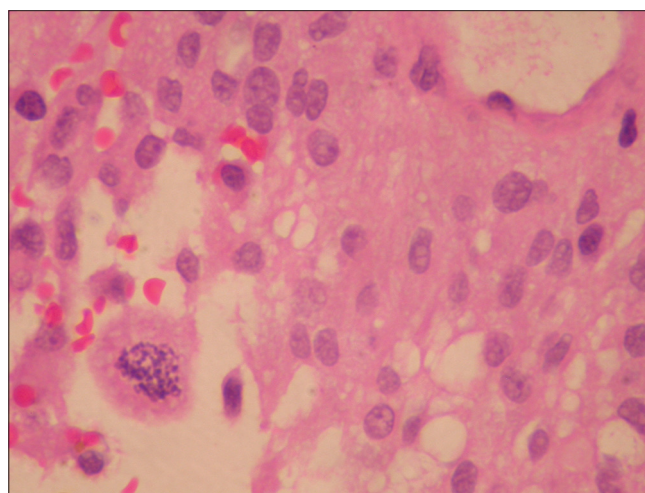
Despite the recurrences, transformation, and progression of this high-grade histology, the patient remained clinically



**Figure 2:** (January 2009): (a) Axial T2-weighted and (b) coronal post-contrast T1-weighted images obtained after resection show complete excision of tumor with residual gliotic and postsurgical changes involving the left temporal lobe.

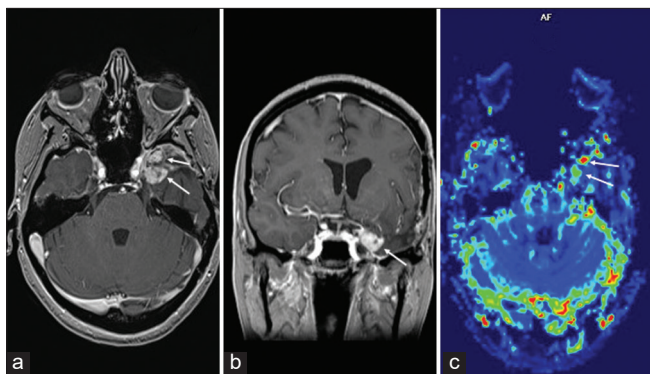


**Figure 3:** A set of light microscopy images from tissue samples taken in October 2008 at initial surgery demonstrating (a) perivascular rosettes of tumor cells with broad processes passing down to the outside of central small blood vessels, (b) hyalinization of the blood vessel wall, and (c) positive (brown) immunostaining for glial fibrillary acid protein.

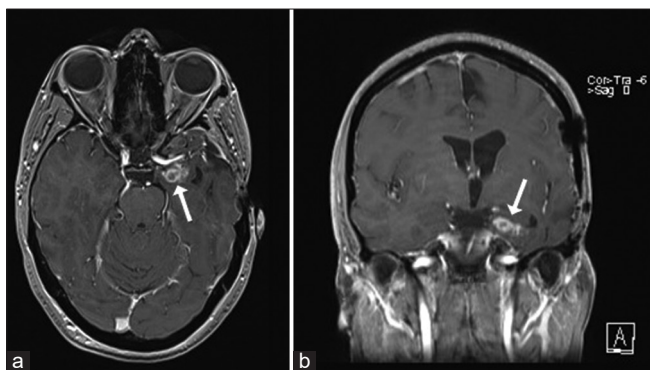


**Figure 4:** (September 2016) – Hematoxylin and eosin stained section of recurrent tumor in 2016, with mitotic figure at the lower left.





**Figure 5:** (July 2020): axial (a) and coronal (b) postcontrast T1-weighted images showing recurrent nodular enhancing tumor (arrows) along the inferior aspect of the medial temporal lobe, also showing increased perfusion on the relative cerebral blood flow map (c) obtained from dynamic susceptibility contrast planar imaging.



**Figure 6:** (July 2021): axial (a) and coronal (b) postcontrast T1-weighted images showing overall regression of the previous tumor with a new small indeterminate nodular area of contrast enhancement (arrows).

stable at the time of paper submission, with a performance status of 0 and no new or evolving neurological symptoms. Further chemotherapy with reduced temozolomide will be considered in case of recurrence on upcoming interval imaging.

## DISCUSSION

### General aspects

Astroblastomas are infrequent neoplasms of the CNS, accounting for approximately 0.48–2.80% of all gliomas.<sup>[22]</sup> Studies suggest that astroblastoma has a bimodal age distribution, with peaks at 5–10 years of age and again at 21–30 years, alongside a strong female predilection of the tumor, with a male-to-female ratio of 1:11;<sup>[2,3,13,19,23,25]</sup> the latter facts fit the characteristics of our patient case quite well. Clinically, as in our case, presenting symptoms are usually associated with the sequelae of raised intracranial pressure,

such as persistent headache, vomiting, and focal neurological deficit.<sup>[1,2,24]</sup>

On MRI, astroblastomas are typically seen as large, lobulated, supratentorial, well-demarcated, solid, and cystic masses.<sup>[11,23]</sup> As compared to infiltrative gliomas, these show relatively little surrounding vasogenic edema or tumor infiltration for their large size. Ependymomas also show cystic components but are more commonly placed centrally and have an infratentorial location. In general, the solid components in astroblastomas have been described as appearing hypointense to gray matter on T1-weighted images and having a bubbly appearance on T2-weighted images. Further imaging features include intratumoral hemorrhage with a fluid-fluid level and less commonly adjacent bone scalloping and dural “tails.”<sup>[6]</sup> As previously explained, our case demonstrated most of these traits at presentation. Notwithstanding the aforementioned characteristics, radiographic distinction between well-differentiated and malignant astroblastomas, as well as with infiltrative gliomas, might prove difficult and warrants experience.

### Treatment: trends and controversies

Ahmed *et al.* presented the largest series of astroblastoma cases in 2013 ( $n = 239$ ), followed by a subsequent review of 54 cases by Hammas *et al.* in 2018.<sup>[2,13]</sup> The group found that increased age, a supratentorial anatomical location, and treatment before 1990 were poor prognostic factors, almost certainly reflecting advancements in imaging modalities and the increased likelihood of early symptomatic presentation in an infratentorial tumor. Regarding management, OS in patients receiving surgery was seen to be 43 months, statistically improved in comparison to patients not receiving surgery, in which patients had an OS of 13 months. In this context, as in our patient, GTR was the surgical procedure of choice, due to increased survival over subtotal resection (83% vs. 55%).<sup>[2,26]</sup> The same authors found that there was a trend toward improved OS in patients receiving radiotherapy versus no treatment; however, the difference was not statistically significant. In our patient’s case, radiotherapy may have delayed tumor growth to some degree, particularly adjunctive fractionated radiation treatment; however, the latter cannot be confirmed due to insufficient evidence. Of note, the Ahmed study did not identify or stratify tumors between low-grade and high-grade classifications.<sup>[2]</sup>

Similarly, Hammas *et al.* commented on the benefits of GTR over subtotal resection, stating the benefits of the former in providing superior tumor control rates. Also mentioned was that the addition of adjuvant focal radiotherapy after subtotal resection does not appear to provide equivalent outcomes to GTR. Also, the authors reiterated the overall benefits of

adjuvant therapy for high-grade and recurrent cases.<sup>[2]</sup> Again, this observation is well in keeping with the patient's clinical evolution throughout follow-up.

In terms of recurrence, the role of chemotherapy remains a question of debate; despite the controversy, some groups have reported on the benefit of systemic treatment in some of these patients.<sup>[7-9]</sup> Similar to our case, Bergkåsa *et al.* reported on a case of anaplastic astroblastoma promptly recurring after surgery and radiation; the patient was treated with 3 cycles of PCV (procarbazine, CCNU, and vincristine), later switched to 3 cycles of temozolomide due to bone marrow toxicity. The ensuing imaging showed good response to treatment throughout 7 years of follow-up.<sup>[7,8]</sup>

Based on our institutional experience and previous review of the medical literature, we strongly recommend the use of temozolomide in cases of high-grade histology recurring at postsurgery and/or postradiation; as it was the case here, a satisfactory degree of response might be achieved, even at reduced doses. Should temozolomide not be indicated due to local failure or toxicity, PCV (alternatively PC or single-agent CCNU depending on the patient's fitness to chemotherapy) might prove beneficial in terms of local control although bone marrow toxicity remains a precluding factor, particularly at full dose.

### Histogenesis and molecular profile: A complex subject

Despite being part of the diagnostic neuro-oncology portfolio since 1926, the histogenesis of astroblastoma continues to be debated by neuropathologists. This comes as no surprise as these glial neoplasms contain complex features overlapping both astrocytomas and ependymomas as well as other entities such as high-grade neuroepithelial tumor with MN1 alteration (HGNET-MN1).<sup>[27]</sup> In this histological framework, astroblastomas are characterized by an abundance of astroblastic pseudorosettes, with perivascular hyalinization.<sup>[16]</sup> Yet, in a similar manner, ependymoma can also expose features of perivascular pseudorosettes; despite this, the general consensus among experts is that broad-based (rather than fibrillary) processes forming pseudorosettes remain more associated with astroblastoma than ependymoma.<sup>[21]</sup> Making matters more complicated, astroblastomas are neither entirely astrocytic nor "blastic." The name is believed to be associated with the assumption that astroblastomas originate from the astroblast, an intermediate of glioblasts and astrocytes.<sup>[8,14]</sup> However, more recently, astroblastomas have become more controversial due to the emergence and increasing use of molecular profiling of CNS tumors alongside traditional standalone histological diagnosis. This, in tandem with the rarity of astroblastoma, has resulted in a paucity of molecular information and increasing doubts surrounding historical diagnoses based

purely on tumor morphology. In the most recent 2021 update of the WHO Classification of CNS tumors, astroblastoma has been categorized as "circumscribed astrocytic gliomas," within "gliomas, glioneuronal tumors, and neuronal tumors." New changes include the introduction of meningioma-1 proto-oncogene alteration (MN1) as a molecular marker for astroblastoma to provide more diagnostic focus for the entity.<sup>[18]</sup> MN1 is a transcriptional regulator in which inactivation through a [4:22] balanced translocation results in meningioma pathogenesis and has been identified as being altered in 48% of astroblastoma cases and associated with favorable prognosis.<sup>[10,17,18,28]</sup>

In the case presented here, molecular profiling was not carried out at initial diagnosis as molecular profiling was not available then. Despite this, the tumor was diagnosed as an astroblastoma according to the current WHO classification, that is, based on the histological features of the neoplasm. As a result, some molecular analyses were subsequently carried out on the recurrent tumor (2010). A nonsense variant in RB1 and a frameshift variant in PTCH1 were identified while no pathogenic variants were detected in BRAF. Moreover, no mutations were detected in the following gene panel: ALK, ATRX, BCOR, CDKN2A, CDKN2B, CTNNA1, DDX3X, EZH2, FGFR1, H3F3A, HIST1H3B, HIST1H3C, IDH1, IDH2, MSH6, MYCN, NF1, NF2, PMS2, PTEN, SMARCA4, SMARCB1, SMO, SUFU, TERT, TP53, TSC1, TSC2, and YAP1 genes. HIST2H3C analysis failed. Of note again, although MN1 testing was not available at that particular point in time, the above collected data excluded the presence of CNS primaries other than astroblastoma.<sup>[15]</sup>

### High-grade transformation capability and its impact on survival

It is important to note that astroblastomas have not yet been given a WHO classification grade and that the histopathologic subtyping into low and high grade, although used locally by some centers, has not yet been fully integrated in the formal WHO classification, which makes the literature somehow inconsistent. However, we believe that subtyping remains a crucial element of diagnosis likely to shape management.

Indeed, as early as 1989, Bonnin and Rubinstein categorized astroblastoma into low- and high-grade subtypes, with high-grade tumors demonstrating particular features such as microvascular proliferation, necrosis with pseudopalisades, increased cellularity, and nuclear atypia with a high mitotic MIB-1 proliferative index.<sup>[9]</sup> In the context of prognosis, groups have reported a clear correlation between high-grade tumors and poor survival.<sup>[20]</sup> Two years postdiagnosis, our case was further classified as a high-grade astroblastoma due to the recurrence of the tumor, along with increased mitotic activity. We can always query as to whether this

neoplasm harbored mixed foci of low and high grade at initial diagnosis (2008) or if the tumor underwent a genuine, short interval transformation (2008–2010); this question might never be elucidated. Regardless, the patient's clinical evolution has remained remarkably "indolent" throughout follow-up despite of underlying high-grade features. With this considered, several observations ought to be considered when explaining the attained long-term survival:

1. In the case of low-grade tumors, complete resection may indeed be curative and appears to play a major role in postsurgery outcome for both grades of astroblastoma.<sup>[9]</sup> In our patient, although not achieving full local control, repeat surgery might have been helpful as to delay recurrence, particularly in the framework of high-grade tumor and supratentorial location. Furthermore, in this case, surgery proved useful as to delay chemotherapy, hence toxicity.
2. In accordance with our review of the literature, adjuvant treatment appears to remain critical in the management of high-grade lesions. In this patient case, radiotherapy and radiosurgery might have deferred recurrence to some degree, at least in theory. In contrast, the tumor seemed to be highly sensitive to temozolomide and PCV chemotherapy, even on a reduced dose schedule. In spite of the scarce clinical evidence and the ensuing lack of standard protocols for patients presenting with astroblastoma,<sup>[8]</sup> we believe that high-dose radiotherapy and systemic treatment stand as reasonable treatment options, particularly in adjuvant settings and complex patterns of recurrence.<sup>[7]</sup>

## CONCLUSION

We present a rare case of high-grade astroblastoma with complex evolution. To the best of our knowledge, this is the longest surviving case anaplastic astroblastoma in the available medical literature. Alongside increasing numbers of reported cases of astroblastoma, emerging histomolecular analysis of these tumors will further enhance clinical decisions surrounding the management of astroblastomas. In the majority of cases, management requires customized intervention, including surgery, radiotherapy, and chemotherapy. However, the rarity of the entity makes prospective studies hardly feasible and treatment protocols difficult to achieve. In this context, closer international cooperation surrogate to specific databases and multidisciplinary task groups are warranted and encouraged.

## Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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