




High-Grade Astroblastoma in a Young Female: An Enigma with a Rare Cautionary Tale

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Abstract

Astroblastoma is an uncommon neuroepithelial primary brain neoplasm with speculative histopathological origin and unpredictable clinical behavior. They can be easily misdiagnosed, as they are rarely encountered in clinical practice and share common radiological and histopathologic appearances with other glial neoplasms. Herein, we report a case of high-grade astroblastoma in a 27-year-old female with complaints of seizures and loss of consciousness, which was misdiagnosed as atypical meningioma on neuroimaging, due to its rarity and superficial cortical location appearing as extra-axial mass. Although intraoperative findings were also of an extra-axial tumor, the histology and immunophenotype was of an astroblastoma; thus, highlighting the role of histopathology and immunohistochemistry.

Keywords

- ▶ astroblastoma
- ▶ histology
- ▶ neuroepithelial brain tumor

Introduction

Astroblastomas are unusual neuroepithelial tumors constituting 0.45 to 2.8% of all neuroglial tumors and belongs to the “other glioma” category without any World Health Organization (WHO) grading in the 2016 edition of the WHO Classification of Tumours of the Central Nervous System.^{1,2} Although classically scrutinized as pediatric brain tumors, but astroblastomas tend to display a bimodal age distribution, with a peak prevalence in children aged 5 to 10 years and in young adults between 21 and 30 years.³ These are peripheral or superficially located large tumors, preferably in the cerebral hemispheres, but they have also been described in the cerebellum, brainstem, corpus callosum, hypothalamus, and the ventricular system.^{4–8}

The limited knowledge and extreme rarity created challenges in timely accurate diagnosis of these uncommon tumors, as its radiologic and histopathologic

features is shared with other glial neoplasms and causes delay in appropriate treatment.

Herein, we report a case of high-grade astroblastoma in 27-year-old female patient who was initially misdiagnosed as meningioma on neuroimaging, due to its superficial location, appearing as extra-axial, as well as on morphology. Later, it recurred after 1 year of initial presentation as recurrence of meningioma, but diagnosed as high-grade astroblastoma by histomorphology together with immunohistochemistry (IHC) study.

Case Report

A 27-year-old female patient was admitted to our institution with complaints of recurrent seizure and loss of consciousness since the last 6 months. One year prior to this presentation, she had complaints of headache, multiple episodes of vomiting and seizures. On magnetic resonance

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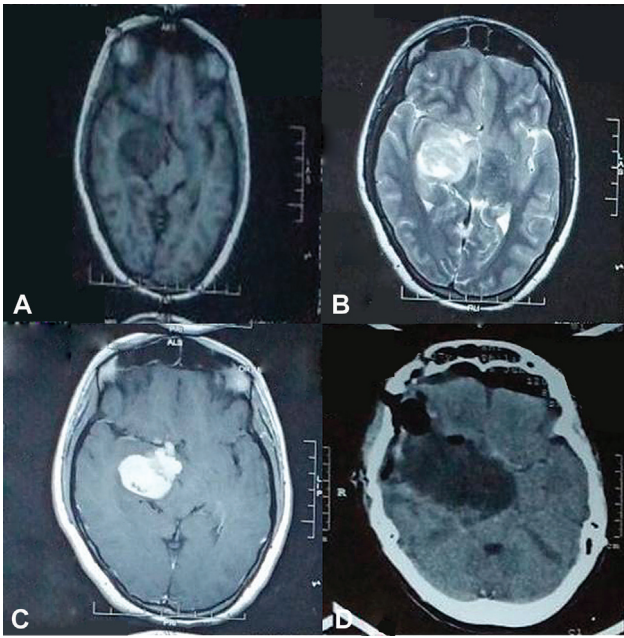


Fig. 1 (A–C) Contrast-enhanced magnetic resonance imaging (MRI) imaging showing iso to hypointense T1W, iso to hyperintense signals on T2W signals, and postcontrast intense heterogeneous enhancement, respectively, in middle cranial fossa. (D) Postoperative computed tomography scan showing complete excision of tumor.

imaging (MRI), it was suggested sphenoid wing meningioma as heterogeneous contrast-enhancing extra-axial large lesion in middle cranial fossa with parenchymal compression in adjacent frontotemporal lobe. She underwent a surgery (craniotomy and decompression) at peripheral hospital and diagnosed as meningioma, WHO grade I on morphology after clinicoradiological correlation there. But due to worsening symptoms and loss of consciousness, she was admitted to our institution. MRI of the brain revealed a large ovoid mass in middle cranial fossa showing iso to hyperintense signals on T2W and iso to hypointense T1W signals without internal hemorrhage. Postcontrast intense heterogeneous enhancement seen with peripheral areas of necrosis and broad base against falx measured $5.9 \times 5.7 \times 6.4$ cm in size. There was compression/infiltration of parenchyma of adjacent frontotemporal lobes and third ventricle, interpeduncular cistern, sella and prepontine cistern, suggesting recurrent meningioma (atypical; ► Fig. 1).

The patient underwent gross total resection of the lesion; intraoperatively, a large, soft-to-firm, moderately vascular, suckable tumor was present in right sphenoid wing and right temporal lobe. Histopathology showed a highly cellular neoplasm composed of well-formed papillae with central fibrovascular core as well as perivascular pseudorosettes. Pseudorosettes showed broad processes from tumor cell bodies to the adventitia of the central vessels. Tumor cells lining the papillae were polygonal with abundant eosinophilic cytoplasm, round to oval eccentrically placed pleomorphic nuclei, vesicular chromatin, conspicuous nucleoli, and intranuclear inclusions in few of the cells. Foci of necrosis, interstitial hemorrhage and congested

blood vessels without fibrillary background were noted. The mitotic index was 10–12/10 high-power field (HPF). On IHC, tumor cells showed diffuse strong positivity for glial fibrillary acidic protein (GFAP), olig2 and dot positivity for EMA; and negative for IDH1 (R132H), synaptophysin, progesterone receptor (PR) and cytokeratin. MIB-1 labeling index was approximately 30% in the cellular zones (► Fig. 2). Taking into consideration the histopathology and IHC, a final diagnosis of high-grade astroblastoma was offered. Postoperatively, she received radiotherapy due to high grade and on regular follow-up.

Discussion

The term “astroblastoma” is confusing, since the tumor is neither astrocytic nor is it “blastic.” These were initially delineated by Bailey and Cushing in 1926 as a separate glial tumor and further specified by Bailey and Bucy in 1930; however, in the following decades, much uncertainty has arisen regarding criteria for their diagnosis.^{9,10} Histogenesis of these tumors has been clarified recently by histologic and comparative genomic findings but disputes exist regarding its cellular origin and validity as a distinct entity, because it shares features of both astrocytomas and ependymomas in terms of presence of intermediate filament on ultrastructure, showing ependymal differentiation along with positive staining for glial fibrillary acidic protein (GFAP) and S100. Now, these tumors are thought to arise from brain tanycytes, which are a variety of ependymal cells present in the floor of the fourth ventricle with their processes ending on vessels and neurons and are involved in neuropeptide transport.¹¹ The most common clinical symptoms of astroblastoma include headache, seizures, and vomiting, as observed in this case. The studies performed to date show striking female preponderance with a male-to-female ratio of 1:11, similar to our case. Bell et al reported the largest series of supratentorial astroblastomas exclusively located peripherally, as they developed in the cortex, subcortical area, and periventricular area of the cerebral hemispheres, which also happened in our case.¹²

Radiologically, astroblastomas usually appear as well-circumscribed, often solid cystic mass, located near or at the surface of the brain; thus, appearing as extra-axial neoplasms, as in our case, and misdiagnosed. Astroblastoma tends to be isodense to brain parenchyma on the plain CT scan.

Macroscopically, astroblastomas are well-circumscribed, soft lobulated lesions with foci of necrosis and hemorrhage. Bonnin and Rubinstein divided this entity histologically into two groups: low-grade tumors with low-to-moderate mitotic figures, little cellular atypia, uniform perivascular arrangement, minimal or no proliferation of vascular endothelium, and prominent sclerosis of vascular walls; high-grade tumors related to high cellularity, anaplastic nuclear features, elevated mitotic indices, vascular proliferation, and necrosis with pseudopalisading.⁴ Immunohistochemically, astroblastomas are immunoreactive for GFAP, S-100 protein

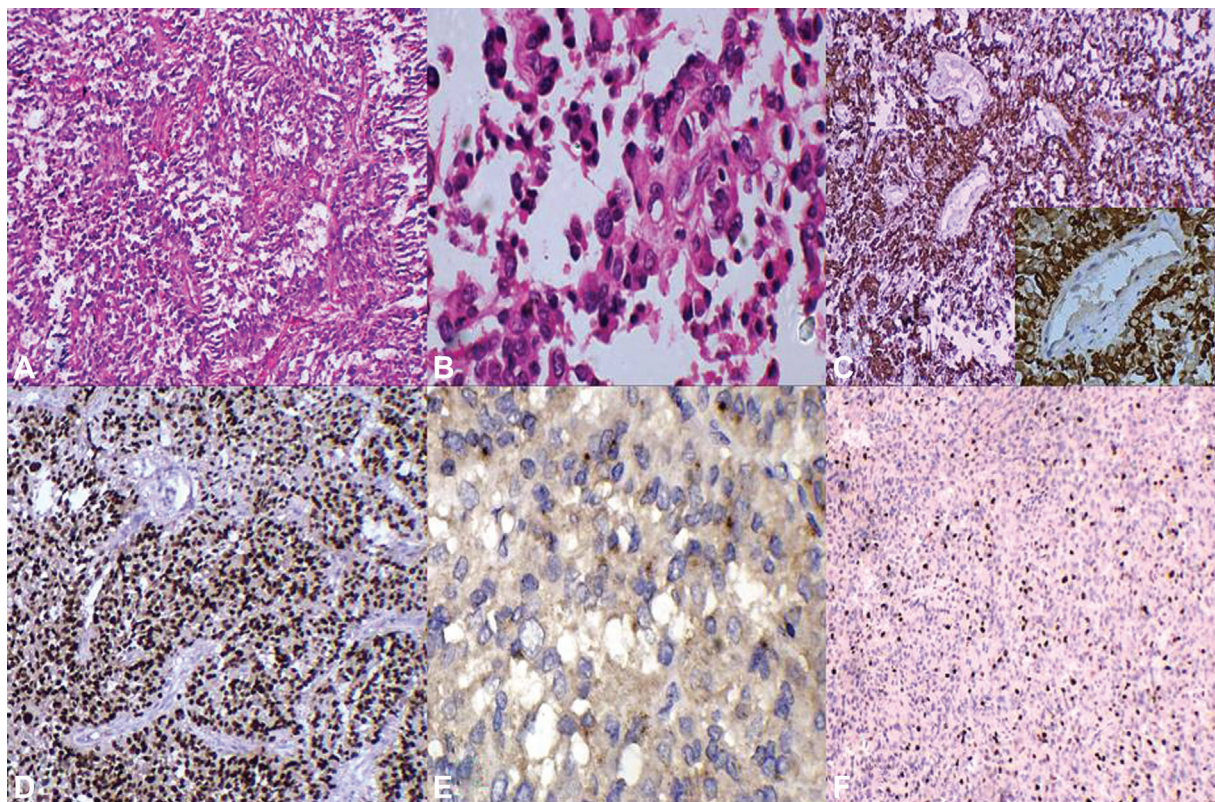


Fig. 2 (A) Papillary and perivascular arrangement of tumor cells (hematoxylin and eosin [H&E], x40). (B) Large polygonal cells with abundant cytoplasm, pleomorphic nuclei with intranuclear inclusion and giant cells (H&E, x400) (C) Cytoplasmic immunoreactivity for GFAP (inset showing perivascular pseudorosette with tumor cells directed toward central blood vessel and lack of fibrillarity; x100, x400) (D) Nuclear immunoreactivity for olig2 (x100). (E) Dot-like immunoreactivity for EMA (x400) (F) High MIB 1 labeling index (30%, x100).

and vimentin. The majority also display at least a focal immunoreactivity for EMA, as seen in current case.

Due to rarity, astroblastomas are difficult to diagnose clinically in early stage. Since tumor descriptions in the literature concern only individual cases or small collections of cases, optimal treatment protocols have not been established. The current consensus is to do surgical excision of the patients with postoperative radiotherapy and chemotherapy for high-grade and recurrent cases as well as regular follow-up with the grade of excision being the major determinant of prognosis.

Conclusion

Case reports like these help in propagation of knowledge on unusual tumors like astroblastomas, with insights on diagnostic and treatment challenges. These studies may help in understanding the key histopathological, immunohistochemical features and treatment options for astroblastomas. To establish a future standard of care, the authors advocate the need for molecular studies to comprehend the properties of this uncommon neoplasm.

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Conflict of Interest
None declared.

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