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Case Report

Astroblastoma: Case Report and Review of Literature

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ABSTRACT

Astroblastoma is a rare and distinct type of aggressive glial tumor for which there is much confusion regarding the diagnostic criteria. We present a case of astroblastoma and review all relevant literature, aiming to discuss astroblastoma from the clinical, pathological, management, and prognostic points of view in an attempt to discover more of its secrets and to introduce a standard approach to its diagnosis and management.

KEYWORDS: Astroblastoma, Brain tumors, Glioma

INTRODUCTION

Astroblastoma is a rare glial tumor of the brain. It is estimated to account for 0.45–2.8% of all primary cerebral gliomas (1,30,31). Astroblastomas are initially described by Bailey and Cushing (3) and further characterized by Bailey and Bucy (2). However, much confusion has arisen regarding criteria for their diagnosis in the following years. Moreover, the origin of these tumors has been debated, because they share common histological characteristics with ependymomas and astrocytomas. Recent studies suggested that astroblastoma is a distinct histological and genetic entity (6, 7, 31, 39). It has a distinctive characteristic of perivascular pseudorosettes with frequent vascular hyalinization. Perivascular pseudorosettes in astroblastoma have short and thick cytoplasmic processes and blunt-ended foot plates (30).

We introduced a case of astroblastoma and reviewed all relevant literature of astroblastoma in order to discuss astroblastoma from the clinical, pathological, management, and prognostic view points. We tried to discover more secrets of astroblastoma and to introduce a standard approach to its diagnosis and management.

CASE REPORT

A forty-year old male patient complained of headache, nausea, vomiting, and deterioration of conscious level with

a gradual progressive course for the last two weeks. He presented with recurrent attacks of vomiting and confusion that rapidly worsened. Neurologically, the patient was confused and uncooperative with recurrent attacks of vomiting and bradycardia but no other neurological symptoms or signs. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a well-demarcated enhancing space-occupying lesion, that was partially cystic, partially calcified in the subependymal area of the left frontal lobe partially projecting into the frontal horn of the lateral ventricle and causing obstructive hydrocephalus (Figure 1). We managed the patient as an urgent case in our intensive care unit (ICU) and performed urgent external ventricular drainage to manage the increased intracranial pressure (ICP). About one week later, we performed surgery for tumor resection by left frontal craniotomy. The tumor was pink-yellow in color, highly vascular, soft in consistency in parts and firm and calcified in other parts and was partially succable and well circumscribed which allowed total resection of the tumor. After the surgery, the patient was transported routinely to the ICU for 3 days. Follow-up CT showed gross total resection (GTR) with mild postoperative brain edema at the site of surgery. The patient was followed up periodically every month for the first 3 months and then every 6 months without postoperative chemotherapy or radiotherapy and without recurrence. After one year, he showed a recurrent mass at the same site as the primary lesion in the follow-up MRI that was managed



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surgically (Figure 2). In the histopathological examination of the specimens, the perivascular pseudorosettes were observed throughout the tumor. Tumor cells were aligned along the fibrovascular stalk by one or two cell layers. Tumor cells were multilayered in some areas of the specimens. Perivascular pseudorosettes extended eosinophilic cytoplasmic processes toward the vessel wall. Cytoplasmic processes were short and thick, of which blunt footplates were attached to the vessels. Multinucleated cells were rarely seen and some of them had atypical nuclei. Cytoplasmic processes of tumor cells were strongly positive for glial fibrillary acidic protein (GFAP).

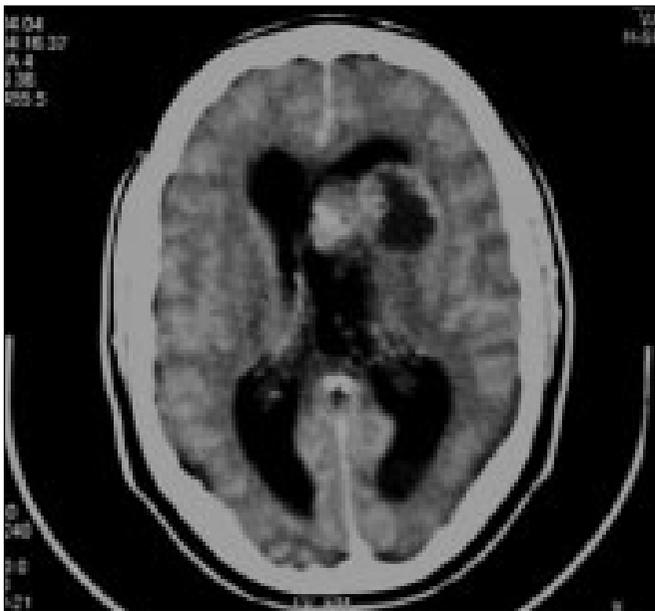


Figure 1: Preoperative MRI with contrast showing astroblastoma.

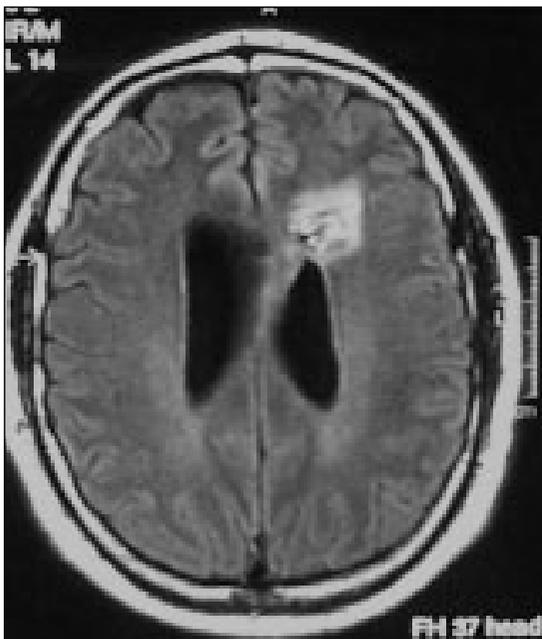


Figure 2: Postoperative MRI T1 with contrast showing recurrent lesion.

Moreover, the tumor cells were also strongly positive for S-100 protein, vimentin and neuron specific enolase (NSE). Focal positivity was present for epithelial membrane antigen (EMA) and CAM (Figure 3).

■ DISCUSSION

Astroblastoma was first described by Bailey and Bucy (2) and some isolated cases were reported afterwards (1,10-13, 17-19, 21, 24, 27, 28, 30, 36, 37, 40). Bonnin and Rubinstein (5) have published general characteristics of astroblastoma based on the review of 23 patients observed by these authors in the last 30 years (12). Brat et al. (7) have also reported 20 patients with astroblastoma. More recently, Navarro et al. (29) reported 8 cases. The presence of astroblastoma as a pure histological entity is still debated (19, 30). However, recent reports have confirmed that it is distinct from the other types of astrocytic glial tumors (5) and it has been distinguished from other gliomas in the recent International Classification of Tumors of the Central Nervous System proposed by the World Health Organization (22, 23, 30, 41, 42).

The epidemiology of astroblastoma is difficult to determine exactly as most reports are case reports. However, in some reports it was estimated as a rare glial tumor to represent only 0.45-2.8% of all neuroglial tumors (30). We tried to collect all cases of astroblastoma reported in the literature consisting of approximately 120 cases reported in about 75 years and we found statistically that the incidence of astroblastoma is about 1.6 new patient/year and in turn this 2.8% prevalence of all primary brain tumors is so high which is found statistically also about 0.004% of primary brain tumors in the collected data. Although our patient is 40 years old, astroblastomas can occur at any age, but in general mostly occur during the first 3 decades of life. Congenital forms have been also described by many authors (6, 7, 20, 30, 39). We found statistically that the mean age of astroblastoma is 14.5 years and the age range is 0- 58 years as documented also by many authors (5, 7, 38); moreover it is seen most commonly at an age of less than 5 years as reported also in the Navarro series (29),

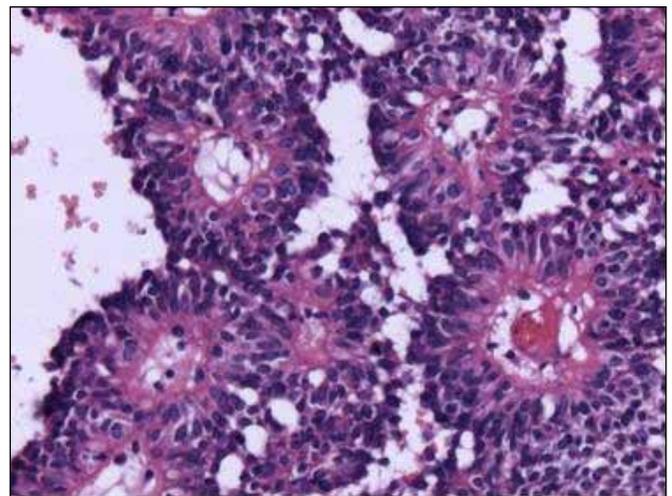


Figure 3: Specimen of an astroblastoma with HE stain, x200 magnification.

in contrast with Husain and Lesstma (17) who found this ratio was unusual. In the two large case series, no predominance between the 2 sexes was reported (8, 26, 31) in contrast to other authors (6, 7, 29). Although our patient is male, we actually found a female predominance (female/ male ratio was about 1.7/1). Astroblastoma typically presents as a spherical tumor located in the cerebral hemispheres (12) but it may also be observed in the corpus callosum (14,16), cerebellum (32), brainstem (35), optic nerve (35), cauda equina (16), and the third and fourth ventricles (29).

Astroblastoma generally develops in the cortex, subcortical area, and periventricular area of the cerebral hemispheres (15). It does not develop in the ventricle itself (26) as present in our patient. The frontal lobe is the most common site of its presentation followed by the parietal lobe.

Considerable confusion has surrounded the diagnosis and cell origin of astroblastoma since its original description as a separate glial tumor by Bailey and Cushing (3). It is now generally believed that astroblastomas are indeed a distinct type of neoplasm characterized by a constellation of histologic findings, each nonspecific in itself, but when taken together lead to the diagnosis (6). Microscopically; astroblastomas and ependymomas have a similar histologic appearance characterized by a radiating arrangement of spindle-shaped tumor cells forming perivascular pseudorosettes (8). These pseudorosettes are similar to those seen in ependymomas, and histopathologic distinction between these two entities is often difficult (30, 31, 38, 40). Distinguishing histological features from ependymoma are;

- 1) Short and robust nature of the processes that constitute perivascular pseudorosettes in astroblastoma, in contrast to the typically compact intravascular architecture of ependymoma,
- 2) Hyalinization of vessels,
- 3) Rarefied spaces between pseudorosettes,
- 4) Absence of abundant fibrillary pattern,
- 5) Difference in nuclear characteristics (9, 12, 21, 22, 33)

Pathological grading of astroblastoma is classified into low and high grade. The low-grade type includes astroblastomas with uniform perivascular arrangement of pseudorosettes, low to moderate number of mitotic figures, little cellular atypia, minimal or no vascular endothelial proliferation, and predominant sclerosis of the vascular walls. High-grade astroblastomas show high cytological atypia, increased mitotic figures, compact cellularity, perivascular cells with high mitotic rates, and hypertrophy of vascular endothelium without hyalinization (5, 12, 21). This has prognostic value, as patients with a low-grade astroblastoma survive for a longer time than those with the anaplastic form. The recurrence rate is very low and commonly occurs within 4.5 years (1, 5, 38, 40) in contrast to our patient who was classified as low grade astroblastoma but had a recurrence after about 1 year with the same pathology despite GTR at the first surgery. This indicates that low-grade astroblastoma may also recur rapidly and that even if the tumor behaviour is more benign, it is still aggressive enough to cause recurrence.

Astroblastoma shows a characteristic appearance on neuro-radiological images. The typical appearance of this tumour is a large, well-delineated, lobulated, superficial, solid, and cystic mass with inhomogeneous contrast enhancing and little vasogenic edema. The solid component of the mass shows a bubbly appearance. The cystic component gives the same signal as cerebrospinal fluid. Punctate calcification may be present. Sometimes astroblastoma resembles an extra-axial neoplasm on MRI, leading to a preoperative misdiagnosis (4, 6,11,12).

The solid portions of the masses are relatively hypointense to gray matter on T1-weighted images and isointense to gray matter on T2-weighted images. Given the large size of the masses, a relative lack of peritumoral T2 hyperintensity exists, representing vasogenic edema or tumor infiltration or both in the surrounding brain parenchyma. Enhancement is heterogeneous, with rim enhancement of the cystic portion and heterogeneous enhancement of the solid portion. CT occasionally depicts punctate calcifications within the solid portion of these tumors; the solid portion can also appear hyperattenuated (6, 31) (Figures 1,2).

The optimum management of this tumor remains in doubt. A total removal, in view of the well demarcated nature, and radiotherapy seems to be the advisable treatment (12, 13, 34). It appears that most neurosurgeons would attempt tumor clearance and administer postoperative radiotherapy (5, 10, 30, 34). In the series by Bonnin and Rubinstein (5), complete surgical excision was achieved in 12 of 23 patients. Eleven patients received radiotherapy and 5 chemotherapy settings. It is of note that the only patient to have radiotherapy following a diagnostic biopsy was alive 12 years after diagnosis, suggesting a benefit for radiation therapy (30, 40).

The behaviour of astroblastoma is unpredictable and the prognosis may be complicated by the potential behaviour of the astroblastoma to convert into a more malignant variant of glioma (13). The prognosis of the astroblastoma seems to place it between the astrocytoma and glioblastoma (5). The low-grade astroblastomas are thought to have better prognosis than the high-grade astroblastomas as we noticed from the follow-up results of our patient and as suggested in other articles. Gross total resection may result in long-term survival. An anaplastic histology has been associated with recurrence and progression, suggesting that more aggressive treatment, including radiotherapy, is necessary for high-grade lesions (5, 7, 21, 25, 29, 34, 38). Combination chemotherapy has had encouraging success regarding the prognosis (30). We found the overall survival of all patients to be 2.4 years and the recurrence rate in the literature to be 34%. As we previously diagnosed our patient as low grade astroblastoma, we found the recurrence of the tumor after about 1 year to suggest aggressive behaviour of the tumor.

■ CONCLUSION

Astroblastoma is a distinct type of aggressive glial tumor that commonly presents in the frontal and parietal lobes. The recurrence rate is 34% after surgical resection and radiotherapy, with an overall survival of 2.4 years.

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