

Supratentorial High Grade Astroblastoma: Report of two Cases and Review of the Literature

Supratentoryal Yüksek Dereceli Astroblastom: İki Olgu Sunumu ve Literatürün Gözden Geçirilmesi

ABSTRACT

Astroblastoma is a rare glial tumor with uncertain histopathological origin and unpredictable clinical behavior. In this report, the authors present two cases of high grade intracerebral astroblastomas. Both tumors occurred in children as supratentorial, well-circumscribed, peripheral masses. The lesions differed radiographically; one contained a huge cystic component and heterogeneously enhancing mural nodule while the other appeared as a prominently contrast-enhancing solid mass lesion. Both patients were treated with surgery and postoperative radiotherapy. They were followed-up long-term and no recurrence of the tumor was detected in either case. We also discussed the radiological and histological characteristics with prognostic features.

KEYWORDS: Astroblastoma, Glial tumor, High grade, Pathology, Radiology

ÖZ

Astroblastom, histopatolojik kaynağı belli olmayan ve klinik gidişatı tam olarak öngörülemez nadir bir glial tümördür. Bu yazıda, yüksek dereceli iki intraserebral astroblastom olgusu sunulmuştur. Her iki tümör olgusu da, çocuk yaşta, supratentoryal, iyi sınırlı, periferik yerleşimli kitle şeklinde görülmüştür. Lezyonlar farklı radyolojik bulgular göstermiştir: İlk olguda büyük kistik bir bölüm ile heterojen kontrast tutan mural nodül görülürken, ikinci olguda belirgin kontrast tutan solid kitle tespit edilmiştir. Her iki hasta cerrahi olarak tedavi edilmiş ve postoperatif dönemde hastalara radyoterapi uygulanmıştır. Olgular uzun dönem takip edilmiştir ve iki vakada da nüks ile karşılaşılmamıştır. Ayrıca, tümörün radyolojik ve histolojik özellikleri ile prognostik değerleri tartışılmıştır.

ANAHTAR SÖZCÜKLER: Astroblastom, Glial tümör, Yüksek dereceli, Patoloji, Radyoloji

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INTRODUCTION

Astroblastomas are distinct and uncommon neuroepithelial tumors of uncertain origin according to the current WHO classification of central nervous system tumors (2,11). Only 0.45-2.8% of all neuroglial tumors have been estimated to be astroblastoma (13, 18). They can be easily misdiagnosed as they are rarely encountered and share common radiological and histopathological appearances with other glial neoplasms. We present two pediatric astroblastoma cases and aim to discuss the radiological characteristics and management of this rare entity in this report.

CASE REPORTS

Case 1

A 6-year-old girl was admitted with complaints of nausea, vomiting, loss of balance and frequent falls, started within a one-month period. She had also been complaining of decreased sensation on the left side of her body recently. Neurological examination revealed slight global left hemiparesis and left central facial nerve palsy. The deep tendon reflexes were hyperactive on the left including a positive left Babinski sign. She also had horizontal nystagmus.

On cranial magnetic resonance imaging (MRI), a huge right frontoparietal mass, hypointense on T1WI and hyperintense on T2WI, was detected. The lesion was prominently cystic on T2WI and contained solid parts that were enhancing heterogeneously on the posterior portion of the mass after gadolinium injection (Figure 1A,B).

She underwent a right frontoparietal craniotomy and the lesion was gross totally resected. Postoperative period was uneventful and neurological examination revealed no additional deficit. The histopathological diagnosis was consistent with astroblastoma grade IV. Microscopically, the tumor showed expansile growth pattern with perivascular pseudorosettes formed as tumor cell processes converge on vessels. There was perivascular hyalinization in focal areas. Immunohistochemically, the tumor cells showed GFAP, EMA, cytokeratin and synaptophysine positivity. The mitotic activity was increased with a Ki-67 labeling index of 7% (Figure 2A,B,C,D,E,F).

The patient received postoperative radiotherapy. She was followed up for 20 months postoperatively and had no neurological deficit or complaints at her last visit. The control MRI at postoperative 20 months showed no pathological contrast enhancement on the surgical field.

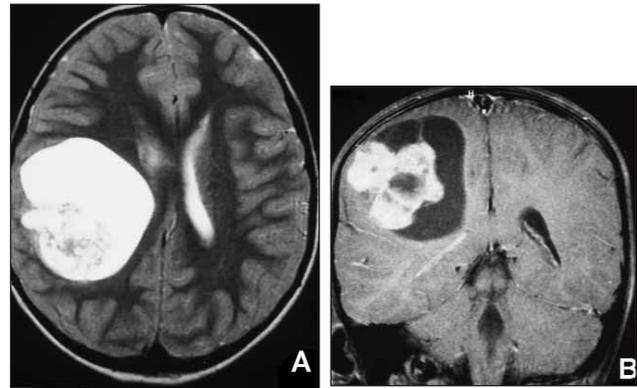


Figure 1: A) Axial T2WI shows the cystic mass in the right frontoparietal area. B) Gadolinium-enhanced coronal T1WI shows contrast-enhanced solid component.

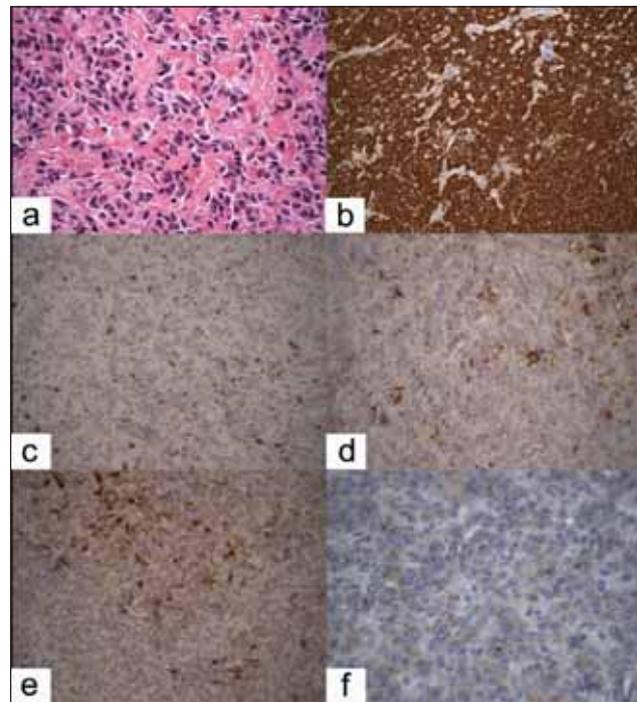


Figure 2: The histopathological microphotographs show A) perivascular hyalinization in focal areas (H&E x400) and B) GFAP (x100), D) EMA (x200), E) cytokeratin (x200) and F) synaptophysin (x400) positivity of the tumor cells. C) The mitotic activity and ki67 immunoreactivity were high (x200).

Case 2

A 7-year-old girl presented with a history of a complex partial seizure one month ago. She was referred for surgery upon detection of an intracranial mass lesion. Neurological examination revealed no abnormal findings. Cranial MRI revealed the solid mass lesion as hypointense in T2WI and hypointense on T1WI with prominent gadolinium enhancement (Figure 3A,B). Additionally, cranial computed tomography (CT)

scans showed calcified mass lesion with vasogenic edema on the right parietal lobe (Figure 3C).

A right parietooccipital craniotomy was performed. A rubbery solid, well-encapsulated but vascular tumor was resected totally. The tumor was histopathologically diagnosed as astroblastoma grade III. Microscopically, the tumor showed palisading necrosis. Immunohistochemical stains revealed GFAP and EMA positivity. Ki-67 labeling index was 5%.

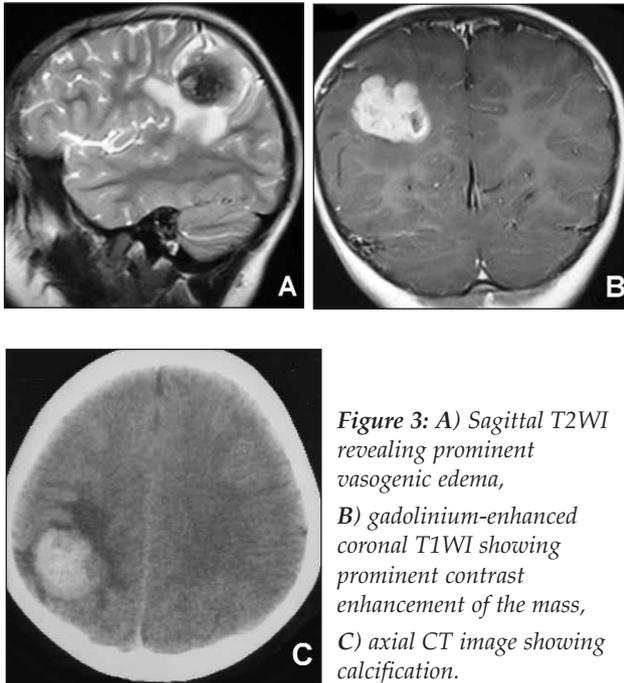


Figure 3: A) Sagittal T2WI revealing prominent vasogenic edema, B) gadolinium-enhanced coronal T1WI showing prominent contrast enhancement of the mass, C) axial CT image showing calcification.

The patient was discharged on the 4th postoperative day without any neurological deficit. She was referred to the oncology clinic for adjuvant radiotherapy. The patient was followed up for 12 months and found to be normal on control physical examination.

DISCUSSION

Astroblastomas are classically located in the cerebral hemispheres. However, tumor invasion has also been reported in the corpus callosum, cerebellum, brain stem and optic nerves (8,18). Clinical signs and symptoms depend on the localization and size of the neoplasm with headache and seizures being the most frequent (12). The first case in this report presented with the classical signs of mass effect, whereas the second one had a history of complex partial seizure. Astroblastomas are mostly seen in young adults, but congenital cases have also been reported (13). Controversy exists on the sex preference of astroblastoma. No preference (3) or remarkable female preponderance (2,4) were reported

in different studies. Both cases reported in this article were also female.

Bell et al. (2) reported the largest imaging series with 12 cases on astroblastomas. As identified in their report, the lesions are almost exclusively supratentorial and peripherally located with both solid and cystic components. They are well demarcated with multiple intratumoral cysts giving it a bubbly appearance. The lesion typically shows rim enhancement on CT and T1W MRI. However, heterogenous gadolinium enhancement may be seen. Less common findings are intratumoral hemorrhage and intraventricular location. Calcification is a consistent imaging feature seen in most reported cases. Imaging of our first case showed typical solid and cystic parts of the tumor without calcification. However, the second case presented with a strongly enhancing solid mass without a cystic component that is usually characteristic for astroblastomas. The CT scan of the second case also showed calcification that was seen hypointense on T2WI. Based on imaging, the differential diagnosis of astroblastoma includes high grade astrocytoma, oligodendroglioma, primitive neuroectodermal tumor, ependymoma, meningioma, and atypical rhabdoid tumor (1,14,17). In a recent report, Eom et al. suggested considering astroblastoma in the differential diagnosis of an extra-axial mass, especially in a young patient (9). Peritumoral edema is usually present in high-grade glial tumors, while astroblastomas have relatively little peritumoral hyperintensity on T2WI despite their large size (4). Oligodendrogliomas may be large with foci of cystic degeneration and nodular calcifications, in contrast to the punctate calcifications seen in astroblastomas (14).

Astroblastomas are defined histologically by the presence of perivascular pseudorosettes and prominent perivascular hyalinization (3). Histologically, astroblastomas may resemble astrocytic tumors, ependymomas, and non-neuroepithelial tumors due to their astroblastic aspects (5,16). Lack of fibrillarity is an essential feature in distinguishing astroblastomas from other glial neoplasms (4).

No definitive cell of origin has been identified yet (2). It may arise from abnormally persisting groups of embryonal precursor cells such as tanycytes, a transitional cell type between astrocyte and ependymal cell during embryogenesis (10, 15). Immunohistochemically, astroblastomas are immunoreactive for GFAP, S-100 protein and vimentin (11). The majority displays a focal cytoplasmic immunoreactivity for EMA (12). Bonnin et al reported two distinct histological types: a

low grade type in which a better differentiated pattern was apparent and a favorable postoperative prognosis may be expected and a high grade type, showing more anaplastic microscopic features, in which postoperative survival was usually short (3). High-grade lesions show focal or multifocal regions of high cellularity, anaplastic nuclear features, elevated mitotic indices, vascular proliferation and necrosis with pseudopalisading (4). Our cases were considered to be in the high-grade group as they had all these features, especially palisading necrosis and vascular hyalinization, together with a characteristic Ki-67 labeling index (7% in case number one and 5% in case number two).

Although malignant astroblastomas may show infiltration of brain parenchyma, most of them are non-infiltrating (5). Total resection is reported to be the best way of treating an astroblastoma (6). Close follow up of all cases and adjuvant therapy for high grade and recurrent cases is recommended (19). The more favorable prognosis is almost invariably associated with circumscription of the tumor that might permit the total resection of tumor in all grades (3). Radiological evidence of a tumor response to chemotherapy has been reported only by Pizer et al. (13). In the series of 23 patients by Bonnin and Rubinstein the only patient who did not receive postoperative radiotherapy had the shorter survival time (3). Caroli et al. reported a high-grade astroblastoma with a five year survival without recurrence after total resection, radiation therapy and temozolomide usage (7). The two patients in this report received postoperative radiotherapy. No recurrences were noted on their last follow-up (20 months for the first case and 12 months for the second case).

In conclusion, two cases of high-grade supratentorial astroblastoma were described in this report. As the radiological features of these rare lesions may be heterogenous, this entity should be remembered in the differential diagnosis of supratentorial intraaxial tumors. Furthermore, as for all malignant tumors of glial origin, we suggest total resection and postoperative adjuvant radiation therapy for high-grade astroblastomas.

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