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CASE-BASED UPDATE

Astroblastoma in a child

Ekrem Unal · Yavuz Koksal · Istvan Vajtai · Hatice Toy · Yalcın Kocaogullar · Yahya Paksoy

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Abstract
Background Astroblastoma, an uncommon neuroepithelial tumor, typically presents in young adults as a well-circumscribed cortical or subcortical spherical mass. Astroblastoma may cause a diagnostic problem to anyone unfamiliar with its architectural and histological features.
Case history We report the case of a 4-year-old boy who was referred for complaints of progressive deficits of balance and difficulty with walking during the previous 3 months. A large fronto-parietal cystic mass with solid mural nodule was discovered. Total removal of the tumor mass was performed, and a diagnosis of high grade (malignant) variant of astroblastoma was made. Postoperatively, the patient received radiation therapy, for a period of 11 weeks, followed by chemotherapy. He is in a good neurological recovery without any evidence of recurrence for 8 months.
Prognosis The best treatment modality for astroblastoma is surgical resection if possible, whereas adjuvant therapy (radiotherapy and/or chemotherapy) can be considered in high-grade astroblastomas, with a close follow-up for all cases.

Keywords Astroblastoma · Brain tumor · Children

E. Unal (✉)
Department of Pediatrics, Meram Faculty of Medicine,
Selcuk University,
42080 Meram, Konya, Turkey
e-mail: drekremunal@yahoo.com.tr

Y. Koksal
Department of Pediatrics, Division of Pediatric Oncology,
Meram Faculty of Medicine, Selcuk University,
Konya, Turkey

I. Vajtai
Neuropathology Service, Institute of Pathology,
University of Bern,
Bern, Switzerland

H. Toy
Department of Pathology, Meram Faculty of Medicine,
Selcuk University,
Konya, Turkey

Y. Kocaogullar
Department of Neurosurgery, Meram Faculty of Medicine,
Selcuk University,
Konya, Turkey

Y. Paksoy
Department of Radiology, Meram Faculty of Medicine,
Selcuk University,
Konya, Turkey

Introduction

Astroblastoma is an extremely rare glial tumor, which typically presents in young adults as a well-circumscribed cortical or subcortical spherical mass in the cerebral hemispheres, with intratumoral cyst-like changes. It is classified as an uncommon neuroepithelial tumor of uncertain origin by the International Classification of Tumors of the Central Nervous System proposed by the World Health Organization [6]. The characteristic histological features of astroblastoma are the presence of typical astroblastic perivasculer pseudorosettes and perivasculer hyalinization. Because a similar histological pattern may be seen in astrocytomas and in glioblastomas, astroblastoma may cause a diagnostic problem to anyone unfamiliar with its architectural and histological features. In addition to this diagnostic challenge, because of the rarity of astroblastomas, there is no consensus regarding their optimal management.

Clinical features

Clinical signs and symptoms depend on the location of the tumor, size, and mass effect of the neoplasm like such other tumors of the brain [7]. This tumor commonly occurs during the first three decades of life, without a sex, race, or familiar predominance [4]. Meanwhile, Brat et al. [3] presented a female predominance in their series (16 women, 4 men) that has not been noted in other studies [2, 8].

Diagnostic investigations

Radiological features

Neuroimaging features of astroblastomas have been reported as generally large, peripheral, solid, and cystic masses with a characteristic bubbly appearance in the solid component and relatively little associated peritumoral T2 hyperintensity for their large size. On computerized tomography scans, astroblastomas may appear slightly higher in attenuation and often have punctuated calcifications [9].

Histology

The entity of "astroblastoma" was firstly announced to English medical literature by Bailey and Bucy [1]. Since the time of first description, clouds of confusion have surrounded the diagnosis, histological origin, and classification of this uncommon tumor. The questions raised for this tumor have been eliminated by reported studies from the middle of the 1980s until today [2–5, 7–11].

Firstly, the term of "astroblastoma" is not definitive in itself because these tumors are not overtly astrocytic, nor are they blastic [3]. Bailey and Bucy [1] believed that astroblastoma originated from the astroblast, an intermediate stage between glioblasts and astrocytes. Conversely, in an in vitro study performed by Rubinstein and Herman [10], it was reported that astroblastoma had originated from the tancycte, a precursor cell that is normally found along the ependymal lining of the embryonel and mammalian brain but is distinct from the epithelial ependymal cell. Recently, Kubota et al. [5] reported that astroblastoma had unique histological features and also stressed that astroblastoma should have been categorized as a specific type of neuroepithelial tumor. Astroblastomas are classified into low grade (well-differentiated) and high grade (anaplastic) according to histological features [2]. The high-grade type consists of tumors with more anaplastic features based on the presence of focal or multifocal regions of high cellularity, anaplastic nuclear features, elevated mitotic indices, vascular proliferation, necrosis with pseudopalisading, and anaplasia with loss of astroblastic architecture [3]. Some of the characteristic histological features of astroblastoma have been reported as the formation of perivascular astroblastic pseudorosettes, absence of true rosettes, looseness of architecture causing pseudopapillae, vascular attachment of the cell main process, lack of an epithelial-free surface differentiation, regional hyaline changes, and pushing borders in regard to the adjacent brain [3, 8]. In a comparative genomic hybridization study performed by Brat et al. [3], astroblastomas were found to be dissimilar to those that have been reported most commonly for ependymomas or astrocytomas. Focusing based solely on the histological finding of perivascular orientation of tumor cells, anaplastic oliodendrogioma, anaplastic astrocytoma, infiltrating fibrillary astrocytoma, and glioblastoma might be misdiagnosed as astroblastoma. The perivascular process of astroblastomas is not fibrillary, nor is there stromal fibrillarity. It should be kept in mind that the lack of fibrillarity is an essential feature in distinguishing astroblastomas from other glial neoplasms [3]. Immunohisto-

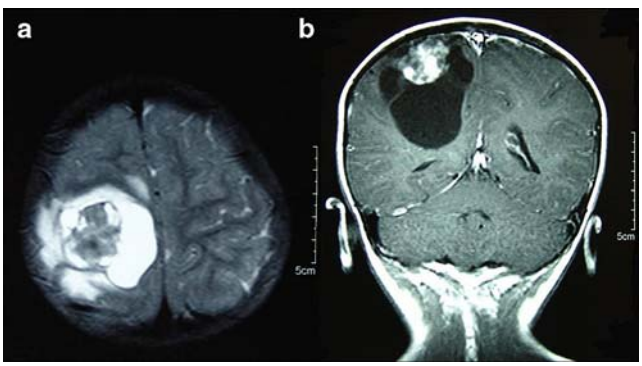

Fig. 1 a T_2-weighted axial MR image shows a mass with solid and cystic components. There is little edema around the mass. **b** Contrast-enhanced T_1-weighted MR image shows contrast enhancement in the solid portion

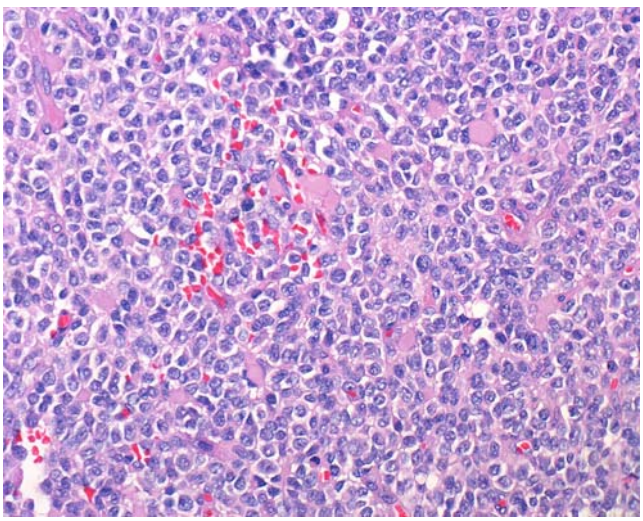

Fig. 2 The tumor cells tend to be cuboidal to slightly elongated (hematoxylin and eosin stain, 400×)

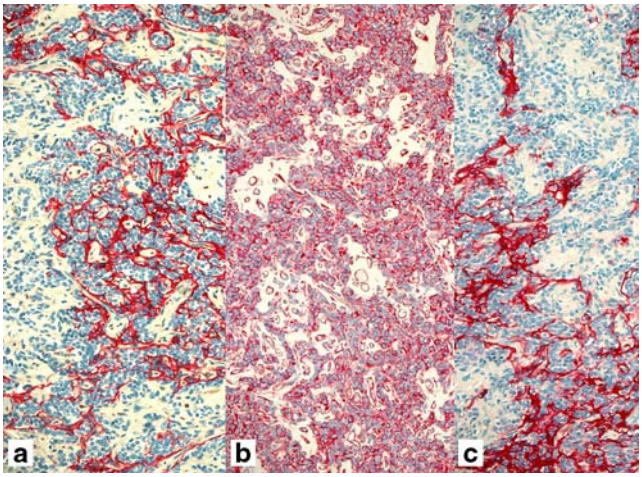

Fig. 3 The tumor was immunoreactive for **a** GFAP (200×), **b** vimentin (200×) and **c** S-100 (200×)

chemically, astroblastomas are strongly immunoreactive for glial fibrillary acidic protein (GFAP), S-100 protein, vimentin, and neuron-specific enolase and focal cytoplasmic immunoreactive for epithelial membrane antigen (EMA) [3, 8].

Prognosis and management

The rarity of astroblastoma precludes its definitive therapeutic studies. A variety of therapeutic strategies such as gross total, near total, or subtotal resection, radiotherapy, and chemotherapy were used among the patients with astroblastomas. In addition, low-grade astroblastomas are often well circumscribed making gross total resection both possible and potentially curable [2, 11]. Several authors have managed the patient with radiation therapy as a first option for adjuvant therapy [2, 3]. Postoperative multimodal therapy consisting radiotherapy and/or chemotherapy have also been experienced in high-grade astroblastomas [4, 8, 11]. These regimes can be used based both on the pathologic diagnosis and the patient's age [8]. Given the small number of the participant and the variety of regimens used in these series, no consensus for the exact modality of the management for the astroblastomas can be drawn.

Prolonged disease-free survival can be expected after gross total resection in managing well-differentiated astroblastoma. High-grade astroblastomas seem to be associated with poor prognosis, and recurrences are more expectable [2, 3, 11]. Long-term survival depends on tumor location, extent of resection, and response to adjuvant therapy [2, 3]. Recurrence may occur in the initial site of the disease, whereas late neuraxis dissemination was also shown [2, 11].

After a review of the literature regarding astroblastoma, we believe the best treatment modality for astroblastoma is surgical resection if possible, whereas adjuvant therapy (radiotherapy and/or chemotherapy) can be considered in high-grade astroblastomas, with a close follow-up for all cases.

Case illustration

A 4-year-old boy was admitted to our hospital with complaints of progressive deficits of balance and difficulty with walking during the previous 3 months. His medical history was unremarkable. On neurological examination, mild left-side hemiparesis with left motor weakness was detected. The other physical findings were within normal limits. Complete blood count, serum biochemistry, and urine analysis were also within normal limits. Magnetic resonance imaging showed a large cystic mass with a solid mural nodule (Fig. 1a). There was little parenchymal edema around the mass. A solid portion of the tumor showed heterogeneous contrast enhancement. The mass was located in the fronto-parietal region (i.e., perirolandic area; Fig. 1b).

For total removal of the tumoral mass, right parietal craniotomy was performed. He was discharged on the ninth day after the operation without any sequel. On histopathological examination, the tumor was markedly hypercellular, infiltrating adjacent brain parenchyma. A solid architecture, focally disrupted by pseudopapillary dehiscences, was present. Individual tumor cells tend to be cuboidal to slightly elongated, including occasional fibrillary processes. With very moderate anisokaryosis, however, fair numbers of mitosis were encountered. Growth pattern included confluent sheets and pseudorosette-like polarized aggregates of tumor cells (Fig. 2). On immunohistochemistry, there was strong and diffuse immunoreactive of vimentin and the S-100 protein, membranous straining for CD56 (NCAM), GFAP, and synaptophysin were also observed (Fig. 3a, b, and c). Conversely, epithelial markers such as pancytokeratin, low-molecular-weight cytokeratin, CAM 5.2, EMA, and CD 99 were negative. A diagnosis of a high-grade (malignant) variant of astroblastoma was made. Postoperatively, the patient received radiation therapy, 5,400 cGy, to the whole brain with ^{60}Co for a period of 11 weeks, followed by chemotherapy, including cisplatin (100 mg/m^2 per day, day 1) and etoposide (100 mg/m^2 per day, days 1–3). He is still under out-patient follow-up without any evidence of recurrence for 8 months.

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