CASE REPORT

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A Case of Astroblastoma

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An astroblastoma is a rare primary glial tumor occurring preferentially in young adults. It is characterized by a perivascular arrangement of the tumor cells forming perivascular pseudorosettes mimicking ependymomas. The histogenesis of astroblastoma is unclear, despite a number of studies to determine its possible cellular origin. We have experienced a case of astroblastoma located at the temporal lobe. It presented as a large, well-circumscribed, and highly enhanced mass lesion on magnetic resonance images(MRI). The tumor was well demarcated and did not infiltrate the brain, which made complete removal possible. Here, we report and discuss the characteristic histological and radiological features of this case.

KEY WORDS : Astroblastoma · Histological study · Glioma.

Introduction

An astroblastoma is a rare glial tumor. These tumors were initially described by Bailey and Cushing and further supported by Bailey and Bucy[1]. Generally, it is a solid, well-circumscribed tumor defined histologically by the presence of astroblastic pseudorosettes and prominent perivascular hyalinization[1-4,6,7,9,10]. However, the classification of astroblastoma as a distinct disease and the nature of its cell of origin have been the subjects of much debate. Recent data from clinicopathological and chromosomal studies have indicated that it should be regarded as distinct from other types of glioma[3,4,6,7,9,10]. We have experienced a case of astroblastoma located at the temporal lobe. In this patient, the tumor could be resected en bloc. We describe its clinical and radiological features, and also discuss its histopathological characteristics.

Case Report

A 25-year-old female patient presented with persistent headache of one-month's duration. The patient had no other symptoms and neurological examinations were normal. Magnetic resonance imaging(MRI) showed a well-circumscribed, solid mass lesion in the right temporal lobe, which was highly enhanced after administration of gadopentetate dimeglumine (Fig. 1). On T2-weighted MRI, the mass appeared heterogeneous and bubbly, and was surrounded by edema (Fig. 1B). The tumor was highly vascularized, as seen on angiography of the right carotid (Fig. 2). The patient underwent a right temporal craniotomy (Fig. 3). The dura was cut open to expose the surface of the tumor, which was pinkish in color (Fig. 3A). We opened the arachnoid membrane along the exposed tumor margin and observed that there was a border of tissue separating the lesion from the adjacent brain tissue. We performed circumferential dissection along the border, as we would do in a patient with arteriovenous malformation. The tumor protruded from the brain parenchyma at the end of the dissection (Fig. 3B). There was no macroscopic evidence of infiltration into the brain. The postoperative course was uneventful. The patient underwent a seven-week course of whole-brain radiation therapy (4,500cGy). There was no evidence of tumor recurrence at the seven-month follow-up MRI.

Histological and immunohistochemical analyses

Perivascular pseudorosettes were observed on hematoxylin and eosin staining (Fig. 4A). The tumor appeared uniform histologically, with no areas of transition to an astrocytoma or a glioblastoma. Perivascular hyalinization or sclerosis was observed. There was no appreciable mitotic activity. On immunohistochemical analysis, the tumor cells were positive

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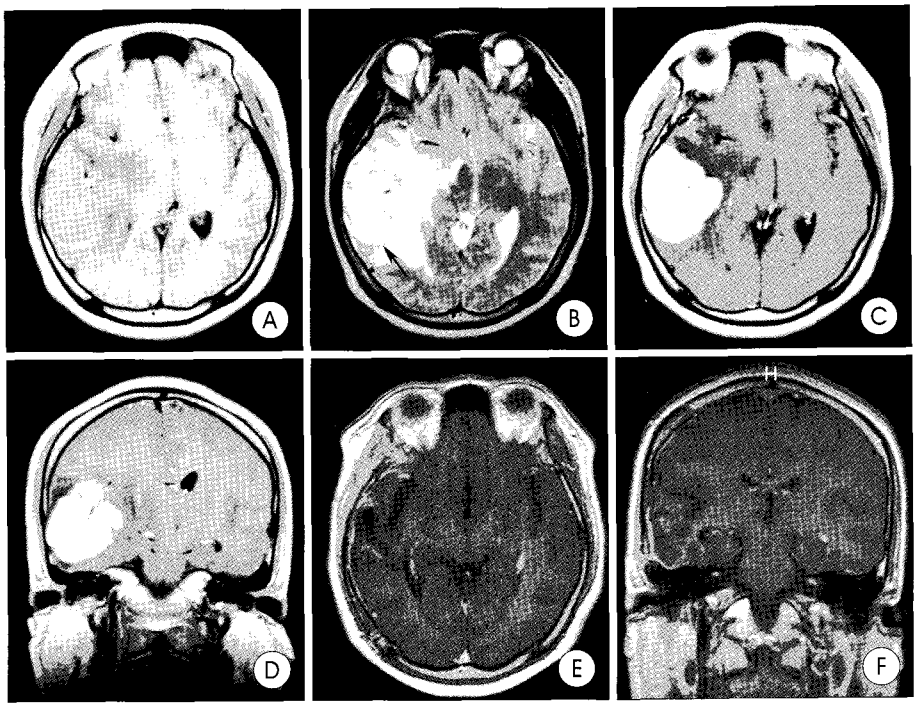

Fig. 1. A : T1-weighted magnetic resonance imaging showing a mass lesion isointense to gray matter in the right temporal lobe. B : T2-weighted axial magnetic resonance imaging showing the heterogenous bubbly appearance of the posterolateral portion of the lesion, and marked peritumoral hyperintensity. C, D : Axial and coronal postcontrast magnetic resonance imaging revealing intense, relatively homogenous enhancement of the lesion. D, E : Axial and coronal postcontrast magnetic resonance imaging obtained 10 months after operation showing no evidence of recurrence.

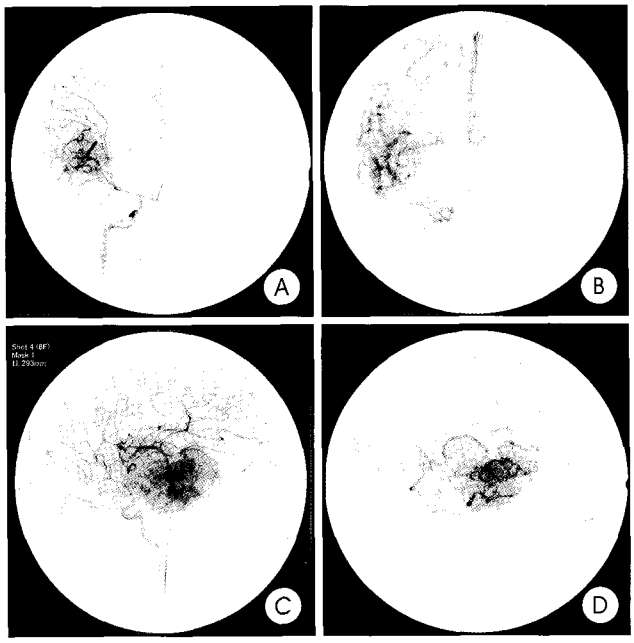

Fig. 2. Right carotid angiograms with anteroposterior (A, B) and lateral (C, D) views showing the high vascularity of the tumor. Arterial phase angiograms showing dilated arteries feeding the lesion and also showing a uniform distribution of fine neoplastic vessels (A, C). The main draining veins are superficial veins (B, D).

for glial fibrillary acidic protein(GFAP), S-100 protein, neuron-specific enolase(NSE), cytokeratin 7, and vimentin (Fig. 4B, C, D, F), but they were negative for synaptophysin. The Ki-67 labeling index was less than 2%.

Discussion

An astroblastoma is a rare glial tumor accounting for 0.45~2.8% of brain gliomas[8,9]. The tumor is generally described as : (1) well-circumscribed and located in the cerebral hemispheres, (2) occurring in young adults, but occasionally reported in children, and (3) having a papillary pattern characterized by perivascular pseudorosettes and hyalinization on histological examination[2-4,6-10]. The tumor was originally described by Bailey and Bucy[1], at a time when astroblasts were thought to be the immediate progenitors of astrocytes. This is an obsolete conception of brain histogenesis and, in view of our current knowledge on glial progenitor cells, astroblasts are unlikely to exist[3,6,7,9]. Finally, the term "astroblastoma" can be regarded as misleading in itself, since these tumors are not overtly astrocytic; nor are they "blastic"[3,8].

In their studies, Rubinstein and Herman[10] postulated that astroblastoma cells are related to tanycytes, which are integral to the ependyma of submammalian species and have features intermediary between those of astrocytes and ependymocytes. These authors suggested that cells similar to tanycytes might exist transiently during normal human embryogenesis, and that astroblastoma may be derived from such embryonal precursor cells that have abnormally persisted[10]. Although a tanycytic origin has been postulated by them, at the present time, tanycytes and ependymocytes are believed to represent a unique cell population[3]. Thus, the cellular origin of astroblastomas remains unknown.

The papillary pattern of astroblastomas can be seen in other tumors such as ependymoma, papillary meningioma, and in metastases from papillary carcinoma[2-4,7,8,10]. It also occurs focally in other types of glioma[2,3,8,11]. In our patient, papillary perivascular pseudorosettes were distributed diffusely throughout the tumor (Fig. 4A). On immunohistochemical examination, these could be differentiated from metastases of nonneuroglial tumors, which are negative for GFAP, NSE, and S-100. However, the tumor was not easily differentiated

from ependymoma, which shows a similar histopathological pattern. The absence of continuity with the ventricular system and histological differences in organization patterns can be helpful in differential diagnosis[3,6,8,9,11]. In astroblastomas, rarified spaces can usually be seen between the pseudorosettes histologically[1-4,6,8,10,11]. In contrast, the intravascular architecture of ependymomas is typically compact[2-4,7,8,11]. Additional features that differentiate these tumors include differences in nuclear characteristics and in the thickness of the perivascular cytoplasmic process[2,3,7,8]. Brat and coworkers[3] conducted comparative genomic hybridization studies on seven astroblastomas and identified specific chromosomal alterations that were not typical of ependymoma or of infiltrating astrocytic neoplasms. These findings suggest that astroblastomas have a characteristic cytogenetic profile that may be useful for differential diagnosis[3].

Radiological features can also help to distinguish astroblastomas from other types of tumor that resemble them. Port et al[9] reported a retrospective study of radiological findings in patients with astroblastoma, from which they defined some characteristic features : (1) astroblastomas are solid and cystic masses, with the solid component being of bubbly appearance, (2) the lesions have little peritumoral T2 hyperintensity for their large size, suggesting the lack of local tumor infiltration into the surrounding brain tissue, (3) the tumors have a relatively low signal intensity on T2-weighted images, and (4) the tumors show intense heterogenous enhancement[9]. In our patient, however, the tumor showed as a solid component with a marked peritumoral edema. On T2-weighted MRI, it had relatively high signal intensity. Despite the marked peritumoral edema seen on MRI in our patient, there was no evidence of tumor infiltration and invasion into the adjacent brain tissue at the time of the operation. Port et al[9] studied only six patients with astroblastoma and larger cohorts of patients must be analyzed in order to define the radiological characteristics of astroblastomas.

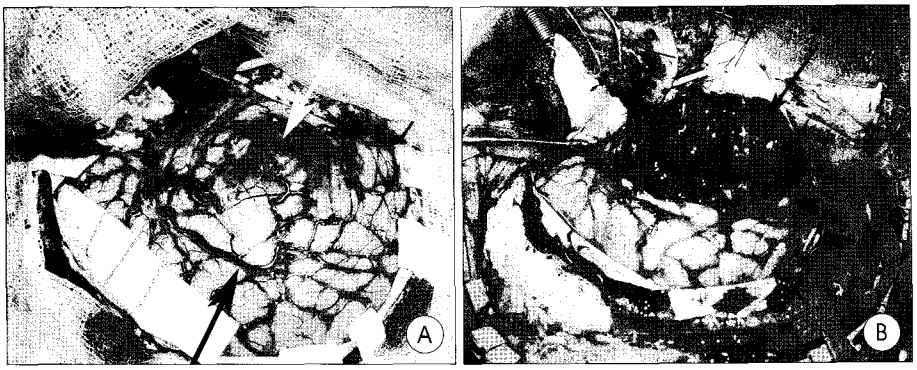

Fig. 3. Intraoperative findings. A : Photograph taken after opening the dura showing the pinkish surface of the tumor (white arrow) located in the temporal lobe and a superficial sylvian vein (black arrow) looking like an arterialized vein in arteriovenous malformation. B : Photograph taken after circumferential dissection of the tumor showing a mass (arrow) protruding from the brain parenchyma.

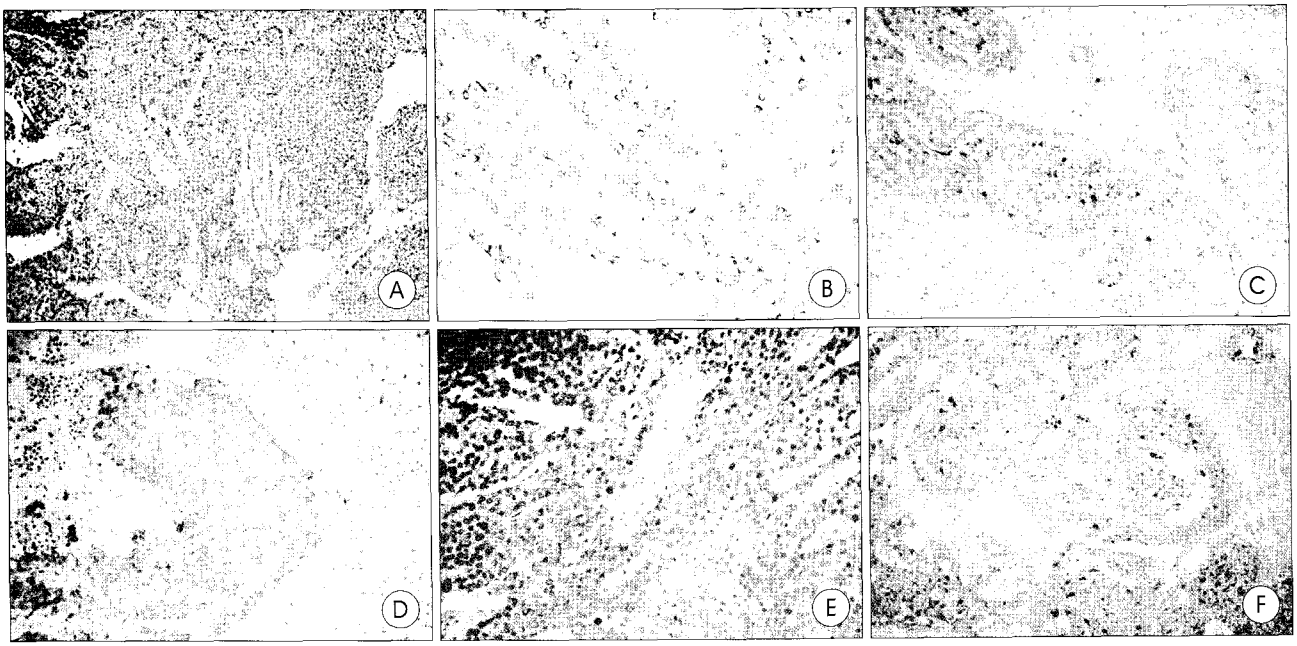

Fig. 4. A : Photomicrograph showing perivascular arrangement of tumor cells forming prominent pseudorosettes, and perivascular hyalinization (H&E, ×200). B~F : Photomicrographs of immunostained tissue samples showing different degrees of immunoreactivity with antibodies. The tumor cells were positive for GFAP (B), vimentin (C), NES (D), cytokeratin7 (E), and S-100 protein (F).

The tumor in our patient was fed via the left middle cerebral artery, not by extracranial arteries (Fig. 2). The tumor vessels become opaque in the early arterial phase. Angiography revealed a uniform distri-bution of fine neoplastic vessels and early filling of the veins. During the operation, we identified early filling veins that resembled arterialized veins in a patient with arteriovenous malformation (Fig. 3). To our knowledge, there have been few reports on angiography of astroblastomas. Because an astroblastoma is a type of glial tumor and shows intense enhancement on MRI, its angiographic features may be similar to those of other glial tumors, as we saw for the tumor in our patient.

An optimum management strategy for astroblastoma has not been established. Bonnin and Rubinstein[2] reported that the best clinical results were obtained after total or subtotal resection of the tumor, followed by radiotherapy. Brat et al[3] suggested that total resection of these tumors should be a therapeutic goal, since there had been no recurrence of the tumor in their study, at least in the short term. Complete resection was possible in our patient because of the clear border between the tumor and adjacent brain tissue. We performed a circumferential dissection around the margin of the lesion, as for arteriovenous malformation. During the dissection, the mass was progressively lifted out of the brain parenchyma (Fig. 3). There was no infiltration and invasion into the brain.

The postoperative course and prognosis of astroblastomas remain unpredictable. Histologically, these tumors are subclassified as low-grade and high-grade types[2,3,10]. However, there is no correlation between the histopathological subtypes and the length of postoperative survival[2,3]. Bonnin and Rubinstein[2] suggested that the prognosis could be complicated by the potential of the astroblastoma to convert into a more malignant type of glioma.

Conclusion

Astroblastoma is a distinct type of glial tumor, usually well circumscribed and supratentorial in young adults, with the characteristic histopathological pattern of perivascular arrangements of tumor cells. In this report, we have described and discussed our clinical, radiological, and pathological findings in a patient with astroblastoma, and have considered previous reports in relation to our findings.

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