Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ

Cukurova Med J 2024;49(4):1107-1110
DOI: 10.17826/cumj.1431427

LETTER TO THE EDITOR

Pilocytic astrocytoma with diffuse angiocentric arrangement and calcification: a case report

Yaygın anjiyosentrik düzenlenme ve kalsifikasyon gösteren pilositik astrositom: olgu sunumu

İsmail Saygın[1], Emel Çakır[2], Sevdegül Aydın Mungan[1], Uğur Yazar[1]

[1]Karadeniz Technical University, Trabzon, Turkey
[2]University of Health Sciences, Istanbul, Turkey

To the Editor,

Pilocytic astrocytoma (PA) is a slow-growing, well-circumscribed grade 1 tumor that occurs in children and young adults. It is the most common glial tumor in the central nervous system of children. Although one third of patients are older than 18 years, only 17% are older than 30 years[1]. In children, 67% of PAs are localized in the cerebellum, while in adults 55% of PAs are supratentorial[2]. The extent of resection is the most important prognostic factor. Adult PAs are less common than pediatric PAs and are associated with a poorer prognosis. The most common alterations are fusions of BRAF[3].

PAs can usually be detected by computed tomography (CT). They are usually well-circumscribed lesions with a round or oval shape and solid, cyst-like features[4]. Macroscopically, PAs are cystic and contain 50% of the mural nodule. Only about 10 % of PAs are completely solid. Microscopically, PAs show a biphasic pattern with alternating dense fibrils and loose microcystic areas. In most cases, only one of these two patterns can be seen. Occasionally, PAs show focal perivascular pseudorosette structures similar to those seen in pilomyxoid astrocytomas[5]. When pseudorosette structures are widespread, the differential diagnosis of other tumors with pseudorosette structures is difficult. Only a small proportion of PAs show thin calcification supporting chronicity[2].

Our case shows extensive calcification and pseudorosette structures. Solid areas are focally present. The differential diagnosis includes tumors with pseudorosette structures or angiocentric arrangement.

A 38-year-old male patient presented to the clinic with a headache. Magnetic resonance imaging (MRI) showed a mass with cystic and solid components in the left parietal lobe. There was no edema or mass effect around the lesion. Extensive calcifications were also observed on CT scan. On microscopic examination after complete resection of the tumor, the neoplastic cells showed a diffuse angiocentric arrangement rather than the classic biphasic pattern of PA (Figure 1a). Diffuse coarse calcifications were also seen in some areas (Figure 1b). Pseudorosette structures, which resemble the angiocentric arrangement, consist of oval-round glial cells surrounding central vascular structures. These vascular structures are usually thickened and hyalinized. (Figure 1c). A few Rosenthal fibers and eosinophilic granular bodies are observed in the solid part of the tumor, which is very rare (Figure 1d).

PA with diffuse angiocentric arrangement is uncommon in the literature. In a case report published in 2013 by Hai-xia Cheng and Yin Wang, they described the first case of cerebellar PA with extensive angiocentric arrangement in the literature[6]. Jun Xie and Hui Wang presented a case report of

Address for Correspondence: İsmail Saygın, Karadeniz Technical University, Department of Pathology, Trabzon, Turkey E-mail: ismailsaygin@ktu.edu.tr
Received: 05.02.2024 Accepted: 10.04.2024

supratentorial PA with extensive angiocentric arrangement in 2016[7].

Pilomyxoid astrocytoma (PMA) was first described by Tihan et al. in 1999 as a subtype that is clinically and histopathologically distinct from PA[8]. It is generally a solid and well-circumscribed mass that most commonly occurs in the hypothalamic region. In contrast to PAs, PMAs typically lack the biphasic pattern and do not contain Rosenthal fibers. PMAs very rarely contain eosinophilic granular bodies. Several mitoses, focal necrosis or vascular proliferation may be seen in PMAs. Radiologically, they are solid, contain little or no calcification and may contain a cystic component. PMAs are classified as grade 1. PMAs have similar genetic alterations to pilocytic astrocytomas. The clinical difference between PMAs and PAs is that PMAs recur more frequently and have a worse outcome[8].

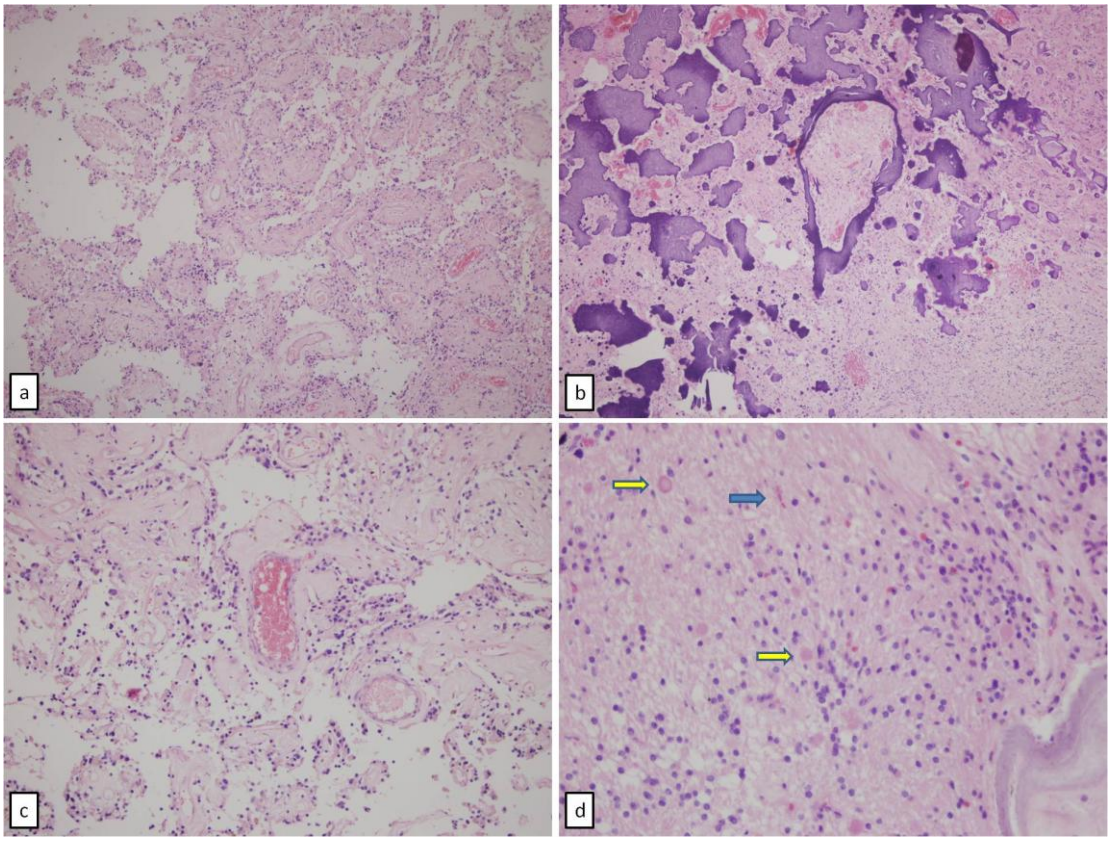

Figure 1.

a. HE X 200: Diffuse angiocentric arrangement of glial cells

b. HE X 200: Widespread calcification in some areas

c. HE X 400: Thickened and hyalinizated vascular structures

d. HE X 400: A few Rosenthal fiber (blue arrow) and eosinophilic granular bodies (yellow arrow)

Angiocentric gliomas (AGs) are pediatric grade 1 diffuse glial tumors. Although AG was described in 2005, it was included as a separate entity in the WHO classification of tumors of the central nervous system in 2007. Almost all AGs show a MYB-QKI fusion. AG consists of uniform, bipolar cells with cigar-shaped nuclei and glial cytoplasmic extensions. In some AGs, the cells may show a pseudorosette-like

radial arrangement around large vessels. In AGs, an eosinophilic structure may be present in the cytoplasm of some cells. Immunohistochemically, most AGs are negative for OLIG2 and show dot-like and ring-like staining with EMA[9].

Papillary glioneuronal tumors (PGNTs) are described radiologically as solid, cystic, well-defined masses. They may also show calcifications. PGNT is a well-circumscribed grade 1 glial tumor. Microscopically, PGNTs consist of glial cells with round nuclei and narrow cytoplasm arranged around hyalinized vascular structures. In addition, neuronal cells are found between these papillary structures. Immunohistochemical markers are helpful for differentiation. In PGNTs, the neuronal cells between the papillary structures react positively to synaptophysin and NeuN. PRKCA gene fusion is a hallmark of PGNTs[9].

Astroblastoma is a rare glial tumor that usually affects children and young adults. Radiologically, it appears as a well-circumscribed, calcified, lobular, solid mass. The most characteristic feature is the Multicystic "bubbly" appearance[10]. No WHO grade has been defined for astroblastoma or anaplastic astroblastoma. Microscopically, perivascular pseudorosette structures and vascular hyalinization are frequently observed. In astroblastoma, the cells of the pseudorosette structures are formed by the luminal orientation of the broad-based cell processes with an oval-round, uniform nucleus and columnar eosinophilic cytoplasm. In general, there is no fibrillar stroma in astroblastomas. Astroblastomas show an MN-1 alteration[9].

Cortical ependymoma is a WHO grade 2 tumor that arises from ependymal cells. It usually occurs in children and young adults. It is often localized in the ventricles and spinal cord and sometimes in the cerebral hemispheres. Supratentorial ependymomas usually occur in older children and adults. Cystic changes and calcifications can be observed radiologically. Microscopically, rosette and pseudorosette structures are observed. Pseudorosette structures consist of ependymal cells arranged around the vessels and fibrillar extensions of these cells extending outwards from the vessel. The absence of Rosenthal fibrils and eosinophilic globules as well as a ring-like positivity for EMA in the rosette structures help to distinguish them from PAs. Ependimomas may show molecular alterations such as the fusion of YAP1 and RELA[9].

In the latest classification of tumors of the central nervous system (2021), the WHO has added further molecular examinations to the morphological parameters for diagnosis[9].

In conclusion, pathologists should be cautious with small tumor specimens containing only papillary structures. PA should be kept in mind in the differential diagnosis of tumors with angiocentric arrangement. In case of clinical and especially radiologic inconsistencies, specimens should be examined very carefully and additional findings should be considered for differential diagnosis.

Author Contributions: Concept/Design : İS, EC; Data acquisition: İS; Data analysis and interpretation: İS, EC; Drafting manuscript: İS. EC; Critical revision of manuscript: SM, UY; Final approval and accountability: İS, EC, SM, UY; Technical or material support: SM; Supervision: SM, UY; Securing funding (if available): n/a.
Ethical Approval: * An informed consent has been obtained from the patient.
Peer-review: Externally peer-reviewed.
Conflict of Interest: Authors declared no conflict of interest.
Financial Disclosure: Authors declared no financial support

REFERENCES

1. Chourmouzi D, Papadopoulou E, Konstantinidis M, Syrris V, Kouskouras K, Haritanti A et al. Manifestations of pilocytic astrocytoma: a pictorial review. Insights Imaging. 2014;5:387-402.
2. Manika M, Rajesh S, Poonam S, Anchal G. Densely calcified pilocytic astrocytoma in the sellar/suprasellar region. Int J Clin Pediatr. 2012;1:129-132.
3. Gregory TA, Chumbley LB, Henson JW, Theeler BJ. Adult pilocytic astrocytoma in the molecular era: a comprehensive review. CNS Oncol. 2021;10:CNS68.
4. Docampo J, González N, Muñoz A, Bruno C, Morales C. Pilocytic astrocytoma. forms of presentation. Neuroradiology. 2014;78:68-81.
5. Love S, Louis DN, Ellison DW. Greenfield's Neuropathology, 8th ed. New York, Routledge, 2008.
6. Chemg HX, Wang Y. Pilocytic astrocytoma with angiocentric arrangement: a case report and literature review. Chinese Journal of Contemporary Neurology and Neurosurgery. 2013;13:342-8.
7. Xie J, Wang H. Pilocytic astrocytoma with angiocentric arrangement:pathological features and differential diagnostic. Int J Clin Exp Pathol. 2016;9:2466-71.
8. Tihan T, Fisher PG, Kepner JL, Godfraind C, McComb RD, Goldthwaite PT et al. Pediatric Astrocytomas with monomorphous pilomyxoid features and a less favorable outcome. J Neuropathol Exp Neurol. 1999;58:1061-8.
9. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D et al. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro Oncol. 2021;23:1231-51.

10. Port JD, Brat DJ, Burger PC, Pomper MG. Astroblastoma: radiologic-pathologic correlation anddistinction from ependymoma. AJNR Am J Neuroradiol. 2002;23:243-247.