Radiology Case Reports 14 (2019) 700–703

Available online at www.sciencedirect.com

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Case Report

Angiocentric glioma mimicking encephalomalacia

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ARTICLE INFO

Article history:
Received 15 February 2019
Revised 10 March 2019
Accepted 13 March 2019
Available online 28 March 2019

Keywords:
Angiocentric glioma
Encephalomalacia
Seizures
Low-grade glioma

ABSTRACT

Angiocentric glioma is a rare low-grade neoplasm of the central nervous system which typically presents with medication-refractory seizures in children and young adults. On magnetic resonance imaging, angiocentric glioma is classically T1 hypointense and T2/FLAIR hyperintense. We present the case of a 40-year-old male who had been followed by our institution for 17 years for management of epilepsy. Initial and repeat brain imaging showed an apparent region of cystic encephalomalacia in the right frontal lobe. In an attempt to control his seizures, the lesion was resected. Grossly, the cut surface of the specimen was characterized by multiple small cystic spaces. Microscopically, the lesion was composed of an infiltrative population of glial cells variably arranged in perivascular "pseudorosettes," nodules, and subpial "palisades." The final diagnosis was angiocentric glioma. This is the second reported case of an angiocentric glioma mistaken for encephalomalacia.

Introduction

Angiocentric glioma is an uncommon neoplasm of the brain which typically presents with medication-refractory seizures in children and young adults [1,2]. The tumor most commonly is centered in the cerebral cortex of the frontal, temporal, or parietal lobes. On magnetic resonance imaging (MRI), the lesion is typically hypointense on T1-weighted images and hyperintense on T2 and fluid attenuated (FLAIR) sequences, does not enhance with gadolinium, and is slow-growing or stable over serial examinations [1]. Additional reported imaging findings include a "stalk-like" extension toward the lateral ventricle and a "rim-like" T1 hyperintensity of the involved gray matter [1]. Microscopically, angiocentric gliomas are characterized by elongate glial cells arranged in "pseudorosettes" around blood vessels; the neoplastic cells also infiltrate surrounding neural parenchyma, form nodules comprised of spindled tumor cells, and aggregate underneath the pia mater [3]. Treatment of angiocentric gliomas involves complete resection, often leading to resolution of the patient's seizures; consequently, this neoplasm is classified by the World Health Organization (WHO) as a grade I tumor [3].

Encephalomalacia is the end-stage of liquefactive necrosis and refers to loss of brain tissue as a result of multiple factors, including infarction, hemorrhage, or trauma. Using MRI, encephalomalacia is recognized as a loss of brain matter and is typically isointense to cerebrospinal fluid on all sequences: T1

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https://doi.org/10.1016/j.radcr.2019.03.015

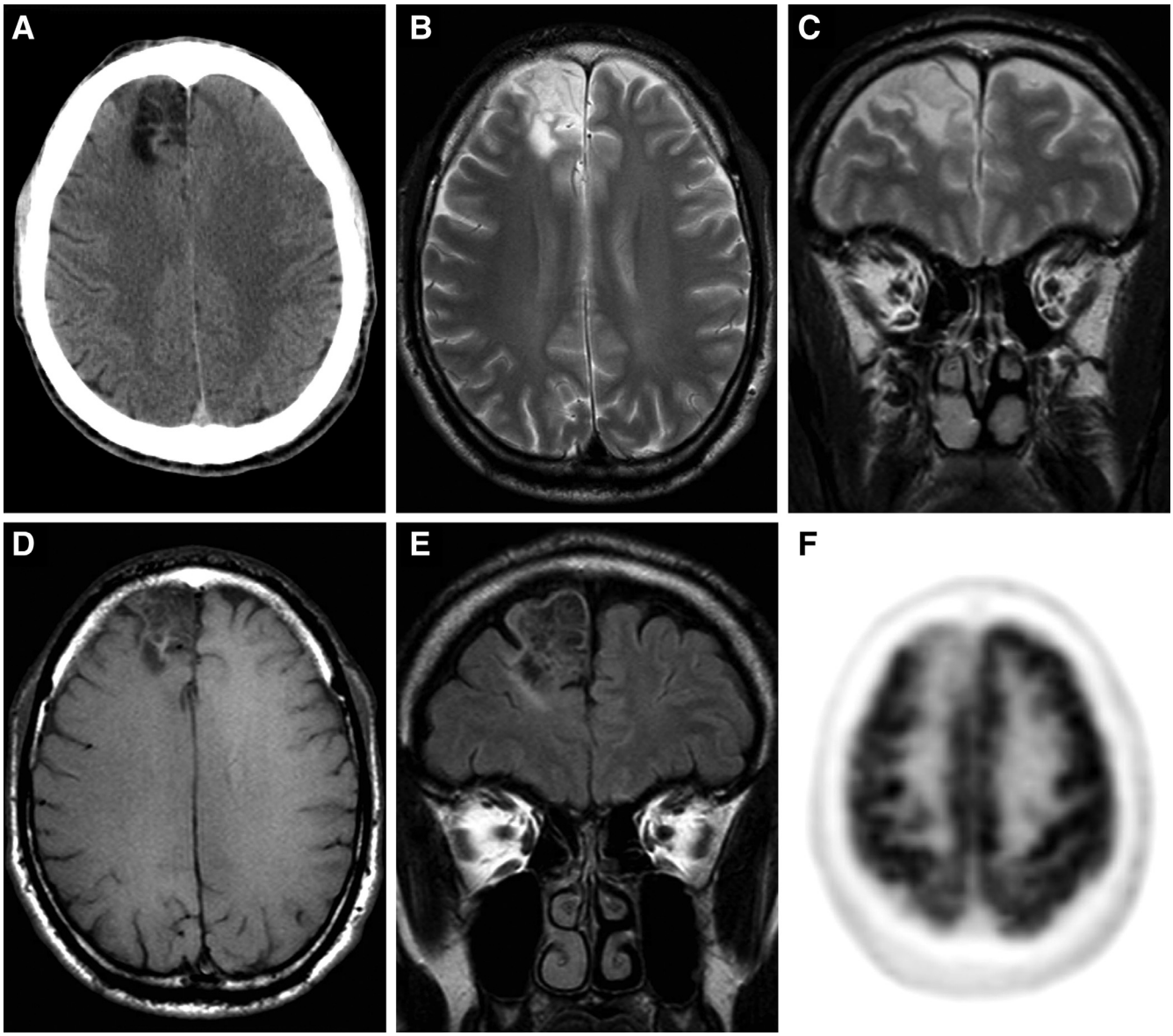

Fig. 1 – Axial noncontrast head CT (A; patient age 25 years) shows a hypoattenuating lesion in the right frontal lobe, involving the cortex and underlying white matter, with no appreciable mass effect, initially interpreted as encephalomalacia and gliosis. Axial (B) and coronal (C) T2-weighted images show that the right frontal lobe lesion has very hyperintense signal and involves the cortex and subcortical white matter. Axial T1-weighted image (D) shows that the lesion is predominantly hypointense but has subtle hyperintense signal at its periphery. The coronal FLAIR (E) image shows that the lesion has predominant hypointense FLAIR signal centrally but has a FLAIR hyperintense rim. (Images B-E, patient age 39 years) Axial PET (F; patient age 39 years) shows that the right frontal lesion is not FDG avid, a finding consistent with encephalomalacia.

hypointense, T2 hyperintense, and FLAIR hypointense. However, encephalomalacia is very often associated with adjacent gliosis which is FLAIR hyperintense.

Case report

The patient was a 40-year-old male with poorly controlled epilepsy despite the use of 4 antiepileptic medications and vagal nerve stimulation. He had been followed at our institution for epilepsy management since the age of 23 years. His history included head trauma as a child, right frontal lobe encephalomalacia, substance abuse, and an episode of cardiac arrest precipitated by cocaine use. A CT scan at patient age 25 (Fig. 1A) showed a hypoattenuating lesion in the right frontal lobe, involving the cortex and underlying white matter, with no appreciable mass effect, interpreted as encephalomalacia. Due to the refractory nature of his seizures, he was evaluated for surgical management. Electrophysiologic

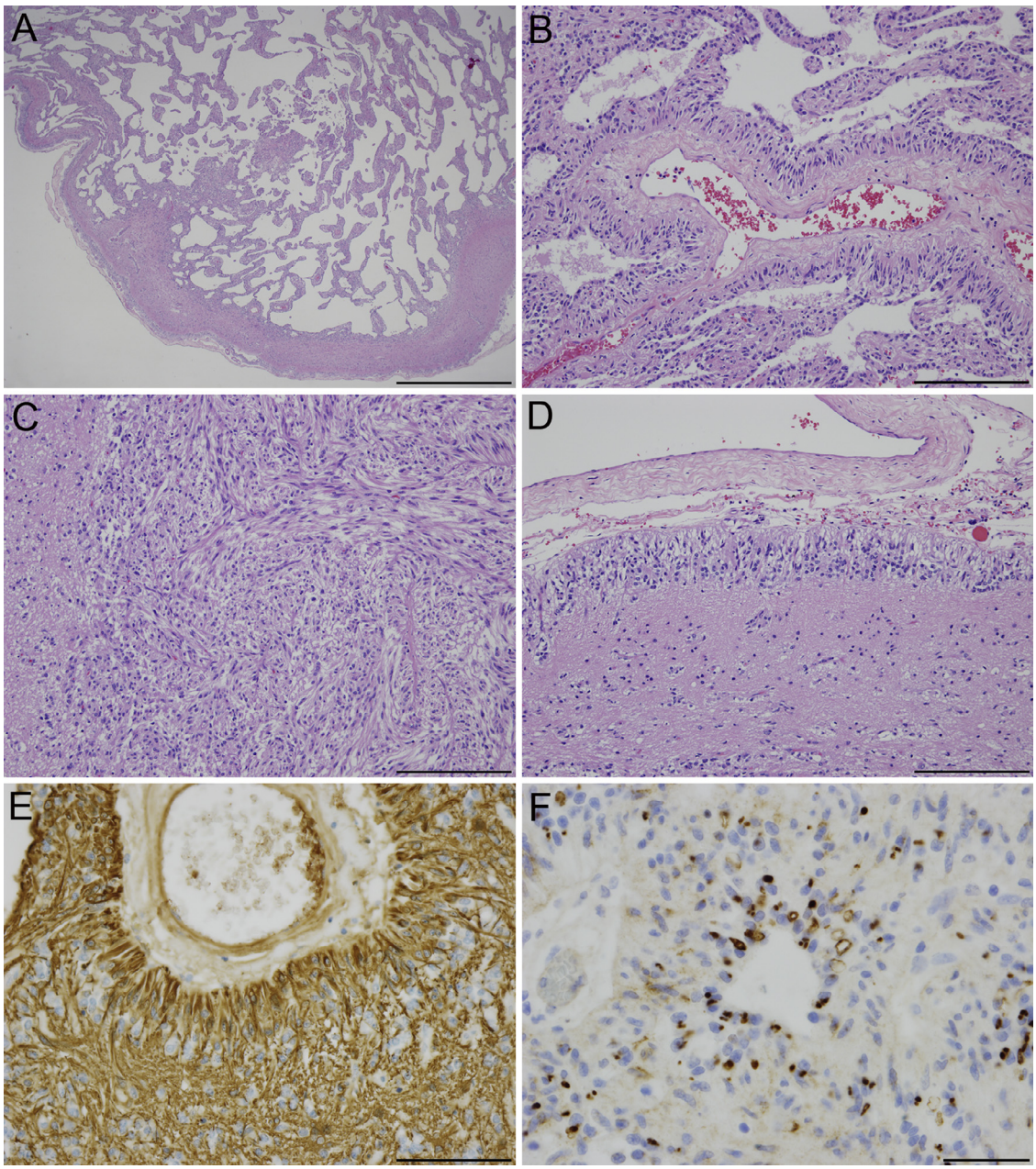

Fig. 2 – The neoplasm is composed of multiple cystic spaces (A, hematoxylin and eosin, original magnification ×20) lined by elongate tumor cells, which are frequently arranged around blood vessels forming perivascular "pseudorosettes" (B, hematoxylin and eosin, original magnification ×200). Occasional nodules composed of spindle cells in fascicles, reminiscent of schwannoma, are present (C, hematoxylin and eosin, original magnification ×200). Tumor cells form palisades along the subpial surface of the brain (D, hematoxylin and eosin, original magnification ×200). The neoplastic cells are immunoreactive for GFAP (E, GFAP, original magnification, ×400), and cytoplasmic dot-like microlumen-type staining is seen on EMA stain (F, EMA, original magnification ×600). (Scale bars: A, 2 mm; B, C, D, 200 microns; E, 100 microns; F, 50 microns.)

studies localized his seizures to the bilateral inferomesial temporal lobes and the right frontal lobe. Brain MRI at 39 years demonstrated a nonenhancing right frontal lobe lesion involving the cortex and subcortical white matter, characterized as predominately T1 hypointense, T2 hyperintense, and predominately FLAIR hypointense (Fig.1B-E), but with a thin rim of FLAIR hyperintense signal. A positron emission tomography (PET)/computed tomography (CT) scan performed as part of a preoperative evaluation demonstrated a paucity of fluorodeoxyglucose (FDG) uptake in the right frontal lobe, corresponding to the previously identified lesion (Fig. 1F). The lesion was interpreted as encephalomalacia.

The right frontal lobe lesion was resected to grossly normal appearing brain parenchyma. Gross examination of the surgical specimen revealed a 2.7 cm in greatest dimension tan-white lesion comprised of small cystic spaces and

covered by a thin layer of superficial cortex. Microscopically, numerous cystic spaces lined by slender cuboidal-to-columnar cells were seen (Fig. 2). The tumor cells showed radial arrangements around blood vessels, reminiscent of ependymal pseudorosettes. Superficially, tumor cells formed radial arrays extending along the subpial surface of the brain, and in the periphery of the tumor spindle-shaped cells formed distinct nodules with infiltration of the adjacent neural tissue by single tumor cells. Immunohistochemical stains showed diffuse GFAP and S100 reactivity in the neoplastic population, with frequent EMA-immunoreactive cytoplasmic "dots." No mitotic figures were seen, and the proliferation rate, measured with an immunohistochemical stain for Ki-67, was low. The morphologic and immunohistochemical findings were diagnostic for angiocentric glioma, a WHO Grade I glioma.

Following the operation for resection, he was seizure-free for 52 days, the longest he had been seizure-free since age 14. In that time, he reported an improvement in his memory and had no residual neurologic deficits. Unfortunately, following an attempt to wean one of his antiepileptic medications, he again presented to our institution following a cluster of seizures.

Discussion

In this case, the lesion mimicked encephalomalacia on all radiologic imaging modalities that were obtained (CT, MRI, and PET). The lesion was hypodense on CT and lacked significant mass effect. On MRI, it was predominantly T1 hypointense, T2 hyperintense, and FLAIR hypointense, characteristics similar to those seen in cavitary encephalomalacia. Furthermore, the lesion lacked FDG avidity on the PET scan.

While the lesion in this case lacked the classic "stalk-like" extension of tumor to the lateral ventricle, the neoplasm did show a subtle T1 hyperintensity of the involved gray matter, a possible clue to the correct radiologic diagnosis. However, T1 hyperintensity may also be seen in the setting of encephalomalacia as an area of laminar necrosis. Grossly and microscopically, the resected lesion was principally composed of numerous small cysts, explaining the predominant hypointense FLAIR signal. Indeed, previously reported partially cystic angiocentric gliomas showed T2 hyperintensity with corresponding hypointense signal on FLAIR [4,5]; the case reported by Kundu and colleagues was also originally interpreted as encephalomalacia. The majority of reported angiocentric gliomas have followed a benign course, and our patient's lesion had been stable for at least 17 years; however, rare cases have been high-grade at presentation or progressed to a higher-grade glioma [6,7].

In summary, we have described a case in which the radiologic findings of angiocentric glioma mimicked encephalomalacia. This is the second report of a largely cystic angiocentric glioma being radiologically misinterpreted as encephalomalacia. Although angiocentric gliomas are rare neoplasms, these lesions should be included in the differential diagnosis for a partially cystic cortical lesion with predominant FLAIR hypointense signal.

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