Case Report

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Necrotizing Primary Angiitis of the Central Nervous System Mimicking Brain Abscess: A Case Report and Literature Review

뇌농양을 모방한 괴사성 원발성 중추신경계 혈관염: 증례 보고와 문헌 고찰

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Primary angiitis of the central nervous system (PACNS) is a rare vasculitis in the central nervous system. Herein, we report a case of diagnosis and treatment of necrotic pattern PACNS, which was difficult to differentiate from a brain abscess. A 19-year-old male presented with blurred vision and a headache. Brain MRI revealed irregular rim-enhancing necrotic masses with central diffusion-high signal intensity in the corpus callosum and peripheral diffusion-high signal intensity in the left parietotemporal periventricular area. Susceptibility-weighted imaging revealed multiple punctate hemorrhages in the lesions. The patient was diagnosed with unusual abscess or tumefactive PACNS. Therefore, we initially treated the patient with antibiotics to rule out brain abscess. However, the brain lesions did not improve on follow-up MRI after the antibiotic treatment. Surgical biopsy was performed, and the histopathological diagnosis was PACNS with a necrotic pattern. The necrotic lesions became smaller on follow-up MRI after high-dose corticosteroid treatment.

Index terms Central Nervous System; Brain Abscess; Vasculitis

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INTRODUCTION

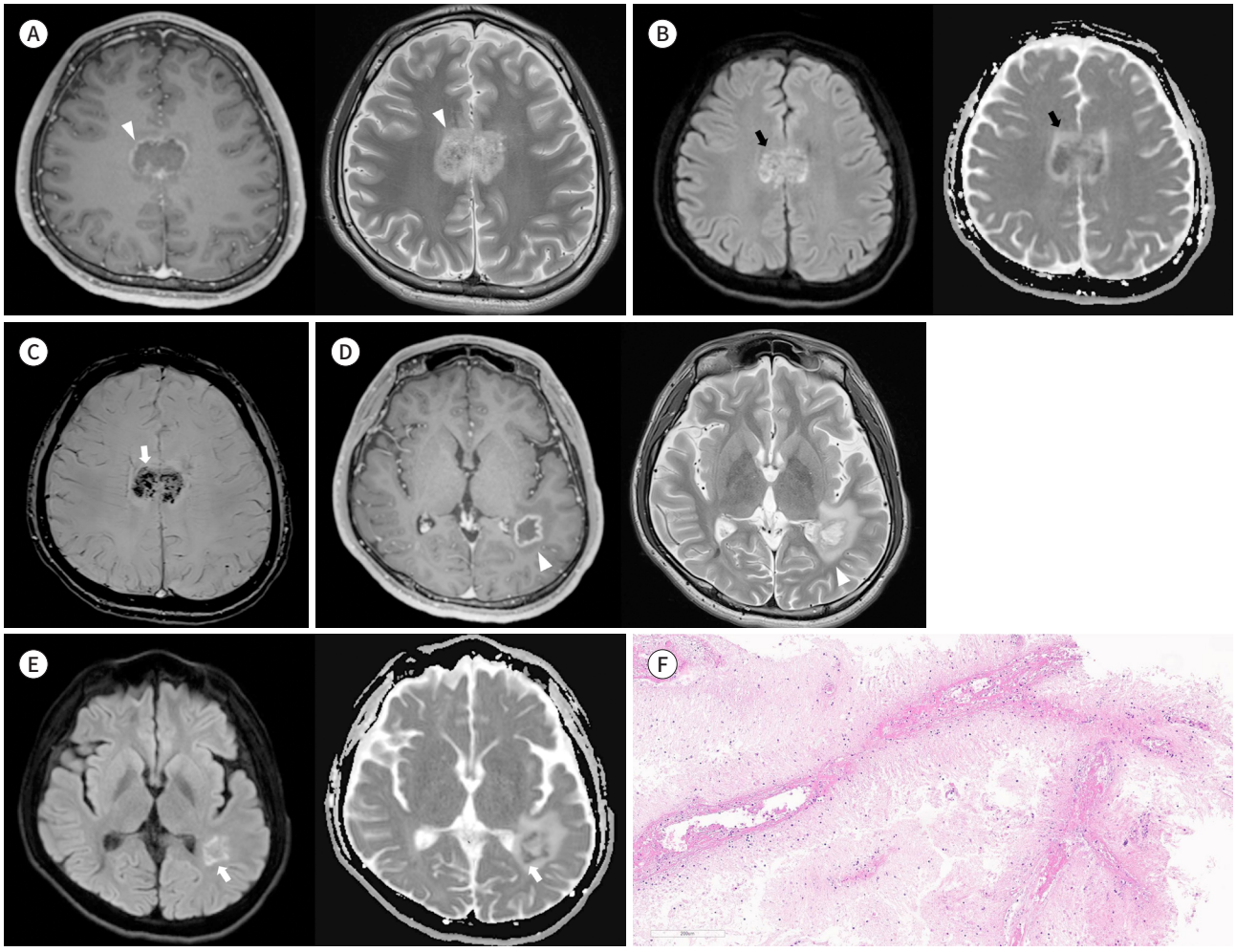
Primary angiitis of the central nervous system (PACNS) is a rare vasculitis of unknown etiology confined to the CNS. Inflammation affects both small and large vessels of the brain, spinal cord, and leptomeninges (1). The clinical features and MRI findings of PACNS are highly variable and non-specific (1, 2). The differential diagnoses of PACNS are broad because diverse neurological diseases with inflammatory, infectious, malignant, and vascular etiologies can resemble PACNS (1). Herein, we report a case of necrotic pattern PACNS, which was difficult to differentiate from brain abscess, diagnosed and treated using a team-based diagnostic approach.

CASE REPORT

A 19-year-old male visited the emergency room with a headache that had occurred 5 hours ago. He also showed symptoms of blurred vision, which had started 2 weeks ago, and mild fever (37.6°C). The patient recognized hand motion but could not count the fingers. Neurological examination revealed no sensory or motor impairments. Peripheral blood laboratory results revealed leukocytosis (white blood cell count, 12710/μL), while cerebrospinal fluid (CSF) examination showed no remarkable findings. Therefore, CT and CT angiography were performed to rule out the possibility of brain lesions, such as brain abscesses or ischemic strokes. Brain CT revealed a low-density mass measuring approximately 3 cm in the body of the corpus callosum. CT angiography revealed no significant steno-occlusion or aneurysm in the intracranial or neck arteries. On the same day, the patient underwent brain MRI for further evaluation. Contrast-enhanced T1-weighted imaging (T1WI) revealed irregular rim-enhancing masses in the corpus callosum and left parietotemporal periventricular area. An approximately 3 cm × 2 cm × 2 cm rim-enhancing corpus callosum mass showed heterogeneous high signal intensity on T2WI (Fig. 1A). Diffusion-weighed image (DWI) and apparent diffusion coefficient (ADC) maps of the corpus callosum lesions showed central diffusion restriction (Fig. 1B). Susceptibility-weighted image (SWI) showed multiple punctate hemorrhages in the corpus callosum (Fig. 1C). The other irregular rim-enhancing lesion (approximately 2 cm × 2 cm × 1 cm) in the left parietotemporal periventricular area on contrast-enhanced T1WI showed heterogeneous high signal intensity with perilesional edema on T2WI (Fig. 1D). DWI and ADC map revealed diffusion restriction at the enhancing rim of the left parietotemporal periventricular lesion (Fig. 1E). SWI also revealed multiple punctate hemorrhages in the left parietotemporal periventricular lesion (data not shown). All the other laboratory findings suggestive of vasculitis were negative. Laboratory test results for infection were negative. Considering the presence of multiple punctate hemorrhages in the necrotic rim-enhancing lesions and the heterogeneity of the DWIs, we interpreted this as an unusual brain abscess or tumefactive vasculitis.

A team-based diagnostic approach was adopted, in which a radiologist, neurologist, neurosurgeon, and pathologist were consulted. After a multidisciplinary discussion, we decided to initially treat the patient with antibiotics to rule out the possibility of brain abscess because the patient had fever and leukocytosis during hospitalization. However, there was no significant change in the rim-enhancing masses on follow-up MRI after 2 weeks of antibiotic treat-

Fig. 1. Necrotizing primary angiitis of the central nervous system. A 19-year-old male with headache.
A. Contrast-enhanced T1WI (left) shows irregular rim-enhancing mass in the corpus callosum (arrowhead). T2WI (right) shows the mass with heterogeneous high signal intensity (arrowhead).
B. Diffusion-weighted image (left) shows central diffusion-high signal intensity at the corpus callosum lesion (arrow). Apparent diffusion coefficient map (right) shows heterogeneous low signal intensity at central portion of the lesion (arrow), suggesting diffusion restriction.
C. Susceptibility-weighted image demonstrates multiple punctate hemorrhages at the corpus callosum mass (arrow).
D. Contrast-enhanced T1WI (left) shows the other irregular rim-enhancing mass in the left parieto-temporal periventricular area (arrowhead). T2WI (right) shows heterogeneous high signal intensity at the mass with perilesional edema (arrowhead).
E. Diffusion-weighted image (left) shows peripheral diffusion-high signal intensity at the left parieto-temporal periventricular lesion (arrow). Apparent diffusion coefficient map (right) shows low signal intensity at the rim of the mass (arrow), suggesting diffusion restriction.
F. Hematoxylin and eosin staining (× 200) of the histopathological specimen shows marked fibrinoid necrosis and neutrophilic infiltration with karyorrhectic debris in the vascular wall of small-sized vessels.
WI = weighted image



ment. Therefore, it was determined that a brain abscess was unlikely and the diagnosis of tumefactive PACNS was possible. A stereotactic biopsy of the corpus callosum lesion was performed. The frozen biopsy diagnosis was a brain abscess, but the final histopathological diagnosis was PACNS with a necrotic pattern. Histopathological specimens showed marked fibrinoid necrosis and neutrophilic infiltration with karyorrhectic debris in the walls of the small vessels (Fig. 1F). After 2 months of treatment with high-dose corticosteroids, the sizes of the corpus callosum and left parietotemporal periventricular lesions decreased on follow-up

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MRI (Supplementary Figs. 1, 2 in the online-only Data Supplement). Headache and blurred vision improved, and fever and leukocytosis disappeared after treatment.

This study was approved by the Institutional Review Board (IRB No. 2022-12-006), and the requirement for informed consent was waived.

DISCUSSION

Here, we report the necrotic pattern of tumefactive PACNS mimicking a brain abscess. Necrotizing PACNS occurs rarely, and its radiographic features are diverse and heterogeneous, making its diagnosis difficult. To the best of our knowledge, there have been no case reports of brain abscesses mimicking PACNS.

Harbitz (3) first described PACNS in 1922 as an unknown form of angiitis in the CNS. PACNS refers to isolated vasculitis of the CNS and the infiltration of inflammatory cells into the vascular walls of the brain parenchyma, spinal cord, and leptomeninges. When immune cells infiltrate and destroy the walls of the CNS blood vessels, the vessel walls become thick with stenosis, ultimately resulting in poor blood circulation. However, the weakening of the vessel wall due to inflammation can lead to vessel rupture and intracranial hemorrhage. Both the small and large CNS vessels can be affected by PACNS (1). Pathologically, there are several patterns of PACNS, the most common of which is granulomatous vasculitis, while other types of non-granulomatous vasculitis include lymphocytic and necrotizing vasculitis. Necrotizing vasculitis is characterized by changes in the acute necrotizing vessel walls, transmural fibrinoid necrosis, and acute inflammation. Fibrinoid necrosis and inflammatory reactions can cause vessel wall thickening, resulting in vascular rupture and aneurysmal dilatation (4).

The annual incidence of PACNS is 2.4 cases per 1 million person-years (5). PACNS can affect younger patients with stroke without cerebrovascular risk factors. Signs of PACNS can occur as early as the late 40s. The clinical features of PACNS are highly variable and non-specific: headache, altered cognition, focal neurological deficits, seizures, and encephalopathy. It has an insidious onset and slow clinical course (1, 5). The primary treatments are high-dose corticosteroids and/or immunosuppressants.

Diagnosing PACNS is difficult because of the rarity and heterogeneity of the disease. The variable clinical presentation and non-specific radiological findings make the diagnosis of PACNS challenging. There are two subtypes of PACNS distinguished by the size of the affected vessels: small-and large/medium-vessel disease (1). PACNS with isolated small vessel involvement is more difficult to diagnose than PACNS with isolated large/medium vessel involvement because there are no usual angiographic features of vasculitis.

The diagnosis of PACNS considers comprehensive laboratory and CSF values, brain biopsy, and clinical and imaging findings. Laboratory findings of PACNS are non-specific. In laboratory tests, serum findings are usually normal, but inflammatory markers can be elevated (particularly the erythrocyte sedimentation rate) (1). A CSF study is abnormal in 80%–90% of patients and shows inflammatory findings, typically lymphocytic pleocytosis combined with elevated protein levels (1). The CT findings of PACNS include low-attenuation lesions, suggesting ischemia. Most MRIs reveal multiple bilateral asymmetrical supratentorial and infratentorial infarctions, mainly in the subcortical and deep white matter. Other MRI findings

include parenchymal and leptomeningeal enhancements, masses, intracranial hemorrhages, and areas of increased signal intensity on T2WI or FLAIR images. Cerebral angiography may reveal dilation or occlusion of the intracranial cerebral arteries; however, negative angiographic findings cannot exclude the possibility of PACNS.

The differential diagnosis of PACNS is broad as it is a diverse neurological disease with inflammatory, infectious, malignant, and vascular etiologies that resemble PACNS (6). In this case, a clinical and radiographic differential diagnosis of brain abscess and tumefactive PACNS was required. Initially, we decided to treat the patient with antibiotics to rule out the possibility of brain abscess because the patient presented with fever and leukocytosis during hospitalization, which is uncommon in PACNS (7). The MRIs were retrospectively reviewed, and the presence of prominent microbleeds in the necrotic rim-enhancing lesions and the heterogeneity of the DWIs suggested the possibility of tumefactive PACNS rather than brain abscess. As in our case, the clinical symptoms and imaging findings may not be correlated. Therefore, a team-based approach is required to diagnose and treat tumefactive PACNS. The team-based approach is a step-by-step diagnostic method that involves multidisciplinary discussions with radiologists, neurologists, neurosurgeons, and pathologists. This team-based approach allowed us to make rational decisions.

Careful consideration of possible differential diagnoses and team-based approaches are necessary to avoid delayed diagnosis and unnecessary treatment of tumefactive PACNS.

Supplementary Materials

The online-only Data Supplement is available with this article at http://doi.org/10.3348/jksr.2023.0005.

Author Contributions

Conceptualization, K.E.; investigation, P.C., C.E., K.E.; supervision, K.E.; writing—original draft, all authors; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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뇌농양을 모방한 괴사성 원발성 중추신경계 혈관염: 증례 보고와 문헌 고찰

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원발성 중추신경계 혈관염은 중추 신경계에서 발생하는 드문 혈관염이다. 뇌농양과 구별하기 힘든, 괴사 패턴의 원발성 중추신경계 혈관염을 진단, 치료를 하였던 본원의 증례를 보고하고자 한다. 시야 흐림, 두통을 주소로 19세 남자 환자가 내원하였다. 조영증강영상에서 가장자리가 조영이 잘되는 괴사성 종괴들의 소견이 뇌량과 좌측 두정-측두 뇌실주위 영역에서 보였는데, 확산강조영상에서 뇌량에 있는 종괴는 중심부에 높은 신호강도를 보였고, 좌측 두정-측두 뇌실주위영역에 있는 종괴는 주변부에서 높은 신호강도를 보였다. 자화율강조영상에서 종괴들 내에 다발성 점상 출혈 소견을 보였다. 비정형적인 뇌농양과 종양성 원발성 중추신경계 혈관염의 가능성을 생각하였다. 먼저 뇌농양을 배제하기 위해 항생제 치료를 시작하였다. 하지만 2주간의 항생제 치료 이후에 시행한 뇌 자기공명영상에서 괴사성 종괴들은 호전되지 않은 소견을 보였다. 병변에 대해 수술적 생검을 시행하였고, 최종적으로 괴사 패턴의 원발성 중추신경계 혈관염으로 진단되었다. 고용량 스테로이드 치료 이후 시행한 뇌 자기공명영상에서 괴사성 종괴들은 작아졌다.

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