Primary central nervous system vasculitis (PCNSV) is rare, with a reported annual incidence of 2.4 cases per million [1]. Although the cause of this disorder is unknown, the pathogenesis is related to the inflammation of parenchymal and leptomeningeal arteries and veins [2]. The clinical manifestations are variable, and the disease usually is aggressive or fatal [2,3]. Making the diagnosis from radiologic findings may be difficult because of its variable radiologic expressions. Early corticosteroid therapy often is effective. Herein we report the case of a 56-year-old man who had a well-enhanced cystic mass with severe edema in the right frontal lobe, which was initially felt to be a malignancy. Histologic examination of tissue removed at craniotomy revealed that it was a PCNSV. Despite early administration of corticosteroids, a new lesion developed within 3 days. The lesions responded to treatment with cyclophosphamide and corticosteroid.

Key Words  Vasculitis; Cysts; Neoplasms; Glucocorticoids.
Fig. 1. Brain CT and MRI revealing a cystic mass-like lesion in the right frontal lobe. A: Brain CT reveals a 6-cm low-density lesion. B: Brain MRI illustrates the lesion with low signal intensity on T1-weighted image (WI). C: Fluid-fluid level of the lesion with upper high and lower low signal intensity is illustrated on T2-WI. D: Well-develop capsule is illustrated on enhanced MRI. E: Diffusion WI reveals no restriction.

Fig. 2. Brain CT on 3rd day after admission illustrating increased peritumoral edema and subfalcial herniation.

cystic fluid with degenerated material, was performed.

Postoperatively, the patient regained consciousness but was drowsy. An immediate postoperative MRI revealed no resid-
Aggressive CNS Vasculitis Mimicking Tumor

**Fig. 3.** Immediate postoperative MRI demonstrates no residual tumor at the primary site (A), but there was a new lesion with well enhancement surrounding the occipital horn and trigone of the left lateral ventricle (arrow) (B and C).

**Fig. 4.** Histological findings of vasculitis in the brain lesion. The hematoxylin-eosin stain demonstrated pseudocapsule (A, original magnification ×20) with prominent lymphocyte infiltration into small vessel wall (arrow) (B, original magnification ×200). Immunohistochemistry stains revealed positive for CD3 (C) and CD31 (D), and these markers are related to lymphocytes (original magnification ×200).

The most remarkable abnormality in PCNSV might be marked elevation of the glutamate and glutamine peaks, which is a kind of neurotransmitter primarily found in astrocytes. In inflammatory conditions, cell breakdown of neural and glial elements occurs, along with an associated astrocytic response, which leads to a local accumulation of many metabolites, including a high concentration of glutamine and glutamate [3,7]. Our patient had the typical findings of a high-grade malignant cystic brain tumor on conventional MRI; if his condition had permitted MR spectroscopy, that
procedure may have made the diagnosis of vasculitis more likely.

Histologically, CNS vasculitis can be categorized into 3 types: granulomatous, lymphocytic, and necrotizing [8]. Granulomatous vasculitis is the most common type and has vasculocentric mononuclear and granulomatous inflammation. Lymphocytic vasculitis, the second most common type, has prominent lymphocytic inflammation associated with plasma cell infiltration and destruction of vessels. Necrotizing vasculitis, the least common type, is associated with transmural fibrinoid necrosis, which occasionally causes intracranial hemorrhage [9]. The present case had a cystic mass-like lesion, and the cyst capsule had prominent lymphocytic inflammation into vessel walls and destruction of vessels.

Regimens for treating cerebral vasculitis, such as with corticosteroid and immunosuppressant, are derived from strategies used for other types of vasculitis. Brain edema responds especially well to corticosteroids. Early corticosteroid treatment of PCNSV can often have favorable outcomes and prevent serious outcomes [10]. To our knowledge, despite the early administration of high-dose corticosteroids, rapid progression of the disease within a few days is very extremely rare. The reason our patient’s disease progressed is not known. Nevertheless, the case suggests that the presence of a large mass-like brain lesion with severe edema should prompt efforts to make a tissue diagnosis promptly, so corticosteroid/immunosuppressive therapy can be instituted if indicated.

In conclusion, an unusual case of cystic vasculitis with rapid progression despite early administration of corticosteroid therapy is presented. If malignant cystic tumor combined with severe brain edema is suspected, early biopsy or resection should be considered to confirm the diagnosis and permit early institution of appropriated treatments.

Conflicts of Interest
The authors have no potential conflicts of interest.

Acknowledgments
We also appreciate Wade Martin of Emareye for his critical English revision. This work was supported by the National Research Foundation of Korea (NRF) grant funded by the Korea government (Ministry of Science and ICT) (NRF-2018R1C1B5085134).

REFERENCES
1. Salvarani C, Brown RD Jr, Calamia KT, et al. Primary central nervous system vasculitis: analysis of 101 patients. Ann Neurol 2007;62:442-51.
2. MacLaren K, Gillespie J, Shrestha S, Neary D, Ballardie FW. Primary angiitis of the central nervous system: emerging variants. QJM 2005;98:643-54.
3. Panchal NJ, Niku S, Imbesi SG. Lymphocytic vasculitis mimicking aggressive multifocal cerebral neoplasm: MR imaging and MR spectroscopic appearance. AJNR Am J Neuroradiol 2005;26:642-5.
4. Imbesi SG. Diffuse cerebral vasculitis with normal results on brain MR imaging. AJR Am J Roentgenol 1999;173:1494-6.
5. Negishi C, Sze G. Vasculitis presenting as primary leptomeningeal enhancement with minimal parenchymal findings. AJNR Am J Neuroradiol 1993;14:26-8.
6. Shoemaker EI, Lin ZS, Rae-Grant AD, Little B. Primary angiitis of the central nervous system: unusual MR appearance. AJNR Am J Neuroradiol 1994;15:331-4.
7. Danielsen ER, Ross B. The clinical significance of metabolites. In: Magnetic resonance spectroscopy diagnosis of neurological diseases. New York: Marcel Dekker; 1999. p. 23-43.
8. Salvarani C, Brown RD Jr, Hunder GG. Adult primary central nervous system vasculitis. Lancet 2012;380:767-77.
9. Lee JS, Jung TY, Lee KH, Kim SK. Primary central nervous system vasculitis mimicking a cortical brain tumor: a case report. Brain Tumor Res Treat 2017;5:30-3.
10. Honda M, Koga M, Kanda T. [Treatment for central nervous system vasculitis]. Brain Nerve 2015;67:287-93.