The Clinical and Radiologic Findings of Primary Angiitis of the Central Nervous System

Primer Santral Sinir Sistemi Anjiiti-Klinikoradyolojik Bulgular

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Dear Editor,

A 52-year-old male patient, who had had ischemic strokes twice before, was admitted with symptoms of vision loss, headaches, and forgetfulness. A neurologic examination revealed time disorientation, attention and memory impairment, bilateral vision loss, left 3/5 hemiparesis, and babinski positivity. Magnetic resonance imaging (MRI) showed hyperintense signaling changes in fluid-attenuated inversion recovery-weighted images in the right occipital lobe suggestive of chronic infarction and in the left occipital lobe suggestive of subacute infarction (Figure 1). In addition, cerebral atrophy, widespread leukoaraiosis, and lacunar infarctions were observed in both hemispheres (Figure 2). Diffusion-weighted MRI showed a diffusion restriction in the left occipital lobe suggestive of subacute infarction (Figure 3A, 3B). Cerebrospinal fluid (CSF) protein was increased (201 mg/dL, normal: 15-45 mg/dL). Vascular irregularities, constrictions, poststenotic dilatations, and segmental occlusions were observed in cerebral angiography (Figure 4A, 4B, 4C and 4D). No finding of systemic vasculitis was detected and the patient was diagnosed as having primary angiitis of the central nervous system (CNS) (PACNS) and was administered intravenous methylprednisolone for ten days at a dosage of 1000 mg/day, then oral prednisone at a dosage of 1 mg/kg/day, and monthly cyclophosphamide infusion. No clinicoradiologic findings related to the new vascular event developed in the follow-up.

PACNS is a rare disease characterized by inflammation of the veins of the brain parenchyma, spinal cord, and leptomeninges (1). Immune infiltration in blood vessels of the CNS leads
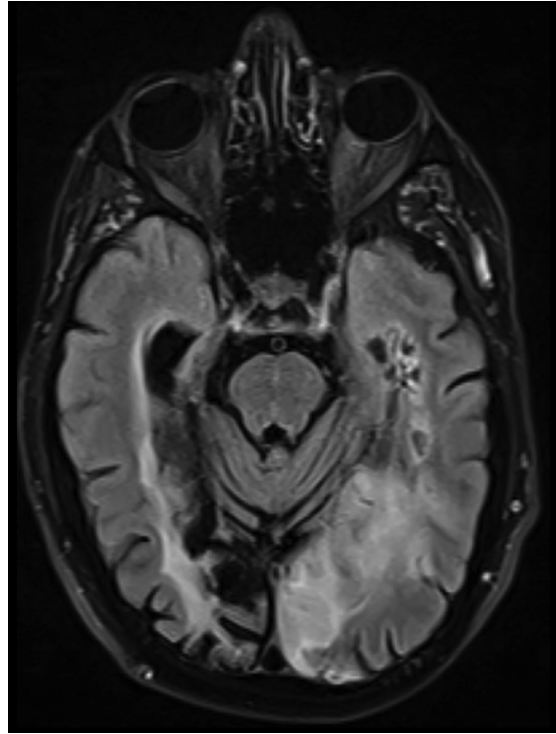
Figure 1. Chronic infarction in the right occipital lobe and subacute infarction in left occipital lobe in axial fluid-attenuated inversion recovery-weighted magnetic resonance imaging

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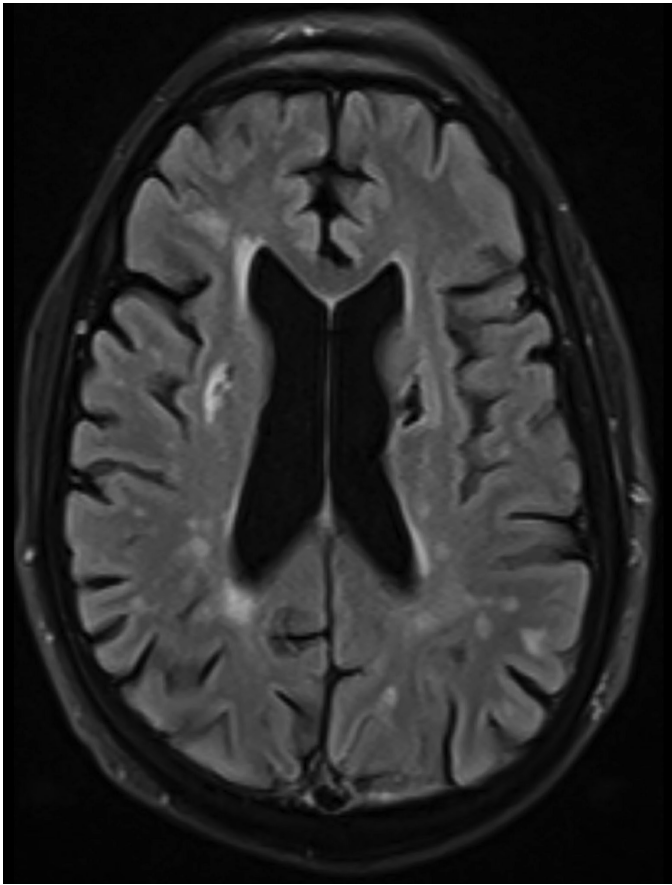
Figure 2. Cerebral atrophy, widespread leukoaraiosis and lacunar infarctions in axial fluid-attenuated inversion recovery-weighted magnetic resonance imaging

to destruction, thickening, and stenosis of vessels, resulting in disturbance in circulation (2). Rupture and bleeding may develop as a result of weakening of blood vessels (2). The clinical characteristics of PACNS are variable and nonspecific; headaches, cognitive disorders, focal neurologic deficits, and epileptic seizures may develop. There are no signs of systemic vasculitis (3). PACNS is diagnosed by clinical presentation, angiography, CSF analysis, and brain biopsy. The diagnostic criteria are as follows: clinical signs of unexplained neurologic deficits; classic angiographic or histopathologic features that indicate CNS angiitis; and no evidence of systemic vasculitis (1). In 2009, the criteria were updated and it was stated that if vasculitic findings are found in tissue biopsy, "definitive" PACNS is diagnosed, and if there are diagnostic MRI and CSF analysis findings along with angiogram
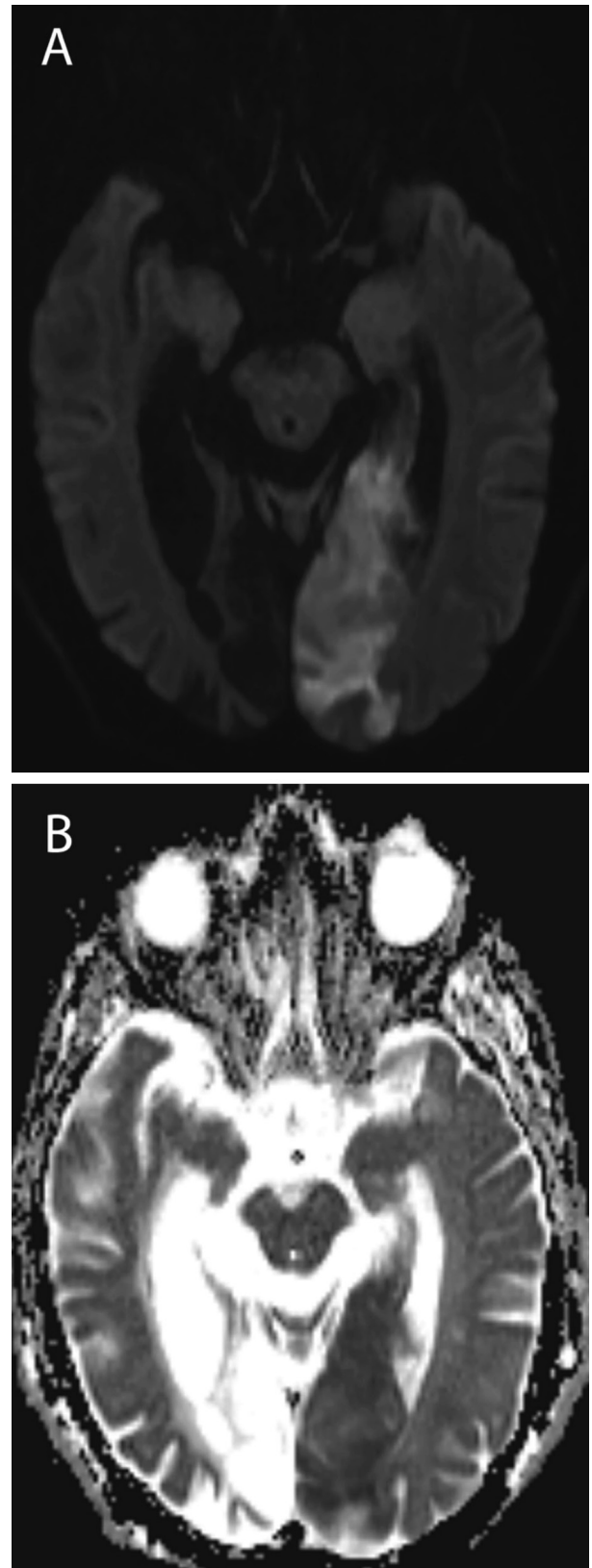
Figure 3. A, B) Diffusion-weighted magnetic resonance imaging showing diffusion restriction in the left occipital lobe suggestive of subacute infarction
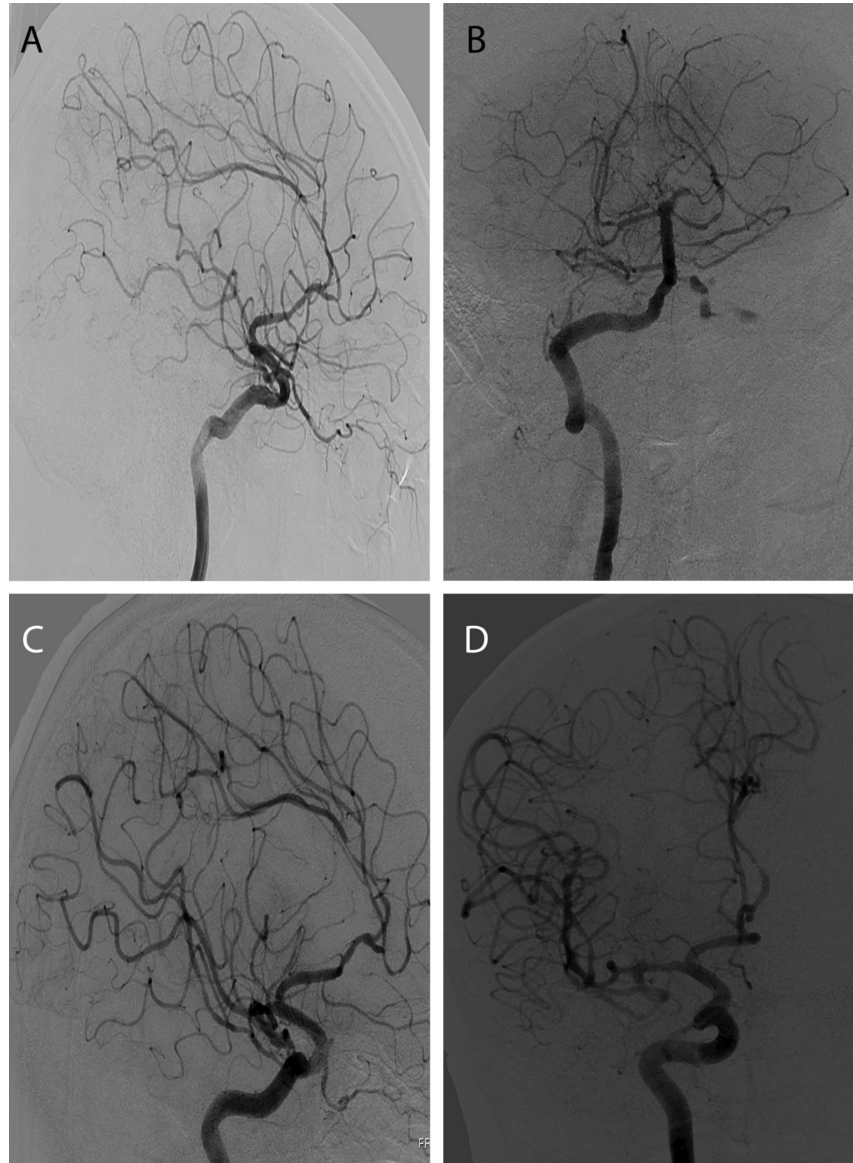
Figure 4. A, B, C, D) Widespread vascular irregularities consistent with vasculitis in cerebral angiography

findings, "possible" PACNS is diagnosed (4). There should be no CNS involvement secondary to systemic vasculitis or to infections for diagnosis (3).

Ethics

Peer-review: Internally peer-reviewed.

References

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