Case Report

Anterior Cerebral Artery Dissection in a Patient With Autosomal Dominant Polycystic Kidney Disease

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Intracranial artery dissection secondary to autosomal dominant polycystic kidney disease is far less common than cerebral aneurysm. A 55-year-old man presented a sudden onset of headache and disturbed consciousness caused by ischemic stroke in the bilateral frontal lobes with minor subarachnoid hemorrhage. The bilateral anterior cerebral arteries were firstly occluded and re-perfused with irregular narrowing and dilation in 3 days after stroke onset, indicating dissection. He was diagnosed with autosomal dominant polycystic kidney disease by abdominal CT findings and by his family history though his renal function was almost normal. Dissection in the anterior cerebral artery has not been reported previously, while some cases with dissection in the vertebral and extracranial arteries were reported in autosomal dominant polycystic kidney disease. His family also had a history of aortic dissection and subarachnoid hemorrhage. Intracranial artery dissection may be a manifestation of systemic arteriopathy with familial clustering in autosomal dominant polycystic kidney disease. Strict antihypertensive treatment is needed in these cases.

Key Words: Cerebral artery dissection—Autosomal dominant polycystic kidney disease—Anterior cerebral artery—Intracranial artery

Case

A 55-year-old man was admitted to a hospital with a sudden onset of headache, disturbed consciousness, and weakness of lower limbs. Brain MRI showed acute infarction in the bifrontal areas with faint subarachnoid hemorrhage (Fig 1, A,B). Initial MR angiography showed the anterior cerebral artery (ACA) occlusion. Three days after onset, cerebral angiogram showed re-perfused bilateral ACAs with irregular narrowing and dilation in the left ACA, indicating dissection (Fig 1, C,D). He was treated conservatively at the initial hospital. He became fully conscious again and was able to walk after 2-month rehabilitation. Seven months after onset, he was evaluated at our hospital. Asymptomatic hemorrhage in the right parietal lobe was newly observed in brain MRI (Fig 1, E). He had a history of hypertension and paroxysmal atrial fibrillation, and took both antiplatelet and anticoagulant drugs at the time of this evaluation. His blood pressure was not fully controlled with some antihypertensive drugs. The serum creatinine level was 0.9 mg/dL and he had mild proteinuria. In his family history, his father and brother had polycystic kidney disease and received dialysis. His brother had a history of subarachnoid hemorrhage. His ant survived aortic dissection. Abdominal CT scan showed multiple cysts in the bilateral kidneys (Fig 1, F). He was diagnosed as autosomal dominant polycystic kidney disease (ADPKD). He started to receive strict antihypertensive therapy and stopped taking antiplatelet agents to avoid hemorrhagic stroke.

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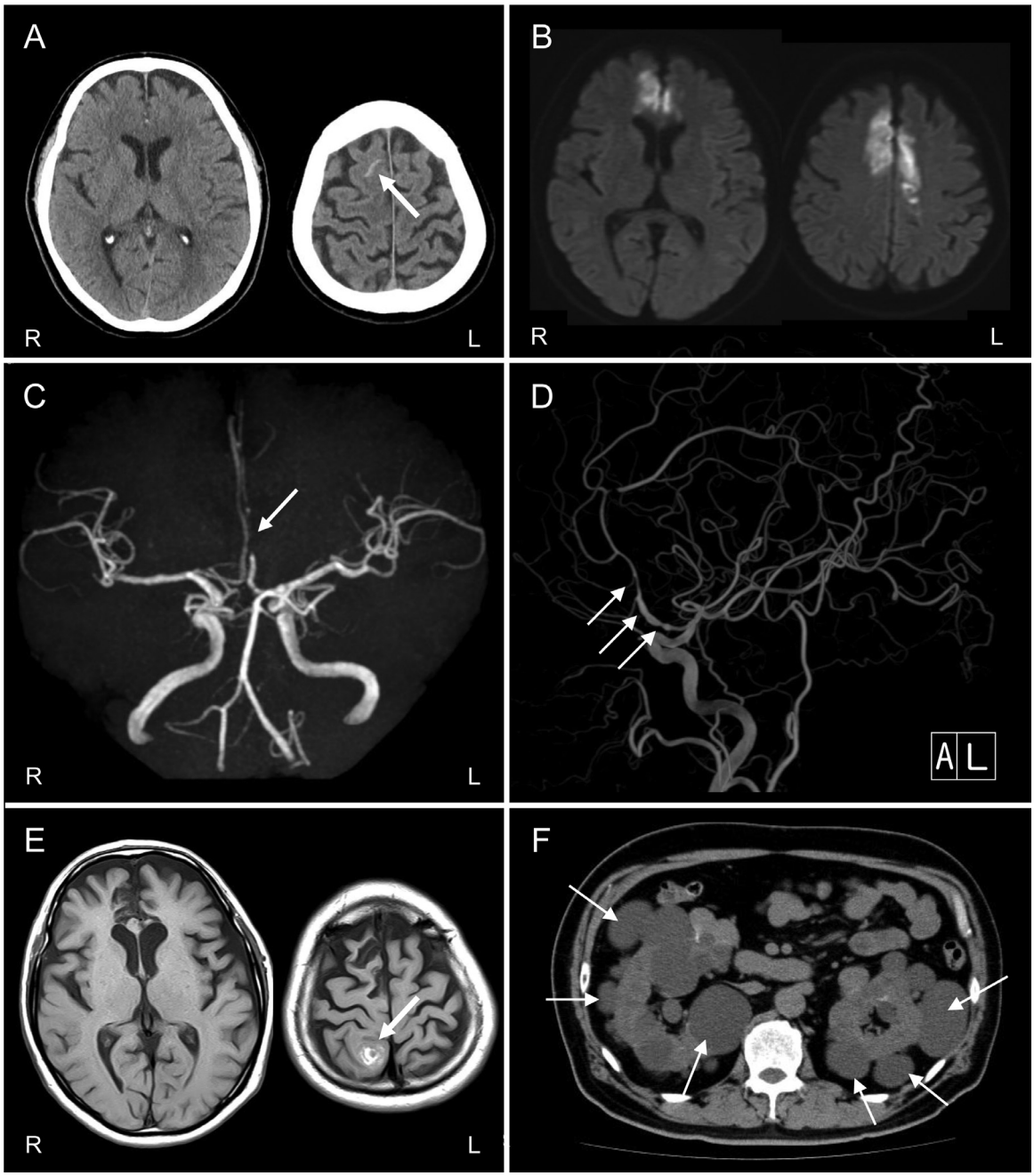

Figure 1. *(A) Initial brain CT revealed faint subarachnoid hemorrhage in the right frontal lobe (arrow). (B) Diffusion-weighted images of initial brain MRI showed acute infarction in the bilateral frontal areas. (C) Brain MR angiography performed 3 days after onset. The right anterior cerebral artery was re-perfused. There remained a stenosis in the left anterior cerebral artery (arrow). (D) Common carotid angiography (anterolateral view) performed 3 days after onset showed an irregular stenosis and dilatation in the left anterior cerebral artery (arrows). (E) T1-weighted images of brain MRI performed 7 months after onset showed an asymptomatic hemorrhage (arrow) in the right parietal lobe. (F) Abdominal CT scan showed multiple cysts in the bilateral kidneys (arrows).*

Discussion

ADPKD is a genetic renal disorder and is associated with an increased risk of cerebral aneurysms ranging from 4% to 33%.[1] Other vascular abnormalities such as aortic dissection,[2] dissection in the coronary artery,[3] and valvular heart diseases[4] were also reported in ADPKD. Intracranial arterial dissection is not commonly considered to be a feature of ADPKD, while several cases of vertebral and carotid artery dissection were reported.[5-7] Dissection in the ACA has not been reported previously. The causal relationship between ADPKD and ACA dissection is unclear because one of the risk factor of spontaneous intracranial artery dissection is hypertension, which is also common in ADPKD.

On the other hand, interruption of the internal elastic lamina in normal and dilated intracranial arteries was reported

in patients with ADPKD.[8,9] Schievink et al reported that the prevalence of intracranial arterial dolichoectasia was significantly higher in the patients with ADPKD (7 of 307 patients) than those without ADPKD (0 of 360 patients), and that some dolichoectasia arteries were suspected to be caused by dissection. They suggested that saccular aneurysm, intracranial arterial dissection, and dolichoectasia may all be manifestations in ADPKD arteriopathy.[9]

Moreover, prevalence of intracranial aneurysms was significantly higher in ADPKD patients with familial history of intracranial aneurysm or subarachnoid hemorrhage than those without such familial history.[10] Arterial dissection and other systemic arteriopathy in ADPKD may also have interfamilial heterogeneity. In the study of Schievink et al, most patients with dolichoectasia had additional vascular manifestations including arterial dissection.[9] In the present case, the patient's family had a history of aortic dissection and subarachnoid hemorrhage, and thus ACA dissection may also be a manifestation of systemic arteriopathy. Both hypertension and genetic vulnerability of elastic tissue may accelerate ADPKD arteriopathy and cause arterial dissection in some ADPKD patients with familial clustering. We should notice the family history in patients with intracranial artery dissection because renal function is normal in some cases with ADPKD. Strict antihypertensive treatment is needed not to develop arteriopathy in ADPKD.

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