The Seoul Journal of Medicine
Vol. 34, No. 1: 69-73, March 1993

Cerebral Aneurysm Associated with Adult Ploycystic Kidney Disease -Report of 2 Cases-

Bong-Soo Kim and Dae Hee Han

Department of Neurosurgery, Seoul National University College of Medicine, Seoul 110-799, Korea

=Abstract=**Two Cases of ruptured cerebral aneurysms associated with polycystic kidney diseases (PKD) are presented. Case 1 is a 60-year-old woman diagnosed as PKD after rupture of an anterior communicating artery aneurysm. Eleven days after successful aneurysmal clipping, she died of sudden massive intra-abdominal bleeding from kidney rupture. Case 2 is a 60-year-old man diagnosed as aneurysmal rupture 15 years after diagnosis of PKD. After anterior and posterior communicating artery aneurysms were clipped he recovered well. Since cerebral aneurysms is far more prevalent in patients with PKD than in the general population and aneurysm rupture is a leading cause of death, screening of the aneurysm by digital subtraction angiography or magnetic resonance angiography and prophylactic surgery of the aneurysm should be done.**

Key Words: *Polycystic kidney disease, Cerebral aneurysm, Subarachnoid hemorrhage, Hypertension*

INTRODUCTION

Cerebral aneurysm associated with polycystic kidney disease was first reported at the beginning of this century (Dunger 1904; Sieber 1905). Thereafter many cases have been reported. Since aneurysm rupture is one of the leading causes of death in polycystic kidney disease, it has been debated how to detect and manage aneurysms.

We report 2 cases of ruptured cerebral aneurysms associated with polycystic kidney disease. One was diagnosed as having PKD before and the other after the aneurysm rupture. The diagnostic and therapeutic implications are to be discussed.

Received January 1993, and in final form March 1993.
서울대학교 의과대학 신경외과학교실: 김봉수, 한대희

CASE REPORT

Case 1

This 60-year-old woman was admitted to Seoul National University Hospital because of a sudden headache, nausea and vomiting which developed 3 days before admission. She had hypertension for 15 years. There were no known family history of subarachnoid hemorrhage or polycystic kidney disease.

She was alert, oriented, and had no abnormal neurologic signs except nuchal rigidity. Her brain CT revealed subarachnoid hemorrhage in the sylvian and the suprasellar

cisterns. A carotid angiogram showed an anterior communicating artery aneurysm (Fig. 1).

She was operated on by left pterional approach. The carotid artery was atherosclerotic. A multilobulated aneurysm of the anterior communicating artery was clipped at the neck.

Postoperatively she did well until the 6th postoperative day when she became drowsy and disoriented. A brain CT showed no abnormal findings. But her abdomen distended and blood pressure dropped. Blood urea nitrogen was 23 mg/dl and serum creatinine was 0.7 mg/dl. Abdominal CT revealed polycystic disease involving the liver and both kidneys (Fig. 2). Eleven days after the operation she died of abdominal bleeding which was thought to be from rupture of kidney cyst.

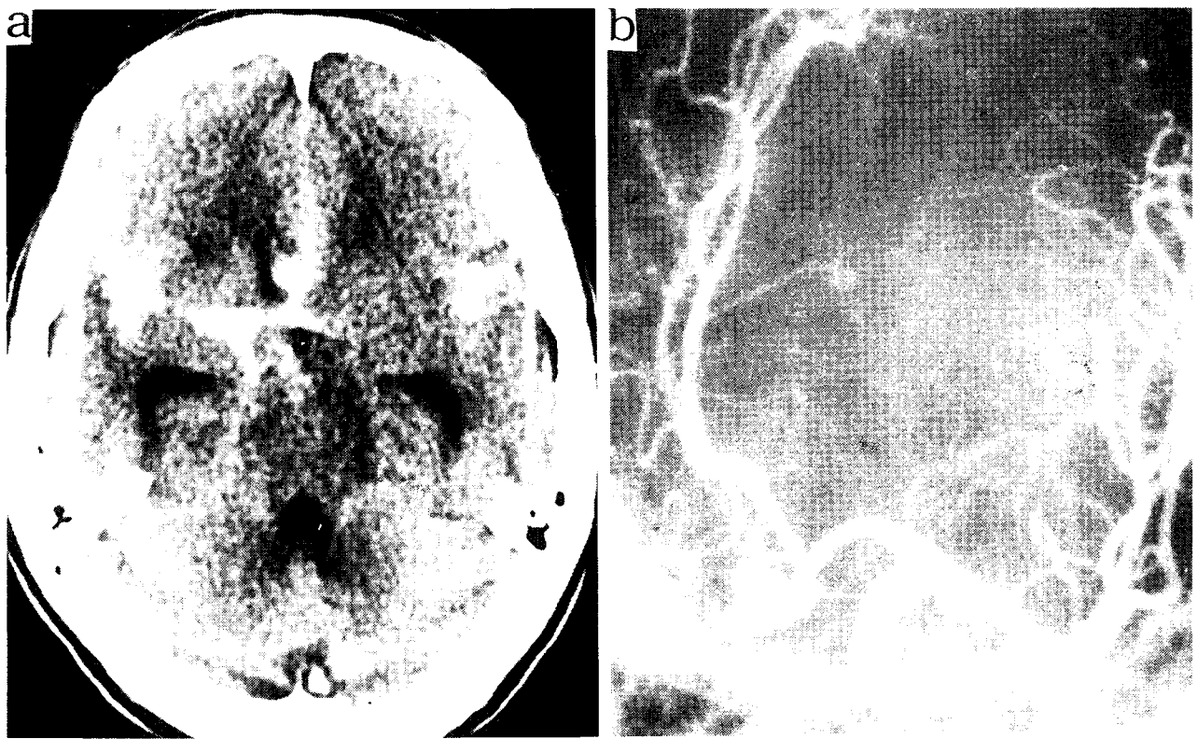

Fig. 1. Case 1. Brain CT showed subarachnoid hemorrhage in both the sylvian and the suprasellar cisterns (a) Right carotid angiogram showed a lobulated anterior communicating artery aneurysm (b).

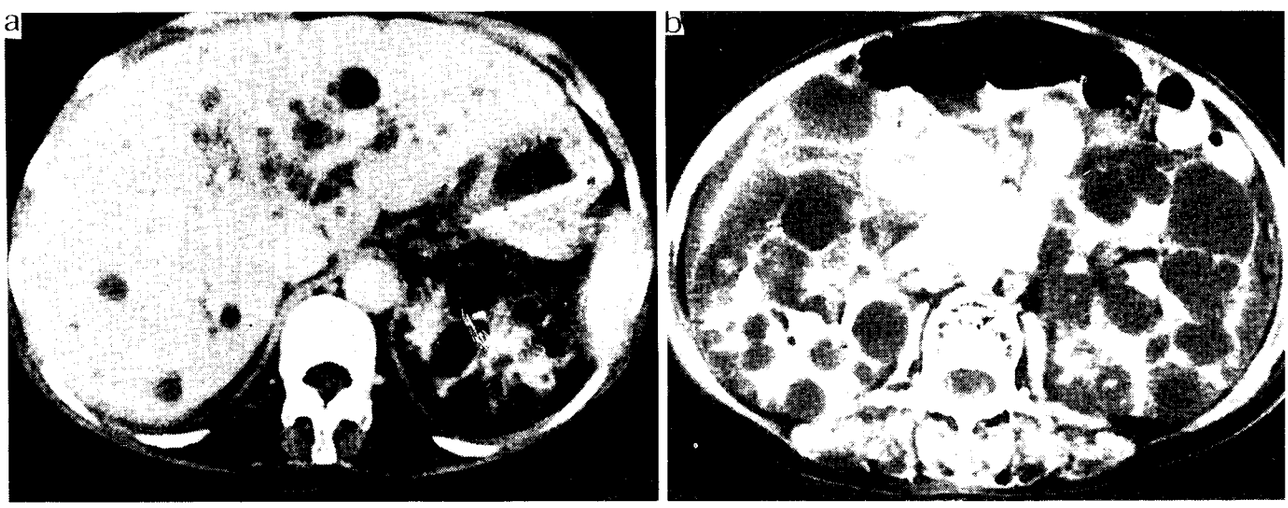

Fig. 2. Case 1. Abdominal CT revealed multiple cysts in the liver (a), and both kidneys (b).

Case 2

This 60-year-old man was admitted complaining of sudden headache which developed on the day of admission. He had had hypertension and polycystic kidney disease for 15 years and gout for 2 years. He had no known family history of subarachnoid hemorrhage or polycystic kidney disease.

On admission he was alert, oriented and had no other neurologic abnormality except nuchal rigidity. The brain CT showed subarachnoid hemorrhage in the sylvian, suprasellar and interhemispheric cisterns and also intracerebral hemorrhage in the right frontal area. A carotid angiogram revealed 2 aneurysms in the anterior communicating artery and right posterior communicating artery and also severe vasospasm (Fig. 3). Ultrasonography showed polycystic disease in both kidneys (Fig. 4).

He was operated on by right pterional approach and the two aneurysms were clipped at the neck in the same operation.

Postoperatively he showed left hemiparesis and memory defect. Brain CT confirmed cerebral infarction in the right middle cerebral artery and both anterior cerebral artery territory. These symptoms improved slowly and he was discharged with mild neurologic deficit.

DISCUSSION

In adult polycystic kidney disease (PKD) the renal tissue is replaced by numerous cysts which results in massive enlargement and functional failure of the kidney (Ziegler *et al.* 1981). The cysts may also involve the liver, pancreas and lung (Dalgaard 1957). It is considered a hereditary disease with autosomal dominant traits (Chester *et al.* 1977). The symptoms usually appear after the age of 30 (Hatfield and Pfister 1972). Cerebral aneurysms are frequently associated with PKD (Chung *et al.* 1982; Choi *et al.* 1985). The incidence is 7.3-16.6% (Sahs and Meyers 1951; Bigelow 1953) in autopsy series, and some angiographic studies show unruptured aneurysms in 41.2% of patients with PKD (Wakabayashi *et al.* 1983). Hypertension is a common presentation in aneurysm with PKD (Gabow *et al.* 1984). Some authors proposed this hypertension as a pathogenetic factor in the development of

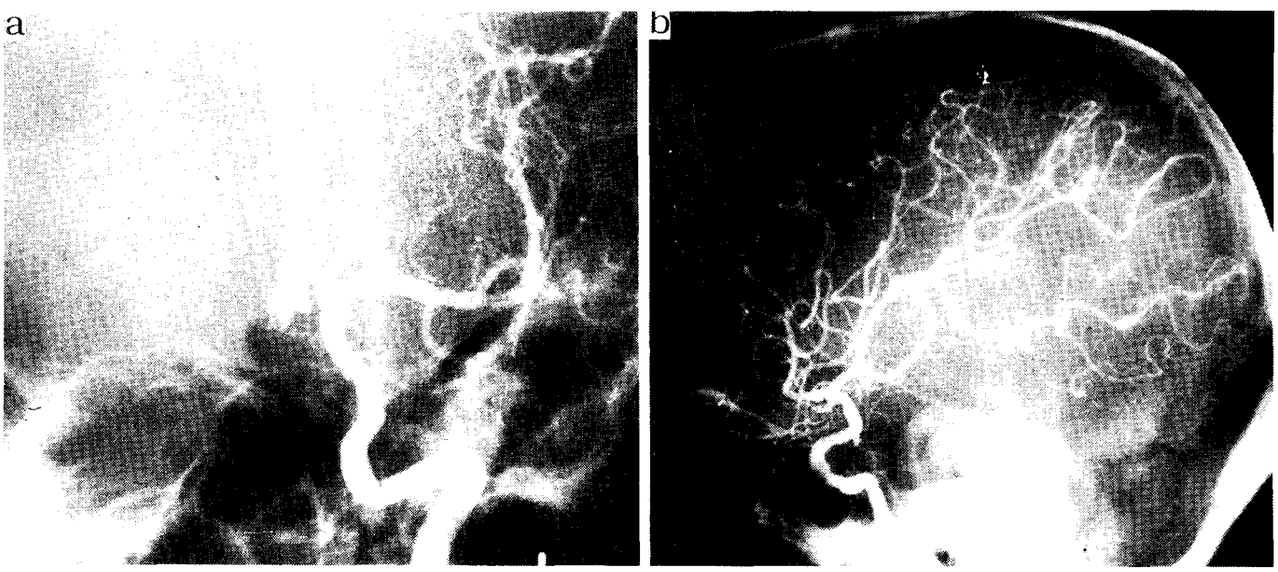

Fig. 3. Case 2. Anterior communicating and right posterior communicating artery aneurysms were seen in the left carotid angiogram (a) and the right carotid angiogram (b).

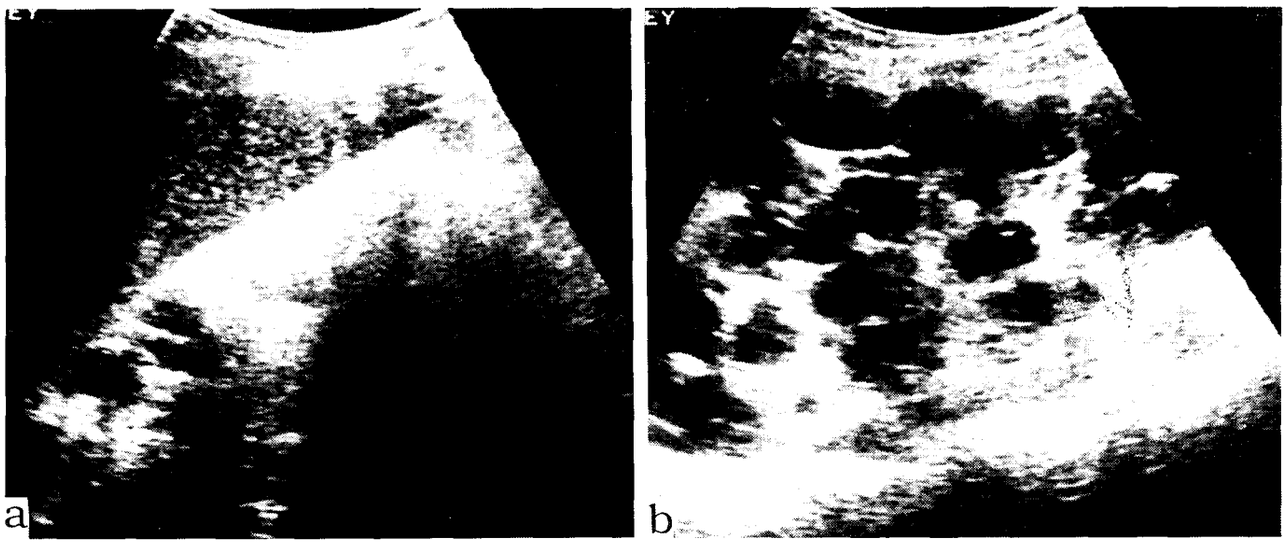

Fig 4. Abdominal sonogram of case 2 showed multiple cysts in the right kidney (a) and the left kidney (b).

aneurysm in PKD (Stehbens 1962; Ryu 1990) but others emphasized congenital factors (Ziegler *et al.* 1981). The location of the aneurysm is similar to that which is found in the general population, and multiple aneurysms are frequently found (Chapman *et al.* 1992). Both of our cases are 60 years old and also have hypertension. One had a single aneurysm in the anterior communicating artery and the other had multiple aneurysms in the anterior and posterior communicating arteries.

Aneurysmal rupture is one of the leading causes of death in patients with PKD. The average age at death in PKD is 73 years in asymptomatic patient, 52 years in symptomatic patients and 47 years in patients with intracranial aneurysms (Hatfield and Pfister 1972). The incidence of aneurysms in PKD is far more frequent than in the general population. So the need to screen for aneurysms in PKD has been emphasized (Wakabayashi *et al.*1983; Matsumura *et al.*1986). Conventional angiography is a definite method to demonstrate an aneurysm but sometimes it may cause serious complications. The screening method should be less invasive. High resolution CT or MRI can detect some aneurysms and it is recommended as an intial test by some authors (Chapman *et al.* 1992). But small aneurysms cannot be detected by this method. Digital subtraction angiography (DSA) or MR angiography (MRA) is a more sensitive and less invasive technique. Thus these methods are needed to screen unruptured aneurysms in patients with PKD. It has been known that patients with a family history of subarachnoid hemorrhage or cerebral aneurysm have a greater risk of aneurysms than those without such a family history (Kaehay *et al.* 1987; Saifuddin and Dathan 1987). In this group, conventional angiography is recommended for screening the aneurysm. Once the asymptomatic aneurysm is found it should be operated on. Aneurysmal rupture can be fatal and in recent days aneurysm surgery has shown relatively low mortality and morbidity rates (Hyun DK *et al.* 1992).

We report 2 cases of ruptured cerebral aneurysms in PKD and emphasize early detection by DSA or MRA and prophylactic surgery for this lesions.

REFERENCES

Bigelow NH. The association of polycystic kidneys with intracranial aneurysms and other related disorders. Am J Med Sci 1953; 225:485-94

Chapman AB, Rubinstein D, Hughes R, Stears JC, Earnest MP, Johnson AM, Gabaw PA. Intracranial

aneurysm in autosomal dominant polycystic kidney disease. N Engl J Med 1992; 327:916-20

Chester AC, Harris JP, Schreiner GE. Polycystic kidney disease. AM Fam Physician 1977; 16(6):94-101

Choi YK, Kim SD, Cho DK, Kim NS, Kim YJ. Two familes of polycystic kidney disease including one case of associated subarachnoid hemorrhage. Kyungpook Univ Med J 1985; 26:501-6

Chung YT, So JH, Choe SY, Chung WC, Won DS, Kim SU. A case of polycystic kidney disease associated with subarachnoidal hemorrahge. Korean J Int Med 1983; 26:191-6

Dalgaard OZ. Bilateral polycystic disease of the kidneys: a follow-up of 284 patients and their families. Acta Med Scand Suppl 1957; 328:22-194

Dunger R. Zur Lehre Von der Cystenniere, mit besonderer Berücksichtigung ihrer Heredität. Beitr Pathol Anat 1904; 35:445-509

Gabow PA, Ikle DW, Holmes JH. Polycystic kidney disease: Prospective analysis of nonazotemic patients and family members. Ann Intern Med 1984; 101:238-47

Hatfield PM, Pfister RC. Adult polycystic disease of the kidneys (potter type 3), JAMA 1972; 222:1527-31

Hyun DK, Bae HG, Lee KS, Yun IG, Byun BJ, Bae WK. Ruptured intracranial aneurysms in patients with polycystic kidney disease, Report of two cases. J Korean Neurosurgical Society 1992; 21:717-22

Kaehny W, Bell P, Earnest M, Stears J. Gabov P. Familial clustering of intracranial aneurysms in autosomal dominant polycystic kidney disease. Abstract from American Society of Nephrology, Kidney Int 1987; 31:204

Matsumara M, Wada H, Ohwada A, Shinoda T. Unruptured intracranial aneurysms and polycystic kidney disease. Acta Neurochir(Wien) 1986; 79:94-9

Ryu Shan-Jin. Intracranial hemorrhage in patients with polycystic kidney disease. Stroke 1990; 21:291-4

Sahs AL, Meyers R. The coexistance of intracranial aneurysms and polycystic kidney disease. Trans Am Neurol Assoc 1951; 76:147-50

Saifuddin A, Dathan E. Adult polycystic kidney disease and intracranial aneurysms. British Medical Journal 1987; 295:526

Sieber F. Uber Cystennieren bei Erwachsenen. Dtsch Z Chir 1905; 79:406-507

Stehbens WE. Cerebral aneurysms and congenital abnormalities. Aust Ann Med 1962; 11:102-12

Wakabayashi T, Fujita S, Ohbora Y, Suyama T, Tamaki N, Matsumoto S. Polycystic kidney disease and intracranial aneruysms: Early angiographic diagnosis and early operation for the unruptured aneurysm. J Neurosurg 1983; 58:488-91

Ziegler TW, Talner LB, Blantz RC. Cystic disease of the kidneys. In Brenner BM, Rector FC (Eds) The kidney, Ed 2, Vol 2. WB Saunders Co, Philadelphia, 1981; pp. 1872-905