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Clinical Analysis of Patients with Ruptured Cerebral Aneurysms associated with Polycystic Kidney Disease

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ABSTRACT

Objective : This study is to define the clinical characteristics and formulate the management strategies of the patients with ruptured cerebral aneurysms associated with polycystic kidney diseases (PKD). **Methods** : During the past 30 years, among of 3,013 patients who were treated with intracranial aneurysms, 7 patients had ruptured cerebral aneurysms associated with PKD. The authors retrospectively reviewed the database and imaging studies of such patients as sources for identification and analysis. **Results** : All 7 patients presented with subarachnoid hemorrhage (SAH). One patient showed Hunt and Hess grade I, 4 of grade II, and 2 of grade III. Six patients showed Fisher group II and 1 patient of group III. Four aneurysms were located at anterior cerebral artery, 2 at middle cerebral artery, and 1 at internal carotid artery. Five patients had small aneurysms and the remaining 2 had large (diameter $\geq$ 10 mm) aneurysms. All aneurysms were in saccular shape. Two of the 7 patients (28.6%) had multiple aneurysms. One patient suffered delayed ischemic neurological deficit. All patients were treated by microsurgery and showed favorable outcome (good: 7). **Conclusions** : The patients harboring PKD had high probability of hypertension. So, intact aneurysms in those patients were exposed to higher rate of being ruptured. Surgery was necessary for ruptured lesions, as well as unruptured lesions which size was increased at follow up imaging study. Multimodality management approach with nephrologist and neuroradiologist are very necessary. The ultimate management outcome was satisfactory. **(Kor J Cerebrovascular Surgery 9(3):212-5, 2007)**

KEY WORDS : Cerebral aneurysm · Polycystic kidney

Introduction

Patients with autosomal dominant polycystic kidney disease (ADPKD) have a higher incidence of intracranial aneurysms than the general population.[12)] These aneurysms also rupture at an earlier age in patients with ADPKD and are associated with high morbidity and mortality. This study is to analyze the clinical and radiologic characteristics of patients with ruptured cerebral aneurysms associated with polycystic kidney disease (PKD) and to define the optimal management strategies for such patients.

Patients and Methods

From September 1975 to August 2005, 3,013 patients were treated with intracranial aneurysms in our institute. Among them, 7 patients had ruptured cerebral aneurysms associated with PKD. The authors retrospectively reviewed the medical records and radiologic images. Database of 7 patients were analyzed for clinical characteristics, aneurysmal features, and management outcomes (Table 1).

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Results

Among 7 patients, sex ratio was 3 males and 4 females. Average age was 46.9 year old (range: 28~60). At admission, 1 patient showed Hunt and Hess grade I, 4 of grade II, and 2 of grade III. Six patients showed Fisher group II and 1 of group III. The aneurysms were located at midline in 3 patients, right hemisphere in 3 patients, and left hemisphere in 1 patient. Four aneurysms were located at anterior cerebral artery, 2 at middle cerebral artery, and 1 at internal carotid artery. The diameter of aneurysm ranged from 3 to 20mm, having average of 8.1mm. Five patients had small aneurysms (diameter<10mm) and the remaining 2 had large aneurysms (diameter≥10mm). All aneurysms were in saccular shape. Two of the 7 patients (28.6%) had multiple aneurysms. Six patients had ruptured cerebral aneurysm at first time, and the remaining 1 was recurred patient. One patient suffered from delayed ischemic neurological deficit. Elective surgery was performed in 5 patients within 3 days of rupture, and 2 patients received delayed surgery after 2 weeks of rupture. The surgical approach was pterional approach in 6 patients and interhemispheric approach in 1 patient. The average postoperative follow up period was 38.6 (range 4~123) months after surgery. Modified Glasgow Outcome Scale showed favorable outcome (good: 7) in all patients.

Discussion

Overview of polycystic kidney diseases

Polycystic kidney disease (PKD) may be hereditary or acquired.[9] The incidence of autosomal dominant polycystic kidney disease (ADPKD) is about 1 in 400~1000 live births and can arise from mutations in 2 different genes, named PKD1 located on chromosome 16p13.3, and PKD2 located on chromosome 4q21~23.[13] ADPKD has characteristics of chronic renal failure having early development of multiple cysts in both kidneys.[14] Therefore, renal function failures such as hypertension, hematuria, nephritis, and renal stones develop.[9] These cysts develop in size as cysts in tubules and collecting ducts grow. They are associated with systemic diseases of liver, testis, ovary, brain, spleen, and pancreas. Also, it is associated with hypertension, cardiac valve defect, cerebral aneurysm, coronary artery aneurysm, aortic artery aneurysm, abdominal and inguinal hernias.[2][9] The allopathic treatment is performed with balanced control of sodium and protein, control of hypertension and renal stones. At the end

Table 1. seven patients were analyzed for clinical characteristics, aneurysmal features, and management outcomes

No	Sex	Age	Fisher Group	Hunt & Hess Grade	Site	Location	Diameter (mm)	Shape	Multiple aneurysm	Number of bleeding	ADPKD / PKD	Other medical hystory	hydrocephalus	Timing of operation (days)	Approach	Outcome
1	M	49	2	2	left	A2-3	5	saccular		1	ADPKD	Liver cysts, HTN	-	27	Interhemispheric	good
2	F	60	2	1	right	ICPCOM	5	saccular	L.ICPCOM	1	ADPKD	HTN	-	26	pterional	good
3	F	43	2	3	right	MCBIF	20	saccular		1	ADPKD	HTN	-	1	pterional	good
4	F	50	2	2	-	ANCOM	5	saccular		2	PKD	ESRD, HTN	+	2	pterional	good
5	M	51	2	2	-	ANCOM	10	saccular		1	PKD	ESRD, HTN	-	3	pterional	good
6	F	47	2	2	right	MCBIF	9	saccular		1	ADPKD		-	2	pterional	good
7	M	28	3	3	-	ANCOM	3	saccular	BABIF, R.VA	1	PKD	Colon cancer, HTN	-	2	pterional	good

(M: male, F: female, A: anterior cerebral artery, ICPCOM: internal carotid artery-posterior communicating artery, MCBIF: middle cerebral artery bifurcation, ANCOM: anterior communicating artery, BABIF: basilar artery bifurcation, VA: vertebral artery, L: left, R: right, ADPKD: autosomal dominant polycystic kidney disease, PKD: polycystic kidney disease, ESRD: ens-stage renal disease, HTN: hypertension)

stage of renal failure, dialysis and renal transplantation are needed.[9] In contrast, ARPKD is relatively rare and has clinical symptoms from the birth. It has high rate of mortality within 1 month of birth. The PKD is diagnosed mainly through ultrasonography.[4]

Cerebrovascular complications are an important cause of morbidity and mortality in patients with ADPKD. According to Fick et al.[5]'s series, the neurologic event was the cause of death in 12% of 129 ADPKD patients (ruptured aneurysms in 6%, hypertensive intracranial hemorrhage in 5%, and ischemic stroke in 1%). The mean age of those who died of ruptured aneurysm was 37 yr. Rivera et al.[11] reported 19 attacks of acute stroke in 14 (9.8%) patients among 142 ADPKD patients. Six (4.2%) patients had intracranial hemorrhage (3 ruptured aneurysms and 3 intracerebral hemorrhages). Nine patients suffered from ischemic events (5 cerebral infarctions and 4 transient ischemic attacks). They observed that patients with ischemic attacks had a better outcome than patients with hemorrhagic attacks.[11]

PCK disease associated with ruptured cerebral aneurysms

Characteristics of cerebral aneurysm in development and rupture

Approximately 8% of ADPKD patients have intracranial aneurysms.[6] The percentage of accompanying cerebral aneurysm in ADPKD patients is 5-times higher than the general population,[8] yet the ratio of ADPKD in cerebral aneurysm patients are very rare.[12] Among these patients, cerebral aneurysm rupture develop at 10 years younger age,[14] dominant in male patients and middle cerebral artery.[3][12] Also, subarachnoid hemorrhage (SAH) is one of the main causes of death.[12] Periodic and long term follow up using magnetic resonance image (MRI) is in demand due to high probability of redeveloping aneurysm.[10] It is also known that there is possibility of underlying aneurysm might grow larger.[1] There is a report that half of the patients had normal renal function at the time of rupture, and 11% were receiving dialysis.[3]

Prophylactic treatment of asymptomatic unruptured cerebral aneurysm

Yanaka et al.[14] reported that degrading of renal function failure can be prevented by prophylactic hemodialysis after the cerebral angiography in patients with intact cerebral aneurysm associated with PKD having normal renal function.

Cerebral aneurysm screening using MRI

Gibbs et al.[6] have reported that almost all of cerebral aneurysm was in saccular shape based on MRI study, especially in 3-dimensional time-of-flight MR angiography and 3-dimensional phase-contrast sequences. All of saccular shaped aneurysm had average of 3.5mm diameter, and 77% was located in anterior circulation. Upon following observation, the development and rupture of cerebral aneurysm had no difference compared to general population. Therefore, ADPKD patients receiving broad screening examination to detect cerebral aneurysm is of no help.[6] Hughes et al.[7] also reported that cerebral aneurysm screening examination in ADPKD patients having no history of SAH is not sufficient based on Risk-benefit analysis.

Conclusion

The patients harboring PKD had high probability of hypertension. So, intact aneurysms in those patients were exposed to higher rate of being ruptured. Surgery was necessary for ruptured lesions, as well as unruptured lesions which size was increased at follow up imaging study. Multimodality management approach with nephrologist and neuroradiologist are very necessary. The ultimate management outcome was satisfactory.

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