Polycystic kidney disease and intracranial aneurysms

Early angiographic diagnosis and early operation for the unruptured aneurysm

TOSHIMITSU WAKABAYASHI, M.D., SHIGEKYO FUJITA, M.D., YOSHIRO OHBORA, M.D., TOHRU SUYAMA, M.D., NORIHIKO TAMAKI, M.D., AND SATOSHI MATSUMOTO, M.D.

Department of Neurosurgery, Hyogo Brain and Heart Center, Himeji, and Department of Neurosurgery, Kobe University School of Medicine, Kobe, Japan

From August, 1981, to August, 1982, the authors performed four-vessel angiography in 17 patients with polycystic kidney disease (PKD) who had no neurological deficit and no history of subarachnoid hemorrhage. Seven cases of unruptured aneurysms were found among these 17 patients (an incidence of 41.2%). Five of the unruptured aneurysms were operated on prophylactically, with no mortality or morbidity. Nine of the 17 patients had hypertension and, of these, two (22.2%) had aneurysms. Of the eight patients without hypertension, five (62.5%) had aneurysms. This study suggests that the coexistence of PKD and intracranial aneurysms might not be due to the hypertension that occurs concomitant with PKD, but instead may be attributable to congenital factors. The authors stress the necessity of early diagnosis and early operation for unruptured aneurysms in patients with PKD.

KEY WORDS • polycystic kidney disease • intracranial aneurysm • early diagnosis • early operation • congenital factor • angiography

There have been many reports on the coexistence of polycystic kidney disease (PKD) and intracranial aneurysms since 1904 when Dunger discussed the frequent association of these two conditions. The incidence of intracranial aneurysms in PKD series has been reported to be 17%, based on autopsy studies. Once rupture of the aneurysm has occurred in patients with PKD with severe renal dysfunction, the prognosis is poor. Therefore, it is reasonable to establish the diagnosis early, and to perform prophylactic surgery for these unruptured intracranial aneurysms.

Although the value of early diagnosis and early operation has been recognized, there have been very few series emphasizing the necessity of cerebral angiography for detecting unruptured intracranial aneurysms in patients with PKD. Furthermore, there has been no report of the use of angiography in the diagnosis of unruptured aneurysms in PKD patients. We have recently performed four-vessel angiography in a series of 17 patients with PKD so as to detect unruptured intracranial aneurysms. This paper presents our findings.

Clinical Material and Methods

The diagnosis of PKD was made in 17 patients from 10 different families. These patients had no neurological deficit and no history of subarachnoid hemorrhage (SAH). Four-vessel angiography was performed in these patients over a 1-year period from August, 1981, to August, 1982. There were seven males and 10 females, ranging in age from 32 to 66 years, with an average age of 42 years.

Computed tomography (CT) was used in the diagnosis of PKD, being a noninvasive method with a high level of accuracy compared to other modalities. A CT scan was ordered for four patients with abdominal pain and two with hematuria. The other 11 patients underwent CT scanning for detection of PKD because they had siblings with polycystic kidneys. The patients were examined for hypertension and renal dysfunction. Hypertension was defined as a blood
TABLE 1
Data on seven patients with unruptured aneurysms and PKD

<table>
<thead>
<tr>
<th>Age (yrs), Sex</th>
<th>Parent with PKD</th>
<th>Location of Aneurysm</th>
<th>Hypertension</th>
<th>Renal Failure</th>
<th>Operation</th>
</tr>
</thead>
<tbody>
<tr>
<td>46, F</td>
<td>mother</td>
<td>ICA-PCoA</td>
<td>-</td>
<td>-</td>
<td>muscle wrapping</td>
</tr>
<tr>
<td>32, F</td>
<td>mother</td>
<td>ICA-PCoA</td>
<td>-</td>
<td>-</td>
<td>muscle wrapping</td>
</tr>
<tr>
<td>50, M</td>
<td>unknown</td>
<td>MCA</td>
<td>-</td>
<td>-</td>
<td>clipping</td>
</tr>
<tr>
<td>42, M</td>
<td>unknown</td>
<td>MCA</td>
<td>-</td>
<td>-</td>
<td>clipping</td>
</tr>
<tr>
<td>66, M</td>
<td>unknown</td>
<td>ACA</td>
<td>+</td>
<td>+</td>
<td>clipping</td>
</tr>
<tr>
<td>48, F</td>
<td>father</td>
<td>ICA-PCoA</td>
<td>+</td>
<td>+</td>
<td>clipping</td>
</tr>
<tr>
<td>66, F</td>
<td>mother</td>
<td>MCA</td>
<td>+</td>
<td>-</td>
<td>not done</td>
</tr>
</tbody>
</table>

* PKD = polycystic kidney disease; ICA-PCoA = internal carotid-posterior communicating artery; MCA = middle cerebral artery; ACA = anterior cerebral artery; BA-SCA = basilar-superior cerebellar artery. + = present; - = absent.

TABLE 2
Incidence of hypertension in 17 cases of unruptured aneurysm

<table>
<thead>
<tr>
<th>Associated Finding</th>
<th>Hypertensive Group</th>
<th>Non-hypertensive Group</th>
<th>Total Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>aneurysm*</td>
<td>2</td>
<td>5</td>
<td>7</td>
</tr>
<tr>
<td>no aneurysm</td>
<td>7</td>
<td>3</td>
<td>10</td>
</tr>
<tr>
<td>total</td>
<td>9</td>
<td>8</td>
<td>17</td>
</tr>
</tbody>
</table>

* These patients are summarized in Table 1.

pressure of greater than 160/95 mm Hg; renal dysfunction was considered to be present when the blood urea nitrogen was greater than 20 mg/dl and the serum creatinine level was greater than 2.0 mg/dl.

During the course of this study, abdominal CT scanning was performed in 36 other patients with ruptured intracranial aneurysms to determine if PKD was present.

Results

Of the 17 patients with PKD, unruptured aneurysms were discovered in seven (41.2%). In two patients the aneurysms were located on the internal carotid-posterior communicating artery (ICA-PCoA), in three on the middle cerebral artery, in one on the distal anterior cerebral artery, and in one patient there were multiple aneurysms, with involvement of the ICA-PCoA and basilar-superior cerebellar artery. Five of these seven patients were operated on for treatment of their unruptured aneurysms, without mortality or morbidity. The operative procedure consisted to clipping in three cases and muscle wrapping in two. The other two patients had hypertension and were not operated on. Details of these seven cases are summarized in Table 1.

Among nine patients with hypertension there were two cases with aneurysm (22.2%), and among eight non-hypertensive patients there were five cases with aneurysm (62.5%) (Table 2). In addition, infundibular dilatation was observed in six of these 17 cases. Polycystic kidney disease was diagnosed by abdominal CT in two of the 36 other patients who were operated on for ruptured intracranial aneurysms during the same period (Table 3).

Discussion

Many definitions of PKD have been proposed, but the classification of Osathanondh and Potter has been adopted in this study. Adult PKD is often confused with other forms. We have chosen to call the adult-type of the disease (Potter type 3) simply "polycystic kidney disease" in this series. In patients suffering PKD, the kidneys are massively enlarged by numerous cysts (Fig. 1). Cysts are sometimes observed in the liver, pancreas, lung, or spleen. The disease is transmitted as an autosomal dominant trait (Fig. 2). The incidence of PKD in autopsy series was reported by Brown to be 0.3%. Polycystic kidney disease is diagnosed in about one of every 3000 patients, and accounts for 5% of the hemodialysis population.

Fig. 1. Abdominal computerized tomography scan of a patient with polycystic kidney disease. Massively enlarged kidneys are observed, with a spot indicating calcification, and numerous cysts.
Symptoms first occur in most of the symptomatic patients at the age of 35 to 50 years. There have been many reports regarding the familial occurrence of intracranial aneurysms. Some authors have attributed this phenomenon to heredity, but in many cases the finding of intracranial aneurysms among family members was fortuitous. There are three disorders noted to be frequently associated with intracranial aneurysms: coarctation of the aorta, Ehlers-Danlos syndrome, and PKD. The coexistence of intracranial aneurysms and PKD has been described in numerous reports since Dunger published his account of the association of these two lesions in 1904. The first case was reported by Borelius in 1901.

The incidence of intracranial aneurysms in PKD series has been variously reported as 7.3%, 16.6%, and 16.5% (Table 4). These findings were based on autopsy studies. Brihaye and Toppet, and Hatfield and Pfister recommended that all patients with PKD undergo cerebral angiography. Brackett and Morantz recommended early diagnosis and early operation for patients with PKD. However, the incidence of intracranial aneurysms in PKD patients who underwent cerebral angiography has not been described previously. Ours is the first report of this kind that we know of.

In our present series of 17 patients, the incidence of unruptured aneurysms was 41.2% in the patients with PKD and no history of SAH (Table 4). This incidence might be excessive, partly because of the small number of cases. Also, the higher incidence might be attributed to the fact that the small aneurysms which are often overlooked in autopsies can be detected by cerebral angiography. Some authors stated the difficulty in finding unruptured aneurysms or small aneurysms in autopsy specimens. In addition, racial difference should be considered in accounting for the marked disparity in incidence between our series and those reported previously.

The connection between PKD and intracranial aneurysms has been the subject of controversy. Congenital factors are emphasized by some, whereas others have proposed that the hypertension that accompanies PKD may result in aneurysm formation. In this series, hypertension was noted in two of seven patients with unruptured aneurysms (28.6%). A comparison of cases with and without hypertension (Table 2) suggests that the coexistence of PKD and intracranial aneurysms might not be attributable to the concomitant hypertension. Consequently, the coexistence of these lesions might be due to congenital factors.

Once SAH has occurred, some patients suffer chronic renal failure, requiring hemodialysis due to blood volume reduction to the kidneys or due to the damage caused by hyperosmotic agents. The average age at death in PKD is 73.0 years in asymptomatic patients, 51.8 years in symptomatic patients, and 46.7 years in patients with intracranial aneurysms.

In the case of an unruptured aneurysm with no surgical treatment, the risk of SAH is 11.5% during a 10-year follow-up period, with a 6.6% incidence of mortality, according to a report by Heiskanen. There was a 10% incidence of mortality during a 5-year period in the series of Mount and Brisman. Salazar reported no deaths among 29 patients operated on for unruptured intracranial aneurysms. Likewise, we experienced no mortality or morbidity in our series, in which 12 unruptured aneurysms were operated on, including the five associated with PKD.

We used CT scanning for screening of PKD because of its accuracy and noninvasiveness; CT can detect very small cysts (1 cm or less). Excretory urography, on the other hand, was reported to confirm the diagnosis in only 47% of cases in a series of patients with PKD. In the management of patients with PKD, it is important not to lower the blood pressure too greatly and not to administer hyperosmotic agents (such as mannitol or glycerol) excessively. Early detection and operation are indicated for the patient with PKD and unruptured intracranial aneurysm.

In this series, a 5.6% incidence of PKD among aneurysm patients (two of 36 patients) was approximately the same as the 3.1% incidence in the series of Sahs and Meyers, 4.4% in the series of Brown, and 6.4% in the series reported by Bigelow.
References

5. Brown RAP: Polycystic disease of the kidneys and interrelationship of these conditions: review of recent literature and report of seven cases in which both conditions coexisted. Glasgow Med J 32:333–348, 1951

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Address reprint requests to: Toshimitsu Wakabayashi, M.D., Department of Neurosurgery, Hyogo Brain and Heart Center, 520 Ko Saisho Himeji, Hyogo, Japan.