Ruptured Cerebral Aneurysm Associated with Coarctation of the Aorta

—Report of Two Cases—

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Abstract

Two cases of ruptured cerebral aneurysm associated with coarctation of the aorta are presented. The aneurysms were successfully clipped in the acute stage prior to correction of the coarctation. Ruptured aneurysm should be treated as early as possible, and unruptured aneurysm should also be treated before aortic repair, if the general condition of the patient allows.

Key words: ruptured cerebral aneurysm, coarctation of the aorta, multiple aneurysms, surgical indication

Introduction

Cerebral aneurysm is associated with coarctation of the aorta in 2–10% of cases, 2.7% of which die from aneurysmal rupture. However, the incidence of coarctation of the aorta among cerebral aneurysm patients is very low, ranging from 0.19 to 1.9% (mean, 0.45%) and in our institute 0.22% (Table 1).

Here, we present two cases of ruptured cerebral aneurysm with coarctation of the aorta, in which the ruptured cerebral aneurysms were successfully clipped during the acute phase of subarachnoid hemorrhage. The clinical features, and timing and order of surgical intervention for the ruptured cerebral aneurysm and coarctation of the aorta in addition to accompanying unruptured cerebral aneurysm are discussed.

Case Reports

Case 1: A 33-year-old female presented on August 15, 1989 with consciousness disturbance. She had a history of hypertension due to coarctation of the aorta 2 years before, and sudden severe headache and paresthesia 1 year before. Intravenous digital subtraction angiograms at a local hospital had revealed a right middle cerebral artery (MCA) aneurysm, but she refused surgery.

On admission to our hospital, she was semicomatose and exhibited left decerebrated posture (Hunt & Kosnik grade IV). The right pupil was dilated and fixed to light while the left was nor-

Table 1 Incidence of aortic coarctation associated with cerebral aneurysm

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>No. of cerebral aneurysms</th>
<th>No. of coarctations</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Walton (1956)</td>
<td>312</td>
<td>1</td>
<td>0.32</td>
</tr>
<tr>
<td>Stenhens (1962)</td>
<td>215</td>
<td>4</td>
<td>1.9</td>
</tr>
<tr>
<td>DuBoulay (1965)</td>
<td>197</td>
<td>2</td>
<td>1.0</td>
</tr>
<tr>
<td>Robinson (1967)</td>
<td>545</td>
<td>2</td>
<td>0.37</td>
</tr>
<tr>
<td>Sedzimir et al. (1973)</td>
<td>1066</td>
<td>2</td>
<td>0.19</td>
</tr>
<tr>
<td>Yokota et al. (1977)</td>
<td>195</td>
<td>2</td>
<td>1.0</td>
</tr>
<tr>
<td>Fukuda et al. (1985)</td>
<td>154</td>
<td>1</td>
<td>0.65</td>
</tr>
<tr>
<td>This work</td>
<td>902</td>
<td>2</td>
<td>0.22</td>
</tr>
<tr>
<td>All series</td>
<td>3586</td>
<td>16</td>
<td>0.45</td>
</tr>
</tbody>
</table>
mal. Blood pressure was 270/160 mmHg, with no discrepancy between the right and the left upper extremities. The bilateral femoral pulses were very feeble and delayed. A chest x-ray film demonstrated no obvious rib notch. Computed tomographic (CT) scans demonstrated subarachnoid hemorrhage with right Sylvian hematoma (Fig. 1). Right direct carotid angiograms showed the large right MCA aneurysm and two right distal anterior cerebral artery (ACA) aneurysms (Fig. 2 left). Nine hours after onset, a right decompressive frontotemporal craniectomy and a neck clipping of the MCA aneurysm were performed.

Two weeks later, she became stuporous with left hemiparesis. Postoperative angiograms by Seldinger's method via the right brachial artery on September 24 visualized only the right carotid, right vertebral, and aortic arteries due to severe elongation (Fig. 3). Direct left carotid angiograms demonstrated a left internal carotid-posterior communicating artery (IC-PCoA) aneurysm and a distal left ACA aneurysm (Fig. 2 right). The left IC-PCoA aneurysm and the three distal ACA aneurysms were clipped and cranioplasty carried out on October 4. Her postoperative course was uneventful except for transient ventricular dilatation.

She was transferred for cardiovascular treatment with mild confusion and left hemiparesis on November 30. However, the aortic coarctation could not be corrected because of extension beyond the renal artery. She is now at home with mild left hemiparesis.

Case 2: A 15-year-old boy was admitted on December 10, 1990 with sudden severe headache. A chest x-ray film had disclosed cardiomegaly when he was 9 years old. Sudden onset of severe headache had occurred at 12 years old.

On admission, his blood pressure was 130/90 mmHg in the bilateral upper extremities. The bilateral femoral pulses were relatively weak. His consciousness was drowsy and confused (Hunt & Kosnik grade III), and mild left hemiparesis was present. A chest x-ray film did not demonstrate the rib notch.
CT scans demonstrated subarachnoid hemorrhage, predominantly in the interhemispheric fissure and basal cistern, intraventricular hemorrhage in all ventricles, and intracerebral hematoma in the right frontal lobe (Fig. 4). Right carotid angiograms showed a large anterior communicating artery (ACoA) aneurysm (Fig. 5). Eight hours later, a right decompressive frontotemporal craniectomy and aneurysmal neck clipping were performed under hypothermic anesthesia.

Two weeks later, he became confused and disinhibited. On January 18, 1991, cerebral angiograms by the Seldinger’s method via the right femoral artery encountered some difficulties due to aortic elongation and stenosis, and demonstrated no residual aneurysm. Aortograms at the same time demonstrated moderate coarctation of the descending aorta (Fig. 6). The pressure gradient was 60 mmHg. Cranioplasty was performed on January 21. He was discharged without neurological deficits on January 30. The coarctation was repaired 6 months later.

Discussion

Ruptured cerebral aneurysm associated with coarctation of the aorta has three characteristic clinical features. First, aneurysmal rupture with coarctation occurs much earlier in life (mean, 25 yrs) than without coarctation (mean, 50–54 yrs). The incidence of bleeding from cerebral aneurysm in young patients with coarctation ranges from 12 to 23%, suggesting that coarctation of the aorta is a very important cause of subarachnoid hemorrhage in youth.

Second, the incidence of multiple aneurysms is higher (30%) than in the absence of coarctation (20%), requiring complete cerebral angiography. Brackett and Morants recommended angiography via the brachial artery for selective catheterization of cerebral arteries, and for identifying aortic lesions. However, the more rapid combination of left direct carotid and right retrograde brachial angiography is the first choice for preoperative examination in the acute phase, because selective catheterization via the
brachial artery might be very difficult and that via the femoral artery often impossible to pass through the aortic stenosis.

Finally, the mortality due to ruptured cerebral aneurysm associated with coarctation is much higher (50–75%), mainly because of rebleeding from the cerebral aneurysm. 1,13) Sedzimir et al. 1) and Schwartz and Baronofsky 10) treated the coarctation before the ruptured aneurysm. This order, however, has a high risk of perioperative rebleeding from the cerebral aneurysm and aggressive induced hypertensive therapy against vasospasm is unsafe. Recent advances in anesthesia, perioperative management, and neurosurgical techniques allow surgical treatment of the ruptured cerebral aneurysm in the acute phase in the presence of untreated coarctation with relative safety. Therefore, the ruptured aneurysm should be treated as early as possible before correcting the coarctation. Generally, unretracted aneurysms should also be treated before aortic repair. However, if heart failure is apparent or clipping of the unretracted aneurysm is difficult, the coarctation should be repaired first.

Early surgery for ruptured aneurysm requires several special postoperative precautions. A most important precaution is to maintain the blood pressure high enough to prevent ischemic complications due to vasospasm, which can worsen the prognosis after clipping of ruptured cerebral aneurysm even in young patients like ours. However, induced hypertension might cause heart failure, cerebral swelling, and rupture of accompanying unretracted aneurysm. Cardiac function was evaluated with pulmonary capillary wedge pressure in Case 1 and central venous pressure in Case 2. External decompression and ventricular drainage were very effective for controlling intracranial pressure during induced hypertension therapy in both cases, although Case 1 required hyperosmolar agents for a short time postoperatively. Blood pressure was easily controlled in our cases by continuous intravenous administration of low doses of nicardipine or dopamine. When the blood pressure must be kept low, care should be to prevent ischemia of the kidney or the spinal cord.2,4,5,7,9-11,14) The urinary output and blood pressure or at least the arterial pulsation and motor function of the lower extremities must be checked every hour.

Both our cases had hematoma, in the Sylvian fissure in Case 1, and in the right frontal lobe in Case 2. Such intracerebral hematoma due to ruptured cerebral aneurysm with coarctation has not been previously described. These hematomas might be incidental or due to hypertension caused by coarctation or subarachnoid adhesions caused by previous minor bleeding from the cerebral aneurysm. This suggests that the intracerebral hematoma with coarctation previously reported6,8 might have been caused by cerebral aneurysmal rupture.

References

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