Intracranial arterial aneurysms in children and adolescents

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Throughout the period 1943 to 1980, 1368 patients with verified intracranial saccular aneurysms were treated in the University of Aarhus neurosurgical department. Forty-three (3.1%) patients (25 boys and 18 girls) were 19 years old or younger, and 33 (77%) had an onset of symptoms typical of subarachnoid bleeding. Using the classification system of Hunt and Hess as a basis for clinical assessment on admission, 58% of the patients could be placed in Grade I or II. Cerebral vasospasm was demonstrated in 53% of the patients undergoing angiography between the 4th and 16th day after hemorrhage. There was no increased morbidity or mortality in the group of patients with vasospasm, and no cerebral infarction was demonstrated at necropsy. Therefore, it is possible that vasospasm is of minor prognostic significance in children. In 15 patients (37%), aneurysm rupture was accompanied by intracerebral hematoma. The mortality rate in this group of 15 patients was 50%, whereas in the group without hematoma it was 26%. The overall mortality rate was 33%. The surviving 29 patients were followed for 3 months to 14 years. Twenty-three patients made a good recovery (80% of survivors and 54% of the total series), five were moderately disabled, and one was severely disabled.

KEY WORDS - intracranial aneurysm □ subarachnoid hemorrhage □ cerebral vasospasm □ intracerebral hematoma □ hypertension □ epilepsy □ anticonvulsant drugs □ children □ adolescents

Ruptured intracranial aneurysm is a condition most frequently found in adults. It does occur in children, however, and intracranial aneurysms have also been reported in the neonate. A review of the literature reveals that the incidence of aneurysm rupture in patients under the age of 20 years is about 3%, and in patients under the age of 15 years is 1%. The purpose of the present study was to investigate some of the factors presumably involved in the rupture of saccular aneurysm at an early age, and to find factors, if any, of prognostic significance.

Clinical Material and Methods

Throughout the period 1943 to 1980, 1368 patients with verified intracranial saccular aneurysms were treated in the Neurosurgical Department of the University Hospital in Aarhus. Of this total group, 43 patients (3.1%) were 19 years old or younger, and these patients are the subject of this report.

By reviewing the patients' case records, we assessed the clinical findings and the outcome of these patients. Patients in whom traumatic etiology was evident were a priori excluded from the series. The classification system of Hunt and Hess' was used for clinical assessment on admission to the hospital and at the time of angiography.

Angiographic studies were performed in all but four patients who died within the first 24 hours. Depending on the clinical condition, angiography was performed to evaluate the most suspicious area, usually the carotid regions. Pan-angiography was performed in 17 cases (44%). The severity of intracranial spasm was assessed on the basis of the preoperative arteriograms. Slight spasm was defined as a minimal narrowing of the arterial diameter limited to the location immediately around the aneurysm. Spasm was classified as severe either if it was generalized or if there was a pronounced narrowing within an extensive vascular region.

Summary of Cases

Patient Population

The distribution of the sex and age of the 43 patients appears in Fig. 1. There were 25 boys (58%) and 18

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FIG. 1. Sex and age distribution in 43 children with intracranial saccular aneurysms.

Girls (42%). In the pediatric age group (that is, patients 14 years old or younger), there were nine boys and five girls. In one case we found a positive family history of cerebral arterial aneurysm. The patient was an 11-year-old boy with coarctation of the aorta and multiple intracranial aneurysms. His grandmother had died at the age of 56 years because of rupture of an internal carotid artery aneurysm.

Associated Clinical Features

Four children (9%) had hypertension before the present admission, one because of coarctation of the aorta, one with polycystic kidneys, one with nephritis due to diphtheria in early childhood, and one with illness of unknown etiology. Three of these four hypertensive children had multiple aneurysms. Ten patients (23%), three boys and seven girls, suffered from recurrent headache in the years before their aneurysm rupture. In five patients the headache pattern fulfilled the criteria of migraine.5 One patient, an 11-year-old boy with a left internal carotid artery aneurysm, suffered from epileptic seizures.

In addition to the boy with hypertension, we found a second patient with coarctation of the aorta. This patient had been operated on for her condition 7 years earlier, at age 12 years, and showed no signs of hypertension. She had a single intracranial aneurysm. However, only selective angiography of the right carotid region was performed.

Six patients had a history of minor head injury that occurred 2 to 5 weeks before admission to the neurosurgical department. In two patients, minor head injury occurred 10 to 16 months before subarachnoid hemorrhage (SAH). Previous intracranial hemorrhage was suspected in nine patients, and in eight of these it occurred in the 5 weeks preceding rupture. Subarachnoid hemorrhage, proven at lumbar puncture, had occurred previously in one patient. Four-vessel angiography revealed no aneurysm in this patient. Three months later, after a repeat SAH, angiography disclosed a small saccular aneurysm arising from the left internal carotid artery.

None of the 43 patients had any clinical evidence of atherosclerosis, inflammatory angiopathy, subacute bacterial endocarditis, syphilis, blood dyscrasia, or a collagen disorder.

Presentation

Thirty-three (77%) of the patients had an onset of symptoms typical of SAH. Their presentation was characterized by acutely occurring intense headache, stiffness of the neck, nausea, and vomiting. About one-third of the patients had the characteristic symptoms without loss of consciousness, one-quarter had a typical attack with unconsciousness for less than 1 hour, and one-third were unconscious for more than 1 hour.

Among the patients with an atypical course were four patients who had had no signs of SAH. Three of these patients were admitted to the hospital because of oculomotor paresis, and one had developed a left hemiplegia. The circumstances surrounding rupture of the aneurysm were known in 29 patients (Table 1).

Patients were assessed on admission to the hospital according to the grading system of Hunt and Hess14 (Table 2). Twenty-five patients (58%) were in Grades I and II (that is, in good clinical condition).

Angiographic Findings

A total of 51 saccular aneurysms were demonstrated angiographically in the 43 patients. Five patients
Circumstances associated with rupture of the aneurysm in 29 patients

<table>
<thead>
<tr>
<th>Activity During Onset</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>walking, standing, or sitting</td>
<td>11</td>
</tr>
<tr>
<td>running</td>
<td>3</td>
</tr>
<tr>
<td>playing football</td>
<td>3</td>
</tr>
<tr>
<td>common play</td>
<td>2</td>
</tr>
<tr>
<td>asleep or lying in bed</td>
<td>2</td>
</tr>
<tr>
<td>riding a motorbike</td>
<td>2</td>
</tr>
<tr>
<td>swimming</td>
<td>1</td>
</tr>
<tr>
<td>dancing</td>
<td>1</td>
</tr>
<tr>
<td>during coitus</td>
<td>1</td>
</tr>
<tr>
<td>working (lifting a weight)</td>
<td>3</td>
</tr>
</tbody>
</table>

(11%) had multiple aneurysms (Table 3). The locations of the aneurysms are given in Table 4. Neither side had a significant preponderance of aneurysms. Based on the arteriograms and the operative findings, the size of the aneurysm could be assessed in 37 patients with 44 aneurysms. The diameter of the aneurysm was less than 5 mm in 12 cases (27%), between 5 and 10 mm in 22 cases (50%), and larger than 10 mm in 10 cases (23%). Two patients had giant aneurysms, 35 and 50 mm in size.

On the basis of the preoperative arteriograms, the severity of intracranial vasospasm could be assessed in 38 patients. In 13 of these patients, angiography was performed within the first 72 hours after SAH. Spasm was demonstrated in two of the 13 patients. In 19 patients, the angiographic studies were performed after the 3rd day and before the 17th day following SAH. Nine (47%) of these patients revealed no spasm, five patients (26.5%) demonstrated slight spasm, and severe spasm was present in five other patients (26.5%). None of the four patients admitted without any signs of SAH revealed vasospasm. The remaining two patients underwent angiography about 1 month after the acute attack. Angiography demonstrated slight vasospasm in one of them.

The correlation between the patients' clinical condition at the time of angiography and the degree of vasospasm in the group of patients undergoing angiography between the 4th and 16th day after SAH is shown in Table 5. It should be pointed out that none of the patients with vasospasm was in Grades IV or V. Of the two patients with vasospasm demonstrated within the first 72 hours, one had slight spasm and was in Grade V. This patient had a very large intracerebral hematoma and died within 48 hours. The second patient, a 16-year-old girl with classical migraine and severe vasospasm, was in Grade III. One patient demonstrated slight vasospasm at angiography performed 1 month after the acute attack, at which time she was classified in Grade I.

It is remarkable that four of five patients who suffered from migraine developed vasospasm. One of them was a patient with vasospasm demonstrated at angiography within the first 72 hours after SAH. In another, vasospasm was demonstrated about 1 month after the hemorrhage. In the remaining three, angiography was performed on the 5th, 6th, and 8th day, respectively.

Recurrent Hemorrhage

In five patients, rebleeding occurred during the hospital stay, for a rebleeding rate of 13%. Two episodes occurred on the 7th day after SAH, the remain-
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TABLE 5
Relationship between clinical grade at time of angiography and cerebral vasospasm

<table>
<thead>
<tr>
<th>Grade*</th>
<th>No Spasm</th>
<th>Slight Spasm</th>
<th>Severe Spasm</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>II</td>
<td>5</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>III</td>
<td>2</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>IV &amp; V</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

* Grading system of Hunt and Hess.14

ing on the 8th, 13th, and 16th day, respectively. One of these patients had hypertension and four aneurysms, of which two ruptured within a 7-day period.

Intracranial Hematoma

In the 14 patients in the pediatric age group (that is, 14 years old or younger), seven had a hematoma. In the total series of 43 patients, intracerebral hematoma accompanied the aneurysm rupture in 15 patients (37%). The frequency of hematomas was highest (42%) in relation to aneurysms of the middle cerebral artery (MCA) and lowest in relation to aneurysms of the anterior communicating artery (Table 6). Most patients with hematoma were classified in Grades III to V on admission (Table 2).

Mortality

Eight patients (19%) died preoperatively. Six of them presented in Grade V and died within the first 48 hours of admission. Two died after rebleeding on the 8th and 13th days, respectively. The remaining 35 patients were operated on. The interval from SAH to operation and from angiography to operation appears in Table 7. One patient died during an emergency operation. He had a very large intracerebral hematoma and a ruptured aneurysm arising from the MCA.

Five patients died on the 1st through the 6th day postoperatively. Two of them had giant aneurysms (35 mm and 50 mm in size) which bled profusely at surgery. One patient had multiple aneurysms, and another rebled before the operation. They were both in Grade III at the time of operation. In the fifth patient, autopsy demonstrated massive cerebral edema. Thus, the surgical mortality rate was 17%.

The overall mortality rate was 33%. In the pediatric age group, the mortality rate was 50%, presumably due to the high frequency of associated intracerebral hematoma in these children. There was no difference in mortality between patients without angiographic evidence of vasospasm and patients with vasospasm. Three patients with vasospasm died. However, none of them had evidence of cerebral infarction at postmortem examination.

Observations during operation or autopsy revealed that rupture of the aneurysm was accompanied by intracerebral hematoma in 37% of the patients. The mortality rate in this group was 50%, in contrast to 26% in the group without hematoma. In the hematoma group, almost half of the deaths occurred within the first 48 hours.

Of the five patients with multiple aneurysms, three died. None of the four patients with an unruptured aneurysm died.

Long-Term Outcome

The surviving 29 patients were followed for 3 months to 14 years (mean 2.89 years). No patient died during the follow-up period. We used the Glasgow Outcome Scale15 to assess long-term functional capacity. According to this scale, 23 patients (80%) made a good recovery (54% of the total series), five were moderately disabled, and one was severely disabled. Although all patients received preventive anticonvulsant therapy, seven patients (24%) developed epilepsy. Eleven (38%) complained of recurrent headache.

Discussion

The 43 cases of cerebral saccular aneurysms that comprise this series constitute about 3% of the population of patients with intracranial aneurysms treated in our neurosurgical department during the same period of time (1943 to 1980). This incidence and the
male preponderance demonstrated are in accordance with previously reported series.\(^3,10,11,13,14,24,27\) Since intracranial saccular aneurysms in children and adolescents are even less common as an accidental pathological finding in autopsy studies,\(^1,2,21\) they tend to be symptomatic rather than silent in this age group. Therefore, there might be some special factors that increase the risk of rupture of aneurysms during the first two decades of life. Minor head injuries seem to be of importance and, as stated by Amacher and Drake,\(^2\) antecedent head injury may prejudice the initial diagnosis.

A pathogenic role of hypertension in the formation and rupture of cerebral aneurysms has often been claimed,\(^8,9\) but in recent large series no evident association was found,\(^24\) although hypertension appears to be more prevalent in certain subgroups of the total aneurysm population.\(^9\) In the present study, four (9%) of the patients with cerebral aneurysm had hypertension before admission to the hospital. The incidence of hypertension in a "control" population without aneurysms varies considerably, from 1.6% to 12.2%,\(^17,19\) therefore, a relationship between hypertension and aneurysms cannot be determined from the present study. However, it is remarkable that three of the four hypertensive children had multiple aneurysms, suggesting that hemodynamic factors as well as other causes, possibly congenital, might initiate the development of intracranial aneurysms. A possible difference in the pathogenesis between aneurysms arising in children and those arising in adults has been suggested by Lipper, et al.\(^18\) An unusually large congenital medial defect may be the initiating factor in the pathogenesis of the rare aneurysms occurring early in life, whereas, in the adult, degenerative and hemodynamic factors with or without underlying medial defects may be the basis for the increase in frequency of aneurysms with age. There is still no comparative histological study to confirm this suggestion.

It is generally accepted that vascular spasm is a factor of major importance in the overall morbidity and mortality associated with SAH.\(^12,25\) The incidence of radiographic vasospasm following aneurysm rupture ranges from 16% to 66%.\(^12,23\) The variation is due to differences in timing between SAH and the angiographic studies. Angiographic spasm occurring within the first 72 hours after SAH is a very rare phenomenon. Vasospasm is principally demonstrated after the 3rd day and between 1 and 2 weeks after the hemorrhage.\(^25\) In the present study, 19 patients underwent angiography within this period. Spasm was demonstrated in 53% of the cases. However, it was remarkable that none of these patients was categorized in Grade IV or V. It is possible that vasospasm is of minor prognostic significance in children, since there was no increased mortality rate in the group that had vasospasm and no cerebral infarction at postmortem examination, as has been demonstrated in series with adult patients.\(^6,20\)

In 15 patients (37%), the aneurysm rupture was accompanied by intracerebral hematoma. As demonstrated by others, the frequency of hematoma was highest in relation to aneurysms of the MCA. In the Liverpool series of juvenile aneurysms,\(^27\) five out of seven aneurysms arising from the MCA were associated with massive cerebral hematoma, and four of these patients died. Similarly, in the present study, intracerebral hematomas were accompanied by a high risk of mortality: 50% of the patients died in contrast to 26% in the group without hematoma.

The overall mortality rate of 33% in our series is somewhat higher than in the majority of previously published series.\(^1,2,20,24,27\) The high mortality rate partly reflects results obtained before the introduction of microsurgical techniques and before the use of modern intensive care units. Moreover, the differences in mortality between the different series may be explained by variations in the locations of the aneurysms. The mortality seems to be highest among patients with aneurysms of the MCA, the very location where hematoma was most frequently seen. However, comparison among series may be difficult, mainly because of variations in the time of hospital admission following hemorrhage. The earlier the admission after hemorrhage, the worse is the overall management success.\(^7\) In Denmark, the usual policy in cases in which a ruptured aneurysm is suspected is that all patients, irrespective of clinical condition (except for evident moribund cases), should immediately be admitted to the regional neurosurgical department for diagnostic clarification and medical or surgical treatment. In the present study, 30% of patients were in Grade IV or V on admission, and more than 10% of all patients died within the first 24 hours.

The site of aneurysms in the present study is similar to the distribution reported by Rasmussen, et al.,\(^25\) for all 851 patients with symptom-producing intracranial saccular aneurysms admitted to the neurosurgical departments in Denmark in the 5-year period from 1970 to 1974. In that study, about half of the patients had died before the end of the follow-up period (2 to 7 years after SAH), and only half of the surviving patients were fully functional. In the present study, no patient died during the follow-up period, and only one of the 29 surviving patients was severely disabled; 80% made a good recovery. Thus, although aneurysm rupture is serious in children and adolescents, it appears better tolerated at this young age than in comparable groups of adults. We suggest that the reason for the better outcome in youth is related to a lesser degree of neurological deficit due to cerebral vasospasm following the aneurysm rupture.

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