THE FATE OF PATIENTS WHO HAVE CEREBRAL ARTERIOVENOUS ANOMALIES WITHOUT DEFINITIVE SURGICAL TREATMENTS

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INDICATIONS for operation in any given condition must be drawn from the balance remaining between the risks attending surgery, not only to life but to function, on the one hand, as opposed to the risk to life and to function if surgery is denied. This probably is no more true than in the case of arteriovenous anomalies of the brain, in which the operative mortality rate is at least 11.7 per cent, and in which the risk to function is real because of the fact that these lesions so frequently lie in the sylvian and rolandic fissures.

The literature on the fate of these patients for whom no definitive surgical procedure has been carried out is scanty and conflicting. We have therefore reviewed our series of cases from the standpoint of natural history and course of the pathologic process.

Between 1930 and 1954, 51 patients with proved cerebral arteriovenous anomaly have been studied at the Mayo Clinic. The presence of the lesion was verified either by surgical exploration or by cerebral angiography. We rejected, for purposes of this review, a considerable number of cases in which there was strong reason to suspect that an arteriovenous anomaly was present, but in which adequate verification was not obtained. Of this group of 51 patients, 23 received no definitive treatment. The surviving patients of this group have either been re-examined during 1954 or have answered questionnaires regarding their present status and their histories during the interval. In those cases in which the patients’ answers by letter were not deemed satisfactory, the attending physician was consulted and he supplied the necessary information.

The analysis of these cases will be considered from the standpoint of the usual symptoms presented by these patients.

INTRACRANIAL BLEEDING

Olivecrona wrote that “in the end, probably most, if not all patients die of hemorrhage or are completely incapacitated.” He also wrote that almost

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half of his patients had a history of one or more hemorrhages with hemiplegia, which was often transient, although some degree of motor or speech defects usually remained. Dandy\(^2\) wrote that death resulted from cerebral hemorrhages in 40 per cent of these cases. In Mackenzie's\(^4\) series of 50 patients, 15 presented a history of intracranial hemorrhage. Bassett\(^3\) reported on 18 patients, 11 of whom had subarachnoid hemorrhages, some of them with recurrent bleeding, but he thought that the hemorrhages rarely were fatal and that they were not so devastating as those caused by rupture of an aneurysm. Gould and associates\(^5\) encountered subarachnoid bleeding in 15, and intracerebral bleeding in 9, of 41 patients (an incidence of intracranial bleeding of 56 per cent). The mortality rate from hemorrhage was 20 per cent of the entire series.

In our series of 23 patients, 9 (39.1 per cent) have had intracranial bleeding. Seven of these patients have had subarachnoid hemorrhages and 2 had what appeared historically and clinically to have been intracerebral hematomas.

Of the 7 patients who have had subarachnoid bleeding, 2 have died, both in their first attack; one of these patients (Case 13) had had temporal-lobe attacks for 2 years, while the other (Case 19) had had jacksonian convulsions on the average of two times a year for 22 years. The mortality rate from intracranial bleeding in this group of 25 patients is thus 8.7 per cent.

One patient (Case 4) has had three attacks of subarachnoid bleeding, the first one occurring as the first symptom of her disorder in 1932. The last episode of bleeding occurred in 1942. None of these attacks produced crippling symptoms, and at the time of this report she was in good physical and mental health. In 2 patients some degree of partial hemiparesis developed with their attacks of subarachnoid bleeding. One of these (Case 23) 5 years later had only slight hemiparesis and was working full time, as this report was written. The other (Case 22) became moderately hemiparetic at the time of his subarachnoid hemorrhage, and 5 years later wrote that his hemiparesis was no worse and that he was in good health, except for this disability. The other 2 living patients (Cases 5 and 20) both sustained the bleeding in 1941, and had not had subsequent hemorrhages as this was written. Both were working full time and were in good health, except for occasional episodes of convulsions.

Thus, of the 5 patients who survived subarachnoid hemorrhage, 3 have no residual paralytic or speech defects and 2 have some degree of residual hemiparesis. Examination of Table 1, in which are depicted the locations and approximate sizes of these lesions, suggests that in each instance extirpation of the lesion very probably would have been accompanied by rather marked hemiparesis, with speech defects, in addition, in 2 instances (Cases 4, 5, 20, 22 and 23).

Two patients, as judged on the basis of examination and findings, had intracerebral hemorrhages. One of these patients (Case 29) had had headaches for 21 years before examination in 1941. At that time she had impairment of memory and bilaterally choked optic disks. She was last examined
in November, 1954, at which time she was invalidated because of hypertension, pain in the legs and marked obesity. There was no paralysis. Loss of memory was marked. Extirpation of the lesion undoubtedly would have produced some degree of hemiparesis.

The other patient who in all likelihood sustained an intracerebral hemorrhage (Case 2) had had headaches for a year before she was examined. She returned in 1954 with right-sided weakness and severe headaches, with clear cerebrospinal fluid. The hemiparesis cleared gradually. Repeated carotid arteriography demonstrated a shift of the anterior cerebral artery to the right side. This arteriogram also demonstrated an unusual circumstance in that many of the tangled arteries and venous channels were obliterated. She received deep roentgen therapy during the interim between the two arteriograms, but it is doubtful that this treatment resulted in obliteration of two-thirds or more of the abnormal vascular channels in her lesion.

The mortality rate in our series in those cases in which intracranial bleeding occurred was 22.2 per cent; this figure includes one case in which the episode of bleeding was considered to have been a possibility by the attending home physician, but necropsy was not performed.

CONVULSIONS

Convulsive disorder was the main complaint, or one of the main presenting complaints, of 15 of these 23 patients. In 6 of these patients (Cases 4, 5, 11, 20, 35 and 41) the attacks were generalized; in 6 (Cases 19, 26, 28, 30, 44 and 51) they were jacksonian; and in 3 (Cases 1, 13 and 43) they were temporal-lobe attacks.

The condition of all 6 patients with generalized convulsions was satisfactorily controlled by medication, and operation would not be considered for the convulsive disorder alone. Two of these patients had not had convulsions for many years at the time of this report. One of these (Case 4) had convulsions from 1933 to 1936, but had had none since. The other patient (Case 11) had generalized convulsions from 1934 to 1936, preceded by transient attacks of numbness for a year. He had not had any convulsive disorders since 1939.

The patients with jacksonian attacks fared as well as those with generalized convulsions. One patient (Case 19) had two attacks a year, an occurrence that continued until his death 5 years later from subarachnoid bleeding. In another patient (Case 26) jacksonian attacks occurred two times a year for 4 years, at which time he died from a cause unknown to us. The condition of one patient who had occasional focal attacks for 25 years was unchanged. Another patient had focal attacks once a month for 6 years with her menstrual period. Two patients were better. One of these (Case 28) had focal attacks every 2 months for a year, but had had no attacks since 1942, to the time of this writing. Another patient (Case 51) had had minor jacksonian episodes for 9 years, followed by major attacks with loss of consciousness for 2 years, averaging one every 6 months. For the 2 years, at the time of this report, he had had only occasional minor attacks.
Two patients with temporal-lobe attacks died (Cases 1 and 13), one from carcinoma of the lung and the other from subarachnoid bleeding. In these, the attacks were fairly frequent. In Table 1, which indicates the location of the lesion, it is evident that serious neurologic and speech deficits would have attended surgical removal of these lesions. The condition of the third patient with temporal-lobe attacks was well controlled by means of medication. Her lesion is on the medial aspect of the parietal lobe, and is relatively inaccessible.

Thus, it would appear from this survey that convulsions caused by these lesions usually are well controlled by medication, and that in a few instances the attacks disappear, or, at least, long periods of freedom from attacks intervene.

**PROGRESSIVE NEUROLOGIC DEFECT**

Five patients (Cases 22, 23, 27, 30 and 49) at the time of their initial examination presented varying degrees of hemiparesis. In 3 patients, the hemiparesis developed gradually and progressed over a period of years. One patient, who had had mild hemiparesis which developed over a period of 2 months, died after pneumoencephalography. The cerebrospinal fluid just before death was clear. Of the 2 in this group alive at the time of this report, one was invalided in a county home, and the other was not incapacitated.

In 2 patients (Cases 22 and 23) hemiparesis developed at the time of subarachnoid hemorrhage; it persisted in a slowly progressive form in one patient, while the other patient is no worse. Both patients sustained bleeding in 1950, and their condition was followed to January, 1955.

Table 1, in which the approximate size and location of the lesions are depicted, indicates that surgical extirpation would have resulted in hemiparesis in each of these cases.

**HEADACHES**

Five of these patients (Cases 2, 22, 23, 39 and 44) had severe headaches as one of their main presenting complaints. Three of them continued to have headaches as before, at the time of this report. However, in 2 patients headaches ceased; in one for $3\frac{1}{2}$ years, and in the other for 5 years, as this was written. These patients had had severe headaches for 7 and 20 years, respectively.

**MENTAL RETARDATION**

Olivecrona$^6$ wrote that mental deterioration is a common feature of inveterate lesions, and he believed that this observation strengthens the argument for removal of the lesion. Norlén$^7$ also wrote that mental deterioration is common among these patients and is often profound. Gould et al.$^4$ observed this feature in 7 of 41 patients (17.1 per cent). However, Mackenzie$^4$ found mental deterioration to be rare and detected no instance of severe retardation in his 50 patients. Only one patient (Case 39) in our series exhibited signs of mental deterioration. This patient complained of some loss of memory during the 2 years prior to his admission in 1941. On examination, he
TABLE 1
Salient data concerning 23 patients with cerebral arteriovenous anomalies

<table>
<thead>
<tr>
<th>Case</th>
<th>Age*</th>
<th>Symptom, duration, years</th>
<th>Main presenting symptom</th>
<th>Lesion, location</th>
<th>Sub. hem.†</th>
<th>I.C. hem.‡</th>
<th>Condition*</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>40</td>
<td>5</td>
<td>Temporal-lobe attacks</td>
<td>0</td>
<td>0</td>
<td></td>
<td>Temporal-lobe attacks continued as before. Died Jan. 1955. Carcinoma of lung</td>
</tr>
<tr>
<td>2</td>
<td>51</td>
<td>4</td>
<td>Intermittent headaches</td>
<td>0</td>
<td>1x</td>
<td></td>
<td>Intermittent headaches as before. Not incapacitated</td>
</tr>
<tr>
<td>4</td>
<td>44</td>
<td>20</td>
<td>Subarachnoid hemorrhage (1942). Occasional convulsions</td>
<td>3x</td>
<td>0</td>
<td></td>
<td>Last subarachnoid hemorrhage 1942. &quot;Getting along fine.&quot; &quot;No convulsions in years&quot;</td>
</tr>
<tr>
<td>5</td>
<td>31</td>
<td>11</td>
<td>Subarachnoid hemorrhage (1941). Generalized convulsions (6)</td>
<td>1x</td>
<td>0</td>
<td></td>
<td>Generalized convulsions 3 times a year. Working full time</td>
</tr>
<tr>
<td>11</td>
<td>41</td>
<td>22</td>
<td>Episodic transitory numbness, left hand. Generalized convulsions (8); none since 1939</td>
<td>0</td>
<td>0</td>
<td></td>
<td>No convulsions since 1939. Occasional numbness, left side. Working. Slight left hemiparesis</td>
</tr>
<tr>
<td>13</td>
<td>31</td>
<td>2</td>
<td>Temporal-lobe attacks every 3 weeks</td>
<td>1</td>
<td></td>
<td></td>
<td>Died in first subarachnoid hemorrhage 2 years after onset of symptoms</td>
</tr>
<tr>
<td>19</td>
<td>62</td>
<td>22</td>
<td>Jacksonian convulsions each year</td>
<td>1</td>
<td>0</td>
<td></td>
<td>Convulsions continued 2 times a year. Died in first subarachnoid hemorrhage 1947</td>
</tr>
<tr>
<td>22</td>
<td>26</td>
<td>10</td>
<td>Headaches. Subarachnoid hemorrhage (June 1950). Hemiparesis</td>
<td>1</td>
<td>0</td>
<td></td>
<td>&quot;Paraparetic&quot; No headaches past 5 years. &quot;Enjoying fairly good health&quot;</td>
</tr>
<tr>
<td>23</td>
<td>42</td>
<td>5</td>
<td>Headaches each month since 1930. Subarachnoid hemorrhage (Feb. 1950); slight hemiparesis</td>
<td>1</td>
<td>0</td>
<td></td>
<td>Slight weakness right side. Working full time</td>
</tr>
<tr>
<td>26</td>
<td>39</td>
<td>9</td>
<td>Transient numbness, left hand. Jacksonian convulsions each 2 months</td>
<td>0</td>
<td>0</td>
<td></td>
<td>Convulsions each 2 months. Died June 1941</td>
</tr>
<tr>
<td>27</td>
<td>53</td>
<td>26</td>
<td>Progressive hemiparesis</td>
<td>0</td>
<td>0</td>
<td></td>
<td>Hemiparetic, otherwise no complaints. Invalid</td>
</tr>
<tr>
<td>28</td>
<td>28</td>
<td>16</td>
<td>Jacksonian convulsions</td>
<td>0</td>
<td>0</td>
<td></td>
<td>No convulsions since 1942. Working full time</td>
</tr>
<tr>
<td>30</td>
<td>28</td>
<td>23</td>
<td>Progressive hemiparesis; jacksonian convulsions</td>
<td>0</td>
<td>0</td>
<td></td>
<td>Hemiparetic, occasional convolution. Not incapacitated</td>
</tr>
<tr>
<td>35</td>
<td>33</td>
<td>9</td>
<td>Generalized convulsions each 2 months</td>
<td>0</td>
<td>0</td>
<td></td>
<td>Convulsive disorder satisfactorily controlled by medication</td>
</tr>
<tr>
<td>39</td>
<td>54</td>
<td>33</td>
<td>Headaches</td>
<td>0</td>
<td>Prob.</td>
<td></td>
<td>Headaches each month, memory loss. Mental deterioration</td>
</tr>
<tr>
<td>40</td>
<td>41</td>
<td>7</td>
<td>Episodic tinnitus</td>
<td>0</td>
<td>0</td>
<td></td>
<td>Tinnitus as before. Not incapacitating. No worse</td>
</tr>
<tr>
<td>41</td>
<td>51</td>
<td>3</td>
<td>&quot;Temporal-lobe attacks&quot;</td>
<td>0</td>
<td>0</td>
<td></td>
<td>No convulsions past 2 years. Taking anticonvulsant medication</td>
</tr>
</tbody>
</table>

* At time of this report.
† Subarachnoid hemorrhage.
‡ Intracranial hemorrhage.
was found to have choked optic disks, and a blood pressure of 170 systolic and 110 diastolic. He was seen again in 1954, at which time his blood pressure was 300 systolic. Loss of memory had increased, and there were other evidences of progressive mental deterioration.

**SUMMARY**

The fate of 23 patients who had arteriovenous anomalies and for whom no definitive neurosurgical procedures were carried out has been reviewed. Because the series is small, the mortality rate and other data may not be entirely valid. However, the condition of some of these patients has been followed since the 1980’s and provides some indication of the fate of the patients.

The group mortality rate from intracranial bleeding was 8.7 per cent. Only one patient with intracranial bleeding was invalided because of this complication. Two other patients had residual hemiparesis, but continued to work. When it is considered that the mortality rate of radical extirpation of such lesions is at least 11.7 per cent, that excision cannot always be complete, and that because of the location of these lesions residual neurologic defects have to be expected in most cases, this study suggests that intracranial bleeding in itself is a dubious indication for operation.

Analysis of the convulsive disorders presented by these patients indicates that in the majority of patients such afflications can be satisfactorily controlled medically, and that in a significant number of cases convulsions disappear. When, again, the risk of surgery and the fact that surgical extirpation is no guarantee that convulsions will disappear are considered, it is the opinion of the authors that on the basis of the convulsive disorder alone, operation is not justified for many of these patients.

Progressive hemiparesis developed in 13 per cent of patients in this series. This figure corresponds to the mortality rate of surgical extirpation; in
most cases the neurologic defect produced by surgery would be almost as profound as that obtaining had the lesion not been treated surgically.

Mental deterioration was present in one of these 28 patients. Our experience in this regard is similar to that of Mackenzie, and is contrary to that of Olivecrona and Norlén.

It is our opinion, derived from this review, that headache alone rarely constitutes an indication for surgical intervention.

REFERENCES