Vascular Malformations ("Angiomas") of the Brain, with Special Reference to Those Occurring in the Posterior Fossa

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ALTHOUGH vascular malformations ("angiomas") within the posterior fossa have been considered uncommon, or even rare,3,4,12,15,26,27,28,31,34,40 one of us has recently shown that such is not the case.19 This report gives our experience with certain major pathological features of 164 angiomas of the posterior fossa. In addition, the major pathological and demographical features of 330 patients with 346 cerebral angiomas and two dural angiomas are compared. Many of these cases had detailed gross examinations, and all were studied microscopically. These malformations were characterized according to the detailed morphological classification recently published by one of us.21 The dural angiomas have been reported separately.22

Materials

The 164 posterior fossa angiomas were obtained from 37 patients from the autopsy services of the University of Tennessee and the University of Iowa, and from 120 patients from the files of the Armed Forces Institute of Pathology (AFIP). Most of the angiomas from the Universities of Tennessee and Iowa were examined and photographed as specimens, while only a few of the cases from the AFIP were examined grossly by the authors. Hematoxylin and eosin stained sections were used on all cases, and elastic and connective tissue stains were used on many. At times other special stains (for glia, calcium, etc.) were also used. Equivocal cases (those with insufficient tissue to assure the diagnosis) and cases of Sturge-Weber or Lindau's disease have been excluded. The location and type of malformations have been tabulated. The frequency of thrombosis within the angiomas and the frequency and quantity of hemorrhage, calcium, and bone have also been recorded. The lack of clinical data on some of the AFIP cases precluded accurate tabulation of the clinical findings.

The 346 cerebral angiomas (306 from the AFIP and 40 from the Universities of Iowa and Tennessee) were examined in the same manner. Although the cerebral cases are not discussed in detail, they provide a basis for comparing certain morphological and demographical features with those in the posterior fossa malformations.

Results

Tables 1–5 summarize the location, age, morphological type, and major clinical features of the posterior fossa angiomas. The sex ratio derived from our material is probably of little value, as many of the cases were from the predominantly male armed services. There were 26 males and 11 females among the 37 patients with posterior fossa angiomas from the Universities of Iowa and Tennessee.

Tables 3 and 5 summarize the age data available on our patients with angiomas. There is no significant difference between the average age of patients with supra- and infratentorial arteriovenous and venous angiomas and only a questionable difference for the cavernous angiomas. However, there was a significant age difference between the two telangiectasia groups.

Angiomas of the cerebellum were more numerous than those in the brain stem (91 to 69). This difference is probably related to the relative size difference of these two structures.

Of the 164 "angiomas" of the posterior fossa, 70 were A-V malformations, 38 telangiectasia, 31 venous angiomas, 21 cavernous angiomas, and 4 varices. The most common
TABLE 1

Location of posterior fossa angiomas in 157 patients

<table>
<thead>
<tr>
<th>Location</th>
<th>Cerebellum (87 patients)</th>
<th>Brain Stem (66 patients)</th>
<th>Involvement of both Cerebellum &amp; Brain Stem</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right Lobe</td>
<td>Left Lobe</td>
<td>Vermis</td>
<td>Mesencephalon</td>
</tr>
<tr>
<td>23</td>
<td>27</td>
<td>12</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Location Unknown*</td>
<td></td>
</tr>
<tr>
<td>25</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Due to incomplete documentation (photographic or written), the exact location within the cerebellum could not be determined.

TABLE 2

Morphological type of 164 angiomas by location*

<table>
<thead>
<tr>
<th>Type</th>
<th>Location</th>
<th>Cerebellum (91)</th>
<th>Extensive in Brain Stem (6)</th>
<th>Mesencephalon (6)</th>
<th>Pons (48)</th>
<th>Medulla (9)</th>
<th>Cerebellum &amp; Brain Stem (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arteriovenous</td>
<td></td>
<td>52</td>
<td>5</td>
<td>2</td>
<td>5</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Telangiectasis</td>
<td></td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>27</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Cavernous</td>
<td></td>
<td>11</td>
<td>0</td>
<td>1</td>
<td>7</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Venous</td>
<td></td>
<td>20</td>
<td>1</td>
<td>3</td>
<td>7</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Varix</td>
<td></td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

* There were more angiomas (164) than patients (157) as 6 patients had multiple separate angiomas in the posterior fossa.

TABLE 3

Age of 148 patients with posterior fossa angioma*

<table>
<thead>
<tr>
<th>Patient Decade</th>
<th>Cerebellum (81)</th>
<th>Extensive in Brain Stem (6)</th>
<th>Mesencephalon (5)</th>
<th>Pons (45)</th>
<th>Medulla (8)</th>
<th>Cerebellum &amp; Brain Stem (3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-9</td>
<td>3</td>
<td>1</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>10-19</td>
<td>8</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>20-29</td>
<td>20</td>
<td>2</td>
<td>0</td>
<td>4</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>30-39</td>
<td>15</td>
<td>0</td>
<td>0</td>
<td>9</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>40-49</td>
<td>8</td>
<td>2</td>
<td>0</td>
<td>5</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>50-59</td>
<td>14</td>
<td>0</td>
<td>2</td>
<td>10</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>60-69</td>
<td>6</td>
<td>0</td>
<td>1</td>
<td>9</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>70-79</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>6</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>80-89</td>
<td>3</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

* The average age of 148 patients with posterior fossa angioma was 43.6 years. The average age of 81 patients with cerebellar angiomas was 40 years. The average age of 64 patients with brain stem angiomas was 49 years.
type of malformation in the cerebellum was arteriovenous (A-V), while in the brain stem telangiectasis predominated, due primarily to their frequency in the basis pontis. If these typically small (Fig. 1) asymptomatic angiomas are excluded, the A-V malformations predominate. Among the 346 cerebral angiomas, 217 were A-V, 59 cavernous, 46 venous, 22 telangiectasia, and 2 varices.

The majority of the posterior fossa angiomas for which measurements are available (95 cases) measured less than 2 cm in greatest extent and fulfill the essential criteria for "cryptic" angiomas\(^2\) (Figs. 1–5). The size of

\[\text{Average age of patients with angioma by location and type}\]

<table>
<thead>
<tr>
<th>Type of Angioma</th>
<th>Location and Average Age (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Supratentorial</td>
</tr>
<tr>
<td>Arteriovenous</td>
<td>32</td>
</tr>
<tr>
<td>Venous</td>
<td>48</td>
</tr>
<tr>
<td>cavernous</td>
<td>50</td>
</tr>
<tr>
<td>Telangiectasia</td>
<td>44</td>
</tr>
</tbody>
</table>

*The number of patients with angiomas listed here differs from Table 1 because no clinical data are available for 8 patients; 1 of the 4 patients with an angioma involving both brain stem and cerebellum had signs & symptoms of long duration; the other 3 were symptom free.

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**TABLE 4**

**Clinical course of posterior fossa angiomas in 149 patients**

<table>
<thead>
<tr>
<th>Location of Angioma*</th>
<th>Clinical Course</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Sudden Unexpected Death (within 4 hrs)</td>
</tr>
<tr>
<td></td>
<td>Rapid Death with Signs &amp; Symptoms of Short Duration (4 hrs to 1 wk)</td>
</tr>
<tr>
<td></td>
<td>Signs &amp; Symptoms of Long Duration With or Without Hemorrhage</td>
</tr>
<tr>
<td></td>
<td>No Signs &amp; Symptoms</td>
</tr>
<tr>
<td>Cerebellum (80 patients)</td>
<td>16</td>
</tr>
<tr>
<td>Brain Stem (65 patients)</td>
<td>4</td>
</tr>
</tbody>
</table>

\(\text{Fig. 1. Left: Telangiectasia of the basis pontis. These lesions are typically small and clinically silent. } \times 1^{\frac{1}{4}}\)

\(\text{Right: This } 3 \text{ mm soft, reddish-brown area in medulla is a typical telangiectasia, similar to the more common pontine one at the left. } \times 2^{\frac{1}{4}}\)
found in the basis pontis. Of 38 such angiomas, 27 were pontine. None of these 27 cases was associated with significant hemorrhage. Fatal hemorrhage was observed in only one cerebral (diencephalic) telangiectasia.

Calcification was present in 12 of the 164 posterior fossa angiomas (7 in cerebellum and 5 in brain stem), and bone formation was found in 2 (both cerebellar). The calcification was heavy in only 3 of the cases. The calcium deposition was within the walls of the vessels of the angiomas and occasionally in the surrounding gliotic parenchyma, and occurred in 7 A-V, 4 cavernous, and 1 venous angioma. Bone formation (Fig. 7) was found in one A-V and one cavernous angioma, both in the cerebellum. In contrast, bone formation was seen in 7 of 346 cerebral (supratentorial) angiomas and calcification in 64. This calcification was very heavy in 10 of the cerebral angiomas, and moderate in another 21.

An occasional thrombosed vessel was found in a number of the angiomas, but extensive thrombosis was found in only 3 of the cerebral angiomas and in only one of the posterior fossa angiomas (Figs. 7–9).

Two of the posterior fossa angiomas, both pontine cavernomas, were surrounded by relatively extensive Schwannian and fibrous gliosis (Fig. 10). Both of these patients had multiple angiomata, with cerebral as well as brain stem angiomas. Although this reaction may possibly represent part of a complex hamartoma rather than a pure angioma ("simple hamartoma"?), we have interpreted such stromal alterations as an unusual reaction to injury. Their associated cerebral angiomas did not contain this unusual stromal reaction.

Hydrocephalus, of varying severity, was found in 10 of the 157 patients with posterior fossa angiomas. The angioma was located in the cerebellum in 7 of these patients, in both cerebellum and brain stem in 1, and in the brain stem alone in 2. These patients ranged in age from a newborn infant with a mesencephalic and pontine A-V malformation to a 63-year-old man with a medullary A-V malformation. Mechanical obstruction of the aqueduct and/or fourth ventricle was demonstrated in 7 patients with hydrocephalus. Three patients had significant ventricular dilatation without obvious mechanical obstruction to the CSF pathways.
Fig. 3. Clinically silent cavernous angioma of the pons. These lesions usually resemble petechiae more closely than the telangiectasias. ×3.

Fig. 4. Clinically silent venous angioma of cerebellum.
Sixty-three of our 149 patients with posterior fossa angiomas for whom the ages are known were over 50 years of age, and the average age for all of these patients was nearly 44 years. No important difference between the ages of the patients with supratentorial angiomas and those with infratentorial angiomas was apparent in our series (Table 5). The frequency of small, silent ("cryptic") angiomas in the elderly has been discussed by White, et al., and by McCormick and Nofzinger.

Hydrocephalus has been observed in a significant number of patients with posterior fossa angiomas. In most of the reported cases, the hydrocephalus has been obstructive, but significant ventricular dilatation has occurred in the absence of demonstrable obstruction. Both obviously obstructive and apparently non-obstructive types of hydrocephalus were recorded in 10 (6.4%) of the 157 patients with posterior fossa angiomas in our series. Askenasy, et al., have discussed possible mechanisms by which non-obstructive angiomas (including supratentorial ones) may give rise to internal hydrocephalus, and have reviewed some of the literature on this subject. They seem to favor the hypothesis that the non-obstructive forms seen in some patients are due to atrophy of the brain (with ex vacuo hydrocephalus) secondary to "faulty distribution of blood, and subsequently oxygen, to the brain." One of our three "non-obstructive" hydrocephalics had never bled, while the other two had. The absence of a history of morphologic evidence of prior hemorrhage in some cases was also mentioned by Askenasy, et al. We believe that it can be fairly stated that the cause of the ventricular dilatation seen in some angioma patients is still unknown.

Some authors have questioned either the existence or importance of angiomas other than arteriovenous malformations. Such misleading statements as "...we see little merit in attempting to distinguish, as others have done in the past, between arterial, venous or arteriovenous malformations, because arterial and venous channels are common to all these lesions. We believe they should all be classified simply as arteriovenous malformations," and "whether the predominant vessel is arterial, venous or capillary is essentially an academic question for in most cases all

Discussion

Vascular malformations ("angiomas") of the posterior fossa have now been recorded in well over 200 patients. While many recent works cite Verbiest's figure of 108 infratentorial angiomas reported up to 1961, this figure omits the 22 cerebellar and brain stem angiomas reported by White, et al. The most recent figure of 137 cases said to have been reported to 1966 also failed to include many recent reports. A recent review of only the small ("cryptic") angiomas tabulated 147 located in the posterior fossa to which others have now been added. While these varying reports point up the great difficulty of making a comprehensive review of the literature they do indicate that angiomas of the cerebellum and brain stem are not rare.

While posterior fossa angiomas have been found in premature newborns and in patients over 80, many of the patients have been between the ages of 10 and 40. Thus, the commonly held notion that most angiomas are found in young persons does not seem to be on statistically firm ground.
Fig. 6. Fatal hemorrhage into a pontine A-V malformation.

Fig. 7. Bone formation (osseous metaplasia) in cavernous angioma. There are several old thrombosed vessels within this angioma. H. & E., ×12.
FIG. 8. Thrombosed, recanalized vessel in an arteriovenous malformation from the cerebrum. This type of thrombosis is relatively uncommon and was extensive in only a few cases. H. & E., ×50.

types are present," can, if taken seriously, hinder further understanding of nervous system angiomas. As mentioned previously, we have seen fatal hemorrhages from all known types of angiomas and under conditions where adequate tissue examination has proved that they were not all A-V malformations. This is especially true for the angiomas of the posterior fossa, which are often small and a few of which have been studied by us with subserial sections. Tables 2 and 4 summarize our data on this question.

There have been many clinical studies of angiomas but relatively few careful published studies of their morphology. We know of only one ultrastructural (electronmicroscopic) study of angiomas. The relative proportions of the various types (A-V, cavernous, venous, etc.) reported seem to vary widely. For example, cavernous angiomas have been called

Fig. 9. Extensively thrombosed arteriovenous angioma in right caudate nucleus.
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FIG. 10. Photomicrographs showing extensive gliosis associated with cryptic pontine angioma. This type of gliosis was encountered only twice and supports the hamartomatous concept of these angiomas. In the right photograph the cavernous component is readily apparent. Left: P.T.A.H. stain, ×20. Right: Trichrome stain, ×20.

“extremely rare,” yet we have examined no less than 21 infratentorial and 20 supratentorial angiomas of this type. Similar statements have been made concerning venous angiomas, especially those that have bled. Again, our experience with 52 venous angiomas is not in agreement.

There does appear to be at least a rough correlation between the size (extent) of an angioma, regardless of morphological type or location, and the frequency of hemorrhage. In our material, the A-V angiomas have made up the majority of the larger malformations (over 3 cm in size), and have been the type that have most often bled. Unfortunately, our data do not allow us to make an exact quantification of this aspect of an angioma’s natural history.

Calcification and ossification in angiomas of the central nervous system have been mentioned or illustrated on a number of occasions in the literature. Sabra reported its occurrence in 20 of 100 supratentorial angiomas. It seems, judging from these reports, that large amounts of calcification are unusual, yet the rare heavily calcified angiomas cited by Penfield and Ward and Shafey, et al., are apparently exceptions. Similarly, ossification has been uncommonly reported. As previously noted, three of our posterior fossa angiomas had heavy calcification and two others contained bone. Calcification was found in 62 of our 346 supratentorial angiomas, and was abundant in 10. Ossification was present in 7 of the cerebral angiomas.

Amyloid, or amyloid-like material, has been described in the walls of angiomas. This phenomenon was observed in three of our A-V angiomas. Two cerebral and one cerebellar angioma were so involved. Our patients did not have systemic amyloidosis, and we do not know what significance this finding has, if any.

The coexistence of an angioma and a saccu-
lar aneurysm in the same patient is well documented.\textsuperscript{29,31} The cooperative subarachnoid hemorrhage study cites this coincidence of lesions in 37 of 490 patients with angiomas. This coincidence occurred 9 times in our angioma patients, 7 with supratentorial and 2 with infratentorial angiomas. Our figures are almost certainly too low to reflect the true prevalence of this coincidence due to the manner in which most of our cases were obtained. Perhaps a more accurate estimate of the frequency with which these two lesions occur in the same patient can be deduced from the fact that we have found 6 angiomas in dissections of the last 100 consecutive brains with saccular aneurysms.

Summary

We have reviewed 164 vascular malformations of the posterior fossa and presented gross and microscopic photographs of the typical varieties.

We have compared these data with material derived from a series of 346 supratentorial vascular malformations and found that arteriovenous malformations were the most common type in each group, but they occurred with greater frequency supratentorially. Telangiectasia was usually found incidentally and was most common in the posterior fossa, particularly the basis pontis. The other types of vascular malformations occurred with nearly equal frequency in both groups. Although infrequent, calcification and/or ossification occurred more often in the supratentorial lesions. Hydrocephalus occurred in 6.4\% of posterior fossa malformations.

Although bleeding was most frequent with arteriovenous malformations, all types of vascular malformations were associated with fatal hemorrhage, a fact which we believe casts doubt on the belief that all vascular malformations are of the arteriovenous type.

Acknowledgments

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