THE CEREBELLAR ANGIORETICULOMAS

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The angiomatous tumors occurring in the cerebellum have been variously designated as angiopticulomas and angioblastomas, and are also commonly referred to as Lindau tumors, since Lindau's important contribution in 1926 brought the attention of neurosurgeons to this, in the adult, fairly common type of cerebellar tumor. Since the term angiopticuloma appears to be best adapted to the histological appearance of these tumors, I have in accordance with Bergstrand used this designation.

Although arteriography has brought out a surprising number of arteriovenous aneurysms, it is nevertheless true that angiomatous malformations and tumors of the brain are comparatively rare lesions. In the earlier statistics of such as Cushing and Bailey (1928) and Cushing (1932), blood-vessel tumors, in which group are included the true tumors as well as the angiomatous malformations, made up about 2 per cent of the verified brain tumors. In the statistics of this clinic up to Dec. 31, 1934, the corresponding figure was 3.5 per cent, the larger figure in this clinic probably being due to the extensive use of arteriography. In Table 1 are assembled the verified brain tumors observed in this clinic up to Dec. 31, 1950. As may be seen, the angiopticulomas are somewhat less numerous than the arteriovenous aneurysms and make up approximately 2 per cent of our verified tumors. This may seem to be an insignificant number, but when it is recalled that the angiopticulomas occur only in the cerebellum of adults, it becomes

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Count</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Glioma</td>
<td>2,008</td>
<td>47.9%</td>
</tr>
<tr>
<td>Meningioma</td>
<td>806</td>
<td>19.2%</td>
</tr>
<tr>
<td>Neurinoma</td>
<td>356</td>
<td>8.6%</td>
</tr>
<tr>
<td>Adenoma</td>
<td>368</td>
<td>8.7%</td>
</tr>
<tr>
<td>Angioma</td>
<td>111</td>
<td>2.6%</td>
</tr>
<tr>
<td>Arteriovenous aneurysm</td>
<td>104</td>
<td>2.5%</td>
</tr>
<tr>
<td>Aneurysm</td>
<td>77</td>
<td>1.9%</td>
</tr>
<tr>
<td>Craniopharyngioma</td>
<td>74</td>
<td>1.9%</td>
</tr>
<tr>
<td>Cholesteatoma</td>
<td>31</td>
<td>0.9%</td>
</tr>
<tr>
<td>Teratoma, chordoma</td>
<td>10</td>
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</tr>
<tr>
<td>Papilloma</td>
<td>17</td>
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</tr>
<tr>
<td>Granuloma</td>
<td>55</td>
<td>1.3%</td>
</tr>
<tr>
<td>Metastatic</td>
<td>147</td>
<td>3.3%</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>24</td>
<td>0.6%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>4,188</td>
<td></td>
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</table>

In Table 1 the verified intracranial tumors up to Dec. 31, 1950 are classified.
obvious that they form an important group of cerebellar tumors. In Table 2 are assembled the 958 verified posterior fossa tumors observed in this clinic up to Dec. 31, 1950. The angioreticulomas comprise 7.3 per cent of the total number of posterior fossa tumors.

TABLE 2
Age incidence of posterior fossa tumors
1922-Dec. 31, 1950

<table>
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<tr>
<th></th>
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<tbody>
<tr>
<td>Gliomas (408)</td>
<td>175</td>
<td>108</td>
<td>57</td>
<td>34</td>
<td>21</td>
<td>10</td>
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<tr>
<td>Acoustic neurinomas (349)</td>
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<td></td>
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<td></td>
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<tr>
<td>Meningiomas (75)</td>
<td></td>
<td>6</td>
<td>52</td>
<td>81</td>
<td>89</td>
<td>89</td>
<td>31</td>
<td>1</td>
</tr>
<tr>
<td>Angioreticulomas (70)</td>
<td></td>
<td>0</td>
<td>2</td>
<td>5</td>
<td>18</td>
<td>26</td>
<td>19</td>
<td>4</td>
</tr>
<tr>
<td>Tuberculomas (24)</td>
<td></td>
<td>0</td>
<td>6</td>
<td>13</td>
<td>14</td>
<td>14</td>
<td>20</td>
<td>3</td>
</tr>
<tr>
<td>Metastatic (16)</td>
<td></td>
<td>2</td>
<td>6</td>
<td>11</td>
<td>3</td>
<td>0</td>
<td>0</td>
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<tr>
<td>Papillomas, cholesteatomas, chor-</td>
<td></td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>7</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>domas, teratomas (22)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tbody>
</table>

PATHOLOGICAL ANATOMY

From the now classical descriptions of Cushing and Bailey,3 Lindau5 and others, the histological appearance of these tumors is well known and need not detain us here. The macroscopical anatomy of these tumors as they appear at the operating table is as a rule sufficiently characteristic to allow a correct diagnosis as soon as the lesion is seen. The tendency to cyst formation, so typical of these tumors, has been emphasized by all. Tumors unaccompanied by cysts are by no means rare and sometimes the cysts are quite small although they are generally of a size many times the solid tumor nodule. Lindau5 found 2 entirely solid tumors among his 16 cases, and in the material of Cushing and Bailey3 cyst formation was lacking in no less than 4 out of 11 cases. In our material of 70 cases there were 55 cystic and 15 solid tumors, the solid tumors thus comprising 18.4 per cent of the entire series. If to this group are added the cases in which the cyst was quite small, we find that in between one fourth to one third of the cases, cyst formation was lacking or was so small as to be without significance. It would seem, therefore, that the frequency of cyst formation has been somewhat overemphasized in the earlier literature.

The mural nodule in the cystic tumor may occasionally, as pointed out by Cushing and Bailey,3 be quite small, so small in fact that it can be detected only by microscopical examination of the cyst wall, but as a rule it is easily recognized and varies in size between that of a pea and a walnut. In one of the rare cases where a true recurrence was thought to have occurred, the originally cystic tumor was later replaced by a large solid mass, occupying the entire cerebellar hemisphere of one side.

Since these tumors in all probability take their origin from a meningo-
cerebral malformation, the mural node in the cystic tumors and the solid tumors are always located close to the surface of the cerebellum or the medulla and it will be found that they are always at some point in direct contact with the pia-arachnoid. They are therefore in most instances easily detected at operation, even when unaccompanied by a cyst. The exceptions are those cases in which no cyst is present and the solid tumor is located far anteriorly, close to the superior surface of the cerebellum, which may not always be accessible to inspection. Even in these cases the vascular pattern seen on the surface of the cerebellum usually gives a clue to the presence and location of the tumor and it is exceedingly rare that the tumor is not found at operation. The increased vascularity of the cerebellar cortex near the tumor is very characteristic and almost pathognomonic. One or several branches of cortical arteries are greatly enlarged and converge towards the tumor and the draining veins are several times the size of normal cerebellar veins and often contain bright red arterial blood. When the solid tumor is large, the vascular supply may be so abundant that the lesion becomes almost aneurysmatic in appearance. In angiograms of the vertebral artery the vascularity of these tumors can be beautifully demonstrated (Fig. 1).

In our material the tumor was localized to the cerebellar hemispheres in 56 cases, to the vermis in 9 cases and to the region of the fourth ventricle.

Fig. 1. Vertebral angiogram in a case of right-sided cerebellar angioreticuloma. The tumor was largely solid, although accompanied by a small cyst.
in 5 cases. In the last-mentioned group the tumor usually arises from the velum medullare posterius, which separates the tumor from the floor of the fourth ventricle. This situation obtained in 1 of our cases reported in our monograph with Bergstrand and Tönns in 1936. In 2 other cases the tumor had its point of attachment to the medulla in the region of the velum medullare posterius, but the exact relations of the tumor to the velum could not be determined. In 1 case the tumor was located to the uppermost part of the fourth ventricle, growing in a ring-like fashion around the lowest part of the aqueduct, and apparently attached to the floor of the fourth ventricle at its cranial end. In the fifth case there were 2 tumors, one probably arising from the velum medullare posterius but rather firmly attached to the medulla in the region of the calamus scriptorius, and the other attached to the lateral margin of the fourth ventricle, just above the striae acusticae. The opinion generally expressed in the literature that the angioreticulomas located to the region of the fourth ventricle always arise from the velum medullare posterius therefore no longer can be sustained.

HEREDITY—RELATION TO V. HIPPEL’S ANGIOMATOSIS RETINAE—MULTIPLE TUMORS

Familial occurrence of angioreticulomas has been recorded in the literature on several occasions. When associated with angiomatosis retinae, cysts of the pancreas and tumors of the kidney, the condition generally goes under the name of Lindau’s disease. Heredity is frequently, though not invariably, observed in this condition.

On the whole, the combination of angiomatosis retinae with a cerebellar angioreticuloma is very rare and the hope once expressed by Cushing and Bailey that this combination might in the future be of considerable diagnostic importance has not been fulfilled. In the present series angiomatosis retinae was observed only once among 70 cases. Perlmutter, Horrax and Poppen recorded the unusually high frequency of 16 per cent of angiomatosis retinae among their 25 cases.

In this clinic 3 families have been observed in which 2 or more members of the family were afflicted with angioreticulomas. Norlén made one of these families the subject of a short report in 1941. The pedigree of this family is reproduced from Norlén’s paper.

<table>
<thead>
<tr>
<th></th>
<th>No. 1</th>
<th>No. 2</th>
<th>No. 3</th>
<th>No. 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. 1</td>
<td>9, 44 years old, from brain tumor</td>
<td>9, 25 years old, Living, well.</td>
<td>9, 25 years old, Living, well.</td>
<td>9, 25 years old, Living, well.</td>
</tr>
<tr>
<td>No. 2</td>
<td>9, 37 years old, Multiple cerebellar angioreticulomas removed 1937, 1940, 1941.</td>
<td>9, 25 years old, Died, cerebellar angioreticuloma.</td>
<td>9, 18 years old, Living, well.</td>
<td>9, 10 years old, Living, well.</td>
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<tr>
<td>No. 3</td>
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<tr>
<td>No. 4</td>
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</tr>
</tbody>
</table>

9 Died, 44 years old, from brain tumor

9, 37 years old. Multiple cerebellar angioreticulomas removed 1937, 1940, 1941.

9, 25 years old. Died, cerebellar angioreticuloma.

9, 25 years old. Living, well. 9, 25 years old. Living, well.

9, Died at birth. 9, Died at birth. 9, Died at birth. 9, Died at birth.
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In the second family the patient was a man aged 34 at the time of his first admission. His mother had died at the age of 56 from hypernephroma with multiple metastases to the lungs. She had been suffering for some months from a cerebellar tumor which was thought to be metastatic. At autopsy, however, it was found that the cerebellar tumor was an angio-reticuloma situated in the roof of the fourth ventricle. The patient himself had three different angio-reticulomas in the cerebellum, two of which were removed at the first operation in 1947 and the third at a second operation 2 years later. Two of the tumors were situated in the fourth ventricle and one in the right hemisphere. His present condition indicates that he may have still another angioma in the cerebellum.

In the third family, the patient was a man aged 46. His mother became blind at the age of 23. One brother died from brain tumor, and another brother was operated on for brain tumor, diagnosis unverified. One sister has blindness in the left eye, caused by angiomatosis retinae. The patient himself has bilateral angiomatosis retinae, which was first diagnosed in 1941 and which led to blindness in both eyes. Signs of intracranial hypertension began late in 1949 and he was operated on in November 1950, when a large cystic angioma was removed from the left cerebellar hemisphere.

It was characteristic in all 3 cases of familial occurrence that multiple tumors were present, although usually only one tumor was manifest at a time and years elapsed before symptoms of intracranial disease again became apparent. Obviously it is well to keep patients with a hereditary tendency under close observation, as new tumors in other parts of the cerebellum are very likely to occur. The last-mentioned case was the only one in the present series in which angiomatosis retinae was found.

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SEX AND AGE INCIDENCE

There were 41 males and 29 females in our material, indicating a slight preponderance for the male sex. The age distribution presented in Fig. 2 suggests that the tumors tend to manifest themselves at an earlier age in the female than in the male sex. In the female, the greatest incidence occurs.
in the age group 20–40, while in the male the maximum frequency occurs
two decades later. No case occurred before puberty and only 8.7 per cent of
the patients were below 20 years of age when the first symptoms were
noted. The age distribution is not without diagnostic significance as the
vast majority of cerebellar gliomas occur before the age of 20 and most of
these before puberty.

**SYMPTOMATOLOGY**

The first symptom nearly always is headache, which was recorded as
the initial symptom in 50 cases or in 84 per cent of the entire material. In 42
cases vertigo appeared almost simultaneously with headache or very
shortly afterwards. Vomiting occurred very early in 29 cases.

The headache nearly always is of the type associated with intracranial
hypertension but occasionally may occur in a more unusual form as in one
of our cases, in which the first symptom was a sharp pain in the occipital
region, provoked by stooping or the lifting of heavy burdens. Vertigo may
occur in attacks resembling Ménière’s disease but is more often described
as periodically recurring sensations of giddiness, frequently associated with
bouts of increased headache. In some instances, vertigo is so intense that
disturbance of gait and station occurs permanently at an early stage or
as one of the initial symptoms. Although by no means pathognomonic,
the combination of headache, vertigo and vomiting occurring as the earliest
symptoms is a fairly reliable indication of a posterior fossa tumor.

In rare cases some unusual symptom may initiate the disease. In one
case, failing vision because of angiomatosis retinae was the first symptom and
in another case mental symptoms resembling a frontal lobe syndrome were
the first symptoms to be observed. The evolution of the disease is quite
variable and in our material, the length of history before admission varied
between a few weeks up to over 2 years. About two thirds of our patients
came under neurosurgical care within a year after the appearance of the
first symptoms. The history is apt to be shorter and the appearance of
severe symptoms, particularly signs of foraminal herniation, tends to occur
earlier in the younger patients, and a protracted course is perhaps more
often seen in the elderly patients. There appears to be no connection between
type of tumor, whether cystic or solid, or location, and length of history.

The onset is usually gradual but occasionally a very sudden onset of
severe symptoms has occurred. In one case, in which the tumor had not been
found at the first operation and only a suboccipital decompression had been
made, hemorrhage into the tumor precipitated a pressure syndrome of
extreme severity. The course before admission to a neurosurgical clinic is
apt to fluctuate somewhat, with remissions alternating with periods of
more severe symptoms. Occasionally fairly long remissions may occur but
in most cases there is a gradual but steady increase in the severity of symp-
toms.

At the time of admission all patients complained of headache. Vomiting
had occurred in 55 cases or 80 per cent. Choked disc was found in 63 cases; normal eyegrounds were present in only 6 cases. In 1 case, the eyegrounds could not be seen because of secondary changes in the media, following angiomatosis retinae. Four patients were blind at the time of admission, 1 because of angiomatosis retinae, the other 3 because of choked discs with secondary atrophy. Visual acuity was more or less affected in 18 cases. In about one third of the cases, therefore, increased intracranial pressure had existed long enough to affect visual acuity. Evidence of foraminal herniation in the form of cerebellar fits was present in 14 cases and in an additional 8 cases stiffness of the neck was found, although cerebellar fits had not yet occurred. Bulbar symptoms in the form of hoarseness and inability to swallow occurred in 2 cases; both patients died before operation could be performed and these symptoms, as well as other evidence of foraminal herniation, are therefore to be considered as a sign of utmost urgency. Other common symptoms of intracranial tension were double vision, usually temporary, in 17 cases and paralysis of upward conjugate movements of the eyes in 7 cases.

A syndrome of intracranial tension was therefore found in every case and it was well advanced in a large number of cases. Long-standing intracranial hypertension found its roentgenological expression in the form of erosion of the posterior clinoids, increased convolutional markings, etc. in 18 cases. In the early days before the extreme urgency of these cases was recognized, especially when cerebellar fits made their appearance, no less than 5 patients died before operation could be performed and 1, also in the ultimate stage of medullary compression, died in the general surgical clinic after an operation for pleural empyema.

Among the localizing symptoms, disturbances of cerebellar functions are the most frequent and important. Cerebellar disturbance of gait and station was found in 56 cases, asynergia and hypotonia, usually one-sided, in 34 cases, and nystagmus in 29 cases. Unless appearing early before severe intracranial hypertension has set in, disturbance of gait and station is of little localizing value, as any patient who has been severely ill and bedridden for some time is apt to be more or less unable to walk. Among the cerebellar symptoms only nystagmus is of greater localizing value in those cases in which signs of intracranial tension have been present for some time.

Palsies of cranial nerves are not infrequent. Weakness of the facial nerve, usually homolateral, was observed in 15 cases. Homolateral decrease of hearing occurred in 2 cases. More often homolateral tinnitus is observed; this symptom, which is rarely permanent but usually occurs in short periods, was observed in 6 cases. Sensory disturbance in the field of the trigeminal nerve was found in 10 cases: in 2 cases these changes were bilateral; in 8 they were one-sided and located to the side of the lesion. Although double vision occurs frequently, palsies of the 3rd, 4th and 6th nerves are rare; only occasionally has an abducens palsy been observed.
A cerebellar syndrome with severe pressure symptoms, particularly signs of foraminal herniation, in an adult person having a history of less than a year is quite suggestive of an angioreticuloma. However, many other tumors, especially tumors of the brain stem, pinealomas and sometimes even tumors of the cerebral hemispheres, may give rise to a similar syndrome and it is therefore altogether unsafe to rely upon the neurological findings alone. The presence of angiomatosis retinae would of course clinch the diagnosis but this condition is so rare that it is devoid of practical value for the diagnosis of cerebellar angiomatosis. Contrast roentgen examination is therefore necessary to substantiate the diagnosis.

Of our patients who underwent surgery, only 3 were operated on without previous air studies of any kind. In 1 of these cases, a cerebellar cyst had been found at operation in another hospital some years earlier; in the other 2 the diagnosis was based upon the neurological findings alone. These were early cases, however, and increasing experience has convinced me that apparently typical neurological syndromes can lead to diagnostic errors which in critical cases are very apt to lead to disasters. Therefore, in every case in which there is the slightest doubt, it is advisable to substantiate the diagnosis by ventriculography.

Lumbar encephalography is definitely contraindicated when a cerebellar angioma is suspected, unless one is prepared to operate immediately. However, since the encephalograms in such cases are very likely to be negative because no air enters the ventricular system, this principle cannot always be followed. Should this be the case it is advisable, if pressure symptoms exist, to follow up the procedure immediately with ventriculography and operation. Otherwise the lumbar puncture might provoke a fatal foraminal herniation.

Suboccipital puncture and air injection was attempted in 9 cases in which a cerebellar angioma was unsuspected. In 2 of these, the needle struck a cyst in one of the herniated tonsils and the diagnosis was established by filling the cyst with air. In 1 case a cyst had been tapped through a suboccipital puncture in another clinic but no air had been injected. In the remaining 6 cases either no fluid was obtained or the injected air did not penetrate into the ventricles. Ventriculography was subsequently performed in these cases and the tumor was verified and removed at operation. Although no harm resulted in any of these cases which were thought suitable for suboccipital air injection because the symptoms of intracranial tension were mild, suboccipital puncture in cases of cerebellar tumors is attended by a definite risk, particularly when the tumors are angiomatous, because the needle might easily strike a dilated blood vessel in one of the herniated tonsils.

Air injection after tapping the ventricles is by far the safest and also the most reliable method for establishing the diagnosis of cerebellar angioma. It must be emphasized, however, that ventriculography must be followed
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by immediate operation, otherwise the procedure is extremely dangerous. In 2 of our early cases, the importance of immediate operation was not yet appreciated and the patients both died within 24 hours from the increase in pressure following air injection. In another case, in which ventriculography had been performed 24 hours earlier in another hospital, the patient arrived in the neurosurgical clinic in a moribund condition with a temperature of over 40°C. and Cheyne-Stokes respiration. Immediate operation and removal of the tumor failed to save the patient from the effects of this ill-advised ventriculography. In the remaining 59 cases in which operation was subsequently carried out, ventriculography was performed without incident and definitely established the position of the tumor. Sometimes when the tumor is very large no air enters the fourth ventricle and the exact position of the tumor in the posterior fossa may be in doubt. Angiograms of the vertebral artery may then be very useful for completing the diagnosis.

A specific indication for vertebral angiography arises in cases of familial occurrence of angiomas. Experience shows that multiple tumors are frequently present in these cases and angiograms may succeed in disclosing the presence and location of multiple tumors, a fact that ventriculography is quite unlikely to unearth, and which may easily escape attention at operation.

TREATMENT

The operative procedure has not changed much in recent years, but modern methods of anesthesia have contributed materially to the ease and smoothness of its performance. Local anaesthesia has been used in the majority of our cases and this is still the preferable method when the patient is in an advanced stage of intracranial hypertension. If general anesthesia is used, ether should never be employed because its vasodilative action may increase the volume of the brain so much that, in spite of ventricular puncture, there is insufficient room in the posterior fossa for the manipulations attending removal of the tumor. This is of less importance if there is a large cyst present, but when the tumor is largely or entirely solid and of some considerable size, it is necessary to have all the room one can get, particularly if the tumor is situated in an awkward position high up under the tentorium. Intubation is of course essential in any form of general anesthesia and the anaesthetic to be used should be nitrous oxide and oxygen combined, when necessary, with pentothal or nembutal. A method that has recently been introduced by Gordh in this clinic is to combine a high spinal anesthesia with general anesthesia. The reduction of blood pressure associated with spinal anesthesia gives more room in the cranial chamber than any other method and also reduces hemorrhage. The blood pressure can be easily controlled by tilting the table in the appropriate angle. This method has not yet been tried in a case of angioma, but has been used to great advantage in operations on acoustic tumors and other tumors in the posterior fossa, and is considered in this clinic as a method of considerable promise.
Although a unilateral exposure might be sufficient in many cases of angioma, a bilateral exposure is always made in this clinic. The reason is that if some complication causing the intracranial pressure to rise occurs during operation, for instance hemorrhage into the tumor or respiratory embarrassment, it may be impossible to enlarge the dural opening with sufficient rapidity to accommodate the swelling cerebellum and great damage may occur before conditions in the operative field are again under control. The arch of the atlas is of course always removed, as a foraminal herniation is almost invariably present. If a cyst is present, this is opened widely and it is then usually easy to remove the mural node. The solid tumors present a much more difficult technical problem. They are usually large and vascular and when situated in an awkward place, they may be difficult to remove. It is seldom of use to try to remove the contents of a solid tumor with the electric loop; it is always much too vascular for this procedure. It bleeds very easily at the slightest touch and before beginning to mobilize the tumor, it is well to expose it as fully as possible by removing the adjacent cerebellar tissue by suction. If it is in a reasonably accessible position, it can then gradually be mobilized and the entering blood vessels clipped as more and more of the tumor is brought into view. When situated on the superior surface of the cerebellum it may be impossible to see and clip the entering blood vessels and this difficult situation is best handled by rapidly enucleating the tumor with brain spatulas and then clipping the blood vessels afterwards when the tumor no longer obstructs the view. After careful hemostasis the dura is closed by interrupted stitches, no attempt being made to make the closure water-tight.

In cases where the tumor is situated high up under the tentorium, an approach from above might be preferable to the usual cerebellar exposure. This was done in one of our cases, where the tumor was located to the superior part of the vermis, extending upwards to the quadrigeminal plate. A large occipitoparietal flap was made, the tentorium was split and the tumor, which was entirely solid, was removed without much difficulty.

RESULTS

In 6 cases, the patients died before operation could be performed. Two of them, both early cases in the series, had been subjected to ventriculography and died within 24 hours from increased intracranial pressure. In the remaining 4 cases, death occurred suddenly shortly after admission. These patients were all admitted in a late stage of intracranial hypertension; they all had typical cerebellar fits. Under present-day conditions they could probably have been saved by immediate ventriculography and operation, or by ventricular drainage during the days preceding operation.

In 2 cases suboccipital decompression was performed. In one of these, a very large, entirely solid and very vascular tumor was found high up in the vermis. The tumor was thought to be inoperable, and after biopsy the operation was terminated as a decompression. He died the same day,
probably from loss of blood and the effects of intracranial tension. Autopsy showed a large midline tumor, extending to the quadrigeminal plate in the cranial direction (Fig. 3). This tumor could probably have been removed from the transtentorial approach. In the other case, no tumor was found at operation. Angiography some days later showed the angiomata to be located high up under the tentorium in the right hemisphere (Fig. 1). The patient refused a second operation and did very well for more than a year, when he had a sudden hemorrhage into the tumor and was brought to

Fig. 3. Autopsy specimen of large mid-line angioreticuloma.

the clinic in a comatose condition. At the second operation a large solid tumor, accompanied by a small cyst, filled with recent clot, was removed from the superior surface of the right cerebellar hemisphere. The patient made an uneventful recovery but is still, almost a year after the second operation, unable to work because of cerebellar disturbance of gait.

In the remaining 62 cases the tumor was completely removed. There were 10 fatalities in this group, giving a mortality of 16.1 per cent. Altogether therefore, 64 patients were operated on with 11 fatalities, giving a mortality of 17.2 per cent. As might be expected, the tumors located to the floor of the fourth ventricle carry a very high mortality, the operation having terminated fatally in 2 out of 5 cases.

Five patients came to operation in a comatose condition and died within a day or two because of the combined effects of long-standing intracranial
tension, operative shock and loss of blood. Two patients died of meningitis after particularly long and difficult operations. Two died from the effects of a postoperative clot and 2 from pulmonary complications, pneumonia in one case and pulmonary embolism in the other. These results are not very brilliant, considering the fact that the cerebellar angiomas are benign tumors, on the whole favourable for surgical removal. Unfortunately many of these patients arrive in a neurosurgical clinic in a desperate condition, due to the severity of intracranial tension. It should also be noted that this report covers the experience of the author, such as it was, and that of several of his younger co-workers, during a period of almost 30 years. With earlier diagnosis and modern methods of diagnosis and treatment much better results may be expected in the future.

SECONDARY OPERATIONS

In the 3 cases in which familial occurrence of angio reticulomas was observed, new tumors appeared some years after the first tumor had been removed. In one of these, three tumors in different parts of the cerebellum were successfully removed, the first operation having been performed in 1937 and the last in 1949, and in her daughter a second angioma in the other cerebellar hemisphere was removed 3 years after the first operation. In the third case, two angiomas in different locations had been removed at the first operation and 2 years later still another tumor was removed, also in a different part of the cerebellum. As already mentioned, his present condition indicates that he may have still another angioma somewhere in the cerebellum.

In 2 other cases additional tumors occurred, but since they were localized to the same cerebellar hemisphere, it is impossible to say whether they were true recurrences or new tumors. In the first of these, a cyst-forming angioma was removed from the left cerebellar hemisphere in 1934; the removal was thought to be radical. In the beginning of 1938 another angioma in the same hemisphere was removed, but this obviously recurred very rapidly because at the third operation 10 months later a very large tumor, also in the left hemisphere and almost filling the entire posterior fossa, was removed, this time with an early fatality. At autopsy in this case multiple angio reticulomas in the kidneys were found, indicating a tendency to phakomatosis. In the second case, a cyst-forming angioma was removed from the left cerebellar hemisphere in 1941. In 1948 another tumor of the same type and location was removed, but it was impossible to decide from the operative findings whether a recurrence or a new tumor was present, and even though the first tumor was thought to have been radically removed, the case should perhaps best be classified as a recurrence.

LATE RESULTS

Of the 53 patients who survived the operation, 6 died later. One, as has already been mentioned, died as a result of removal of his second recur-
rencence 4 years after the first operation. Three patients died some months up to 6 years after operation from intercurrent disease. In none of these cases was there any suspicion of recurrence. One patient died 11 years after operation with symptoms indicating a lesion of the brain stem, but ventriculograms 3 months before her death did not indicate the presence of an expanding lesion. She had been well and able to work for 10 years after operation. One patient died in his home 7 months after operation of "cerebral hemorrhage." Since the patient was only 45 years old and did not have hypertension, it seems possible that the cause of death might have been hemorrhage into a recurrent tumor.

Among the patients still living, 3 are invalids because of blindness, 1 because of cerebellar incoordination, and 1 because of mental impairment. The remaining 42 patients are well, although some of them may have some minor defects such as some impairment of vision. Because of the location of these tumors near the surface of the cerebellum, cerebellar incoordination is rarely a cause of disability, as it frequently is with more deeply situated tumors, such as the acoustic tumors.

SUMMARY

In a series of 4,101 verified brain tumors, 70 cases of cerebellar angio-reticuloma were observed. In adults they occupy fourth place among the posterior fossa tumors, occurring with practically the same frequency as meningiomas, but less frequently than gliomas and neurinomas.

Cyst formation was found to be less frequent than generally thought, and almost one fifth of the tumors observed were entirely solid. In 5 cases, the tumor was located to the region of the fourth ventricle, in 9 to the vermis and in the remaining 56 to the cerebellar hemispheres. In 1 case von Hippel's disease of the retina was observed. Three families were observed where two or more members of the family were afflicted with angio-reticulomas.

Among the symptoms, intracranial pressure is the most important and also the first symptom in most cases. Cerebellar symptoms were found in most cases, but were often late in appearance. Cranial nerve palsies, especially facial weakness and sensory disturbance in the trigeminal field, were observed in several cases.

Air studies were carried out in all but 3 cases. In cases of familial occurrence, vertebral angiograms are indicated, as multiple tumors are frequent in such cases.

Six patients died before operation. In 1 case the tumor was considered to be inoperable and decompression only was performed with an early fatality. In 63 cases the tumor was completely removed, with 10 fatalities. In 64 cases the mortality therefore was 17.2 per cent. Tumors located to the floor of the fourth ventricle carry a very high mortality, the operation having terminated fatally in 2 out of 5 such cases.

Among the survivors 6 died later from intercurrent disease. Five of the survivors are invalids, 3 because of blindness, 1 because of mental
impairment and 1 because of cerebellar incoordination. The remaining 42 patients are well and able to work.

REFERENCES