THE CEREBELLAR ASTROCYTOMAS
A REPORT ON 98 VERIFIED CASES

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In 1931 Harvey Cushing² published his memorable article on cerebellar astrocytomas, presenting a series of 76 cases and describing in great detail the clinical features of the disease. He emphasized the relatively benign features of the tumour and the high percentage of surgical success that could be achieved. Of the patients operated upon, 18.6 per cent died in the post-operative period, 57 per cent were still alive at the time of writing, and 38.6 per cent had survived for 5 years or longer.

Cushing clearly established the very different nature of the cerebellar astrocytomas from most astrocytomas of the cerebral hemispheres. He also suggested that the surgical results were likely to improve with greater experience. In a subsequent report¹⁰ his case mortality in the last 29 consecutive new cases of cerebellar astrocytoma was only 3.4 per cent.

Since Cushing’s contribution a number of reports concerning the same tumour have been published, notably by Loisel²¹ (30 cases), Bergstrand⁴ (56 cases), Bucy and Gustafson⁵ (25 cases), Mabon et al.²² (131 cases), Ringertz and Nordenstam²⁹ (140 cases), Wertheimer and Corradi²¹ (44 cases), Matson²⁶ (34 cases) and Holub¹⁶ (54 cases). In addition to these, collections of similar cases have been referred to in publications dealing with intracranial neoplasms in general.¹,³,¹¹–¹⁵,¹⁷,¹⁹,²⁰,³⁰,³²

The present paper deals with 98 cases of confirmed astrocytomas of the cerebellum and roof of the fourth ventricle. There were 6 of the latter type of growth and they were included because of the similarity of their behavior to that of the cerebellar tumours proper.

All patients in this series were under the care of one of the authors (W.McK.) at the Maida Vale Hospital, Leavesden Emergency Hospital, the Atkinson Morley’s branch of St. George’s Hospital, the National Hospital, and the Hospital for Sick Children.

ETIOLOGY

The dominant incidence of this growth in childhood prompted Cushing⁶ to ascribe it to a congenital origin. In this he was supported, among others, by Marburg,²³ who described embryonic ependymal rests, usually found in children, which were originally connected with the ventricles of the cerebrum and cerebellum. In the cerebellum he described a median Hochstetter’s canal and median or sometimes lateral Essick’s cysts — both ependymal remnants. Marburg believed that gliomas could arise in and around these struc-

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tures and that ependyma-lined cysts could develop from these rests even without tumour formation. Surprisingly, Marburg described examples of ependyma-lined cavities only in cerebral gliomas, as Stroebbe had already done in 1896. One would have expected the finding of similar cavities also in cerebellar gliomas according to his thesis. In our material no record could be found of such ependymal inclusions.

Bergstrand also believed that cerebellar astrocytomas are congenital, describing neuroblasts as well as glial cells in his sections. He proposed, therefore, that the tumour be named glio-neuroblastoma. He found considerable support on the Continent but his findings were completely rejected by Bailey, who ascribed the appearance of neuroblasts to errors in staining and interpretation.

One point suggested by Cushing in connection with the etiology was that all cerebellar astrocytomas originated in the midline and only invaded the cerebellar hemispheres secondarily, but many authors have since noted growths that were purely lateral in situation, showing no obvious connection with the vermis. In the present series 36 neoplasms were entirely hemispheral and did not involve the vermis or impinge on the midline, the mural nodule, if present, lying well removed from the vermis.

**OVER-ALL INCIDENCE**

Obrador, in his collection of 4000 intracranial tumours, found 7.1 per cent of cerebellar astrocytomas and spongioblastomas. Cushing, considering only intracranial gliomas, found the incidence to be 10.6 per cent. More recently Penman and Smith noted that of the intracranial gliomas, 8.4 per cent were of the cerebellum and fourth ventricle.

If children only are considered, the percentage becomes much higher; Craig et al. in their collection of intracranial tumours in children up to the age of 15, found a 20 per cent incidence of cerebellar astrocytomas.

Narrowing the field still further to subtentorial gliomas, astrocytomas represented 38 per cent in Penman and Smith's series and 49.4 per cent in Ringertz and Nordenstam's material. In this connection, one must consider the age groups from which the figures were taken, but it will not be far from the truth if one accepts that approximately half of the posterior-fossa gliomas in children are cerebellar astrocytomas.

**AGE DISTRIBUTION**

Cerebellar astrocytoma is a disease of young people. The average age in this series was 13 years, agreeing exactly with that reported by Cushing who noted, however, as we did, a marked range in age distribution. Our material contained, for example, 2 patients aged 1 year, and 2 over 60 years of age. When divided into 5-year groups there were:

<table>
<thead>
<tr>
<th>Patients</th>
<th>Age</th>
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<tbody>
<tr>
<td>38</td>
<td>1–5 years</td>
</tr>
<tr>
<td>34</td>
<td>5–10 years</td>
</tr>
<tr>
<td>11</td>
<td>10–15 years</td>
</tr>
<tr>
<td>25</td>
<td>Over 15 years</td>
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This shows by far the greatest number in the first decade and more particularly in the latter half of it. It should be noted that our material was drawn from hospitals that among them admit patients of all age groups.

SEX DISTRIBUTION

In this series 58 per cent of the patients were male. A similar slight dominance of males was noted by Ringertz and Nordenstam. However, in Cushing's and Wertheimer and Corradi's series the distribution of sex was about equal, and Holub cited a slight preponderance of females.

SYMPTOMATOLOGY

The typical history in this disease is that of a child who for months or even years complains of headaches and vomiting, often made worse by stooping or straining. Exacerbations of symptoms may be separated by long periods of apparently good health. Four-fifths of our patients exhibited the above symptoms and not infrequently vomiting was noted before any complaint of headache was made, this being particularly so in small children. Somewhat later the child's gait becomes ataxic, its hands become clumsy, and it fumbles and drops things. A squint or diplopia may be noticed, this being caused by paresis of one or both lateral recti.

Occasionally dizziness or vertigo are complained of or the sight deteriorates. Finally the child becomes drowsy and shows signs of general deterioration. Slurred speech appeared to be uncommon, being mentioned by less than a tenth of the patients. Tilting of the head to one side or forward, stiffness of the neck, and deafness were infrequent complaints. Attacks of unconsciousness, probably caused by sudden increases of intracranial pressure, occurred in 11 per cent of the cases and were sometimes associated with an opisthotonic posture, the so-called "cerebellar fits."

The average length of history for all cases was 16 months.

So much for the general symptomatology, but if the clinical features produced by the tumours according to their site are contrasted, a distinct picture emerges for tumours placed laterally and for those in the midline. In our series 25 patients had median tumours, 36 had lateral growths, and 31 had lesions that involved both the vermis and one cerebellar hemisphere, i.e. median-lateral growths. The 6 patients with neoplasms arising from the roof of the fourth ventricle will be considered separately.

MEDIAN GROUP OF TUMOURS

The growths in this group originated in the vermis, though most of them had spread bilaterally from it, involving both hemispheres to some extent.

The average age of the patients was 8 years, much lower than the average for the whole series, and the duration of symptoms was 20 months, i.e., rather longer than the average for the entire series.

Headache and vomiting were the most frequent complaints and next in frequency came ataxia of the legs (72 per cent), ataxia of the arms being mentioned by very few (8 per cent). Squint and diplopia, often temporary,
occurred in over half the cases. Periods of stupor or unconsciousness and so-called fits were mentioned by no less than a fifth of the patients or relatives. Drowsiness, dizziness or vertigo, dysarthria, and failing vision occurred in 12–16 per cent. Paresis of one or more limbs and stiffness of the neck were uncommon complaints.

Turning to clinical signs, it was found that papilloedema was an almost constant feature. Ataxia of the lower limbs was noted in 84 per cent but some ataxia of the arms was also present in 64 per cent, even though a far less frequent feature in the history. Clinical hydrocephalus was found in almost half the cases, while squint or nystagmus occurred in just over a third of the patients.

Alteration of tone and reflexes, each noted in 31 per cent, were rather variable and unreliable signs, often an increase rather than decrease being noted. Extensor plantar reflexes were seen almost as frequently. Presumably these features were caused by a mixture of effects of destruction of the cerebellum and direct or indirect involvement of the medulla.

Dysarthria, impairment of vision, paresis, and stiffness of the neck were detected in only a tenth of the cases in this group.

**LATERAL GROUP OF TUMOURS**

These growths, confined to one hemisphere, show an interesting contrast when compared with the median group.

The average age of the patients was 18 years, much higher than in the group with midline growths, but the duration of symptoms was slightly less at 18 months.

The symptomatology of the group again showed a high incidence of headache and vomiting, as shown in the section on general symptomatology. Ataxia of the legs was mentioned by 61 per cent and ataxia of the arms by 42 per cent. Obviously much more attention was paid in this group to ataxia of the upper limbs than in the group with median tumours. Diplopia or squint was only half as frequent as in the median group, occurring in approximately a quarter of the cases. The incidence of dizziness or vertigo and periods of unconsciousness or so-called cerebellar fits was similar to that in the median-tumour cases, occurring in about a fifth of the patients.

When clinical signs were analysed it was found that papilloedema and ataxia of the legs or arms were present in approximately the same proportion as in the median group. It is interesting to note that although ataxia of the upper limbs was equally frequent in the above two groups, it must have been more obvious in the lateral group to be so frequently described by the patient himself. Ataxia was an excellent lateralising sign in this group, only 1 example being found in which it was misleading as to the site of the lesion. Nystagmus, present in two-thirds of the cases in the lateral group, was a far less useful lateralising sign, being correct in only 22 per cent of the cases; in the rest it was either equally intense to both sides or even suggested the opposite side as the site of the lesion. This again demonstrates the mixture of effects of the tumour on the cerebellum and the brain stem.
Facial weakness was noted in a third of the cases but like nystagmus it proved of poor localising value. The same applied to alteration of tone and reflexes, an increase rather than decrease being found often. An excellent lateralising sign, though present in only 22 per cent, was tilting of the head away from the side of the lesion, only 1 patient showing a tilt towards it.

Clinical hydrocephalus was present in 22 per cent, only half the incidence in the median group. Impairment of vision was also detected in 22 per cent and in 1 case had progressed to blindness. Paresis of one or more limbs was found in less than a fifth and deafness or dysarthria in less than a tenth of the patients in this group.

MEDIAN-LATERAL GROUP OF TUMOURS

These cases formed about a third of the series. Little needs to be said about them since they presented a mixture of the features of the above-described median and lateral groups. Of the 31 tumours in this group, 7 appeared to have originated laterally and 5 medially; in the rest the site of origin was uncertain.

The average age was 11 years and the duration of symptoms was 10 months.

ASTROCYTOMAS OF ROOF OF FOURTH VENTRICLE

There were 6 of these cases in the series. The average age was notably low at 7.5 years and the history was of only 8 months’ duration.

The symptomatology, however, was very much the same as in the cerebellar tumours and there were examples of lateral, median and median-lateral growths with clinical pictures appropriate to the site of the growth as already described.

PATHOLOGY

Of all the tumours in the series 86 per cent were cystic, the contents of these varying from 4–110 cc. The latter phenomenal quantity was aspirated from a lesion in a girl of 10 who was admitted with a 6 months’ history of headache and vomiting. Clinically she had a moderate hydrocephalus, some incoordination of the limbs, and papilloedema. At operation she was found to have a large cystic growth of the vermis extending into one hemisphere and containing a large mural nodule. Only the latter was removed completely and radiotherapy was given subsequently since the tumour showed histological evidence of moderate malignancy. Six years later the patient was a successful saleswoman and on examination showed only a slight ataxia of her limbs. Three years later she had lost even these mild signs and was living a normal life.

It is interesting to note that Ringertz and Nordenstam found 78.6 per cent cystic tumours in their series and Mabon et al. cited the incidence of cystic lesions at two-thirds in their cases.

Further subdivision of the cystic neoplasms in our series showed that of 84 such tumours 10 could not be classified because of insufficient data, but of the remaining 74, 47 per cent were of the nodule-and-cyst type in which
the tumour tissue is concentrated in a mural nubbin, the remainder of the cyst being non-neoplastic. A further 45 per cent were of the cyst-within-tumour type in which all of the cyst wall is composed of neoplastic tissue and the cyst itself may form only a small part of the tumour. The remaining 8 per cent were polycystic growths.

The solid tumours were more frequent in the median group than in the rest of the series, amounting to 28 per cent. It must be understood that solid growths, as described here, were entirely solid macroscopically and contained no cystic spaces at all. If, however, “mainly solid” growths had been included, the percentage would obviously have risen and it is therefore easy to understand why other authors have cited a much higher incidence of solid neoplasms.

The histological picture was of low malignancy and did not appear to vary greatly with the site of the tumour. Approximately half of the neoplasms in the series had been classified according to Kernohan’s grading of the degree of malignancy: 79 per cent were grade I, i.e. of very low malignancy, 17 per cent were grade II and therefore still relatively benign, while 4 per cent were grade III, being fairly malignant. There were no highly malignant grade IV lesions in this series. The remaining half of the tumours in the series had been described in many ways and according to various classifications in use at the time, nevertheless it was clear that the great majority of the lesions were of a low grade of malignancy.

Invasion of the medulla occurred in 12 per cent of the cases and although this prevented complete removal the results after partial removal were reasonably satisfactory, as will be shown later.

**RADIOLOGICAL APPEARANCES**

The radiological features of these tumours of the cerebellum and fourth ventricle should be mentioned. For this purpose the films and reports of the patients treated at the Atkinson Morley’s Hospital were reviewed, amounting to 57 cases or rather more than half the series.

Plain radiograms offered a good deal of information, for 76 per cent showed evidence of increased intracranial pressure. This was deduced from the presence of diastasis of sutures, the most frequent single finding, erosion of the clinoid processes, or an obviously hydrocephalic skull. Increased convolutional markings were not taken as reliable evidence because of the difficulty of interpretation.

Enlargement of the posterior fossa appeared to be very variable, often a “normal” fossa being present with a large tumour. No easy way could be discovered to judge what exactly is a large fossa for any particular size of skull.

Considerable attention was paid to thinning or definite enlargement of one side of the posterior fossa as compared with the other and to pathological calcification. The latter feature was present in only 9 per cent of the cases, a slightly lower figure than shown by other authors.6,7,24,25 Taking these signs together, preoperative localisation was made in 20 per cent of cases, and reviewing the films in the light of operative findings the figure
could perhaps be increased to 31 per cent. These figures do not include cases in which a posterior fossa lesion was inferred from other features in the films or the history.

The next stage in radiological investigations was ventriculography, this being a routine procedure throughout the series with only a few exceptions in the early cases. If, then, all the patients admitted in the last 10 years are taken, and this accounts for three-fourths of all the material in the series, it is found that only 2 patients did not undergo the procedure, one of whom was brought in dead and one who died soon after admission.

In 92 per cent the lesion was shown successfully by the ventriculogram, in a further 5 per cent sufficient information was obtained to locate the lesion in the posterior fossa and in only 3 per cent was the ventriculogram inconclusive. Myodil was used whenever information obtained from the air pictures proved inadequate. It may be added that even when all the early ventriculographic reports are taken into account, the rate of success falls only to 95 per cent for the whole series.

An interesting point emerged when measuring the degree of hydrocephalus in the ventriculograms. This was done by calculating the ratio of the maximum width of the anterior horns of the lateral ventricles to the internal width of the skull. Altogether 69 patients were measured in this way, 21 of these having been treated by ventricular drainage before craniotomy; the remaining 48 had not been drained.

Taking the undrained group first, it was found that there were 12 patients with a ventricle-skull ratio of over 0.5 and of these 58 per cent died in the postoperative period, while of the remaining 36 patients, with a ratio of 0.5 or less, only 14 per cent died following operation.

The drained group of 21 patients included 4 with a ratio of over 0.5; none of these died and indeed there was only 1 death in this group.

From these figures it appears that without drainage a high degree of ventricular dilatation carries a very high operative mortality. It is possible that this high death rate could be diminished by ventricular drainage; at least the generally low mortality rate of the patients treated by drainage is impressive and the survival of the 4 patients in this group with a ratio of over 0.5 is suggestive that this may be so. Larger numbers, however, are necessary to make this convincing.

**TREATMENT**

As far as possible radical removal of the tumour was attempted; this consisted of either complete removal of solid or partly cystic growths, or removal of the nodule from a cyst-and-nodule type of lesion. As Cushing had pointed out, the removal of the nodule from the latter type of neoplasm was sufficient to prevent recurrence, provided this, the active part of the tumour, was removed completely.

The total rate of craniotomy was high, 94 per cent of all patients having been subjected to it, and this figure remained approximately constant regardless of the site of the tumour. The remaining 6 per cent were accounted
for by 4 patients who died after ventriculography or burr holes and 2 who succumbed before any treatment could be commenced. One indeed was brought in dead; the house officer, however, examined the fundi and recorded the presence of papilloedema.

Radical removal was achieved in 65 per cent of all the patients and in a further 16 per cent partial removal was accomplished, while in the remainder only biopsy or aspiration was considered advisable.

In cases in which only partial removal, biopsy or aspiration could be achieved, and a satisfactory enough recovery ensued, irradiation was given as a routine measure.

MORTALITY RATES

The total case mortality for the series was 22 per cent and of those patients subjected to craniotomy 17 per cent died in the postoperative period. It should be mentioned here that operative mortality figures included all patients dying in the neurosurgical department, however long that might have been after operation.

In the last 10 years these figures have improved considerably and for the last 67 cases the case mortality was 14 per cent while the operative mortality had fallen to 12 per cent.

With reference to the type of tumour regardless of location, the following figures were obtained:

- Case mortality for solid tumours: 36%
- Operative mortality for solid tumours: 36%
- Case mortality for cystic tumours: 20%
- Operative mortality for cystic tumours: 14%

In view of the above figures it will not be surprising that in the median group of tumours, with its higher proportion of solid tumours, the case mortality was 32 per cent and operative mortality was 24 per cent, these percentages being considerably higher than those for the rest of the series.

SURVIVAL

Of all the patients in the series 65 per cent are still alive and of those admitted over 5 years ago 57 per cent have survived and are still being followed up.

The type of operation or degree of completeness of removal did affect the prognosis, as can be seen from the following table.

- Surviving after complete removal: 80%
- Surviving after partial removal: 64%
- Surviving 5 years after complete removal: 76%
- Surviving 5 years after partial removal: 56%

Medullary involvement was the usual reason for only partial removal.

THE QUALITY OF SURVIVAL

In conclusion this interesting and important feature should be considered, for by it the real success of the operation can be appreciated. For this pur-
pose the patients were divided into four categories. Category A includes only those with no abnormal neurological signs and leading a normal life. Category B patients have mild neurological deficits but still live a normal life. Category C have marked neurological abnormalities and lead a restricted but gainful existence. Category D patients are disabled and cannot maintain themselves or continue with their education in a normal fashion.

<table>
<thead>
<tr>
<th>Type of Survival</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Category A</td>
<td>18%</td>
</tr>
<tr>
<td>Category B</td>
<td>24%</td>
</tr>
<tr>
<td>Category C</td>
<td>15%</td>
</tr>
<tr>
<td>Category D</td>
<td>8%</td>
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</tbody>
</table>

It can be seen that of all the patients in the series 42 per cent are still alive and leading a normal existence while 57 per cent in all are able to maintain themselves, earn their living or continue with their education in a fairly normal fashion. The high rate of return to normal life is the most gratifying aspect of the follow up. Sports and games are mentioned by many, one being a keen "rugger" player while others delight in tennis or swimming. Quite a number have married since the operation and are now happily bringing up a growing family.

SUMMARY

A series of 98 cases of astrocytoma of the cerebellum and the roof of the fourth ventricle is described.

Tumours arising in the cerebellar hemisphere are found to be more common than those originating in the vermis.

An attempt is made to define clinical syndromes for the various situations of the tumour.

It appears that tumours of the vermis have a higher operative mortality than the rest of the growths in this series.

Even partial excision was followed by a good long-term survival rate even when the medulla was involved by the growth.

The radiographic changes of the skull are noted and the accuracy of location of the lesion by air or Myodil ventriculography is commented upon.

It is suggested that the degree of ventricular dilatation present preoperatively may have a bearing on the immediate mortality rate.

We wish to thank Mr. K. W. E. Paine, F.R.C.S., of the Atkinson Morley's Hospital for his very helpful suggestions in connection with this paper.

REFERENCES