Medulloblastomas and Cerebellar Sarcomas
A Clinical Survey

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From 1928 to December 31, 1958, that is over a 30-year period, 109 medulloblastomas and 20 cerebellar sarcomas were submitted to the Department of Neuropathology of the Montreal Neurological Institute and thus classified. This amounts to 4.05 per cent of a total of 2,443 intracranial tumours examined during the same period of time.

It is recognized that the existence of a separate entity of cerebellar tumours histologically very similar to, if not identical with, medulloblastomas has been a controversial subject for some time and the question has not yet been settled satisfactorily. Hanbery and Dugger suggested that the term peri-thelial sarcoma be retained to include similar tumours classified variously as primary reticulo-endothelioma, microglioblastoma, reticulum-cell sarcoma, perivascular sarcoma, and alveolar sarcoma. Recently, Smith et al., after reviewing the literature and evaluating their own therapeutic results, felt that, from a practical clinical viewpoint, cerebellar sarcomas were not distinguishable from medulloblastomas and that the distinction of these sarcomas from medulloblastomas, usually on the basis of the stain for reticulin on histopathological examination, was still a debatable point.

The percentage of medulloblastomas in 1,522 gliomas (Montreal Neurological Institute series, 1957) amounted to 6.7 per cent, which is comparable to the percentage in other large series, such as that of Ringertz and Tola who counted 7.06 per cent or 111 medulloblastomas in a total of 1,571 gliomas, or Christensen’s series of 4.8 per cent or 93 medulloblastomas in a total of 1,928 gliomas (Fig. 1).

In our series, follow-up was obtained in 128 of the 129 cases reviewed (medulloblastomas and cerebellar sarcomas), i.e. 99.2 per cent; 1 patient returned to her native Norway after operation and was not traceable.

In the medulloblastoma group of 109 cases, 4 patients were not treated and there were 22 postoperative deaths (i.e. patients who died within 1 month following operation).

Of these 22 postoperative deaths, 12 occurred following radical removal, 1 following partial removal and irradiation, 6 partial removal without radiotherapy, 2 were twist-drill biopsies with radiotherapy, and 1 underwent twist-drill biopsy followed by right ventriculoperitoneal shunt.

Of the 88 patients suffering from medulloblastoma and surviving longer than 1 month after operation, 7 had radical removal without radiotherapy, 47 had radical removal with radiotherapy, 7 had partial removal without radiotherapy, 11 had partial removal with radiotherapy, 10 had biopsy followed by radiotherapy, 1 of whom had a biopsy by craniotomy and the other 9, twist-drill biopsies, and 1 patient was treated by ventriculoperitoneal shunt.

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Of the 20 patients with cerebellar sarcoma, 1 was not treated; 1 died within the month following twist-drill biopsy without radiotherapy, thus constituting a postoperative death. Ten had radical removal with radiotherapy, 3 had partial removal without radiation, 2 had twist-drill biopsy with radiotherapy, 1 had laminectomy and removal of secondary deposit of tumour without treatment of the primary lesion which was thought to be a tumour of the 3rd ventricle on ventriculography, but at autopsy turned out to be a cerebellar sarcoma which had seeded through the ventriculoperitoneal shunt into the peritoneal cavity.

**Age and Sex Incidence**

In the group of medulloblastomas, the highest incidence occurred in patients aged between 5 and 7, with a peak at 6 years of age (Fig. 2).

If the incidence is computed on the basis of decades (Fig. 3), the highest incidence was noted in the first decade with 65 patients, of whom 40 were male and 25 female. In the second decade there were 18 males and 7 females. In the third decade there were 7 males and 6 females, in the fourth decade 3 males and 2 females and 1 male patient was in the 51–60 age group.

In the group of sarcomas (Fig. 4) the highest incidence was in the first and third decades. Contrary to the figures for medulloblastomas, there were more females than males. The youngest patient with sarcoma was a 3-month-old infant who was not operated upon, the tumour being found at autopsy. The oldest patient was a man aged 55 years.

The incidence of males was 67.8 per cent in the group of medulloblastomas, and 40 per cent in the group of sarcomas.

**Main Symptoms and Signs**

In the groups of medulloblastomas and cerebellar sarcomas the main symptoms and signs are shown in Figs. 5 and 6. It may be seen that vomiting, headache and unsteady gait were the main symptoms in both groups. As far as the objective findings were concerned, the three most important by far were increased intracranial pressure, cerebellar dysfunction and papilloedema. Diplopia, drowsiness and dizziness were of lesser importance. It is interesting to note at this point...
that 9 patients with medulloblastoma and 3 with cerebellar sarcoma were investigated first as a sequel to head injury. Turning now to the main signs presented by the patients, it was found that nystagmus, palsy of the 6th nerve, facial weakness, nuchal rigidity and Macewen's sign were of lesser importance as a presenting sign in the above order of decreasing frequency.

Radiological Appearance

This has been discussed in detail in another paper from the Montreal Neurological Institute and will not be dealt with here. It is, however, interesting to note that on ventriculography two tumours were thought to be lesions of the posterior part of the 3rd ventricle and therefore no other treatment but ventriculoperitoneal shunt was performed. At autopsy both tumours were shown to have been situated primarily in the cerebellum and were medulloblastomas. In both cases there were secondary implants of tumour in the peritoneal cavity via the shunting tubes.

Treatment

In reviewing the literature, one notes that various forms of therapy have been attempted. Spitz et al. removed only enough tumour to open up a pathway for the cerebrospinal fluid. Bailey et al. advocated biopsy for diagnosis after craniectomy and wide decompression. Cushing did a thorough local removal by dissection and suction. Cutler et al. favoured radiological therapy of the suspected cerebellar tumours without histological diagnosis. Frazier et al. performed a biopsy only at the time of the initial decompression. Subtotal removal was attempted in other cases. At the Montreal Neurological Institute, the following methods of treatment were in use up to 1947: total (i.e. to the naked eye) and subtotal removal, with or without radiotherapy. In 1947, the late Dr. W. V. Cone began using twist-drill biopsy of suspected cerebellar tumours; medulloblastomas and cerebellar sarcomas were not operated upon, but irradiated.

Following ventriculography and determination of location of the tumour, the biopsy is carried out in the surgical dressing room with the patient postured on the cerebellar headrest. A twist-drill hole is made in the occipital bone, between theinion and the mastoid process, taking care to avoid the lateral sinus, the whole being performed under local anesthesia. A No. 15 brain-biopsy needle is inserted not more than 7 cm. in the estimated site of the lesion and the material for examination is withdrawn. Biopsy is not attempted
when intracranial pressure is low so as to avoid possible complications from ruptured blood vessels. Following confirmation of tumour, irradiation is begun.

Results of Therapy

Patients who died within 1 month following any sort of operative procedure, including twist-drill biopsy, were designated arbitrarily as postoperative deaths.

Of the 56 patients with medulloblastoma (Table 1) who were operated upon and irradiated, the average survival was 3 years, 3 months and 3 weeks. The average survival of 10 patients treated by twist-drill biopsy and irradiation was 3 years and 4 months. Fourteen patients were operated upon but received no radiotherapy and in this group the survival was markedly shorter, averaging 1 year and 3 weeks.

Of the 13 patients with sarcoma (Table 1) operated upon and irradiated, the average survival was 4 years, 9 months and 1 week. Two patients with cerebellar sarcomas in this series had twist-drill biopsies and irradiation, their average survival being 3 years, 3 months and 2 weeks.

When the combined group of medulloblastomas and sarcomas (Table 1) is considered, again excluding the postoperative deaths, the average survival in 69 patients operated upon and irradiated was found to be 3 years, 6 months and 2 weeks. Compared to this group, the 12 patients who underwent twist-drill biopsy with irradiation showed a slightly shorter average survival time of 3 years and 4 months.

That twist-drill biopsy with or without radiotherapy is not such an innocuous procedure can be seen from the example of the following 3 cases. The first was a 3-year-old male child who received a total radiation of 300 r to the skin and died 2 days postoperatively. The second, an 8-year-old boy, received two roentgen-ray treatments and died 2 days following twist-drill biopsy. The third, a female aged 33 years with a cerebellar sarcoma, died on the day twist-drill biopsy was done, of cerebellar coning.

Five-Year Survival

Of the total of 129 patients in the survey, 12 with medulloblastoma and 7 with sarcoma, operated upon and irradiated, survived 5 years or more. Two (with medulloblastoma) treated with Dr. Cone’s method of biopsy followed by irradiation lived 12 and 7 years respectively, and are, to the best of our knowledge, still living and in good health, with no evidence of recurrence.

Of the other 10 patients—all with medulloblastoma treated by operation and irradiation—7 treated by radical removal of the tumour survived an average of 10 years, 4 being alive at the time of this report with no signs of recurrence. The other 3, treated by partial removal of the tumour, died after surviving 5, 10 and 24 years respectively.

Out of 30 cases of sarcoma in a period of 13 years, from 1945 to 1958, all but 1 of which were treated by some form of operative procedure, there were 6 patients who survived over 5 to 13 years, 4 of whom remain alive at the time of this report: Over 5 years 2 (1 living); over 6 years 2 (1 living); over 9 years 1 (living); over 12 years 1 (living). In 1 case there has been no recent follow-up and this patient is presumed living. The only patient not treated surgically died 2\(\frac{1}{2}\) months after
admission from the effects of a lung abscess. This was a male patient of 64 years, in whom autopsy showed evidence of perithelial sarcoma.

**Statistical Evaluation of Therapeutic Results.** The pairs of "samples" have been compared by means of the "t" test, as shown in Table 2.

Our value of "t" for the samples therefore lies between P 0.1 and P 0.2. In other words, between 10 per cent and 20 per cent of such samples would show by chance alone a difference as great as, or greater than, those obtained. Or, at the 10 per cent level, there is no significant difference between the results obtained, i.e. the difference can be explained by chance.

In conclusion, although the average survival time is longer for patients with sarcoma than it is for those with medulloblastoma, this difference is not significant statistically. Larger numbers of samples, i.e., cases, will have to be studied and followed, before a statistically significant trend might become evident.

**Summary**

A total of 100 medulloblastomas and 20 cerebellar sarcomas were investigated during a 30-year period at the Montreal Neurological Institute. The cases of medulloblastomas are divided into groups according to age, number and sex of patients in each decade, location of tumour and predominance and duration of symptoms. The average survival is calculated, and the possible benefits of different treatments discussed including subtotal or total removal of the tumour without irradiation, operation plus irradiation, and Dr. Cone's method of twist-drill biopsy and irradiation. The merits of each procedure are discussed and an attempt is made to determine whether these differences are significant statistically.

In reviewing the results it appears that there is very little difference in survival between the two groups, i.e. those medulloblastomas treated by twist-drill biopsy and roentgen ray, and those treated by radical removal and radiation. The same may be said about those classified as sarcomas. In addition, longevity in the groups of medulloblastoma and sarcoma is very similar in the categories of treatment just mentioned.

**References**


11. **KINNEY, T. D. and ADAMS, R. D.** Reticulum cell

**TABLE 2**

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<th>Treatment</th>
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<td>Medulloblastoma</td>
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<td>Radical removal with radiotherapy, vs.</td>
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<td>Twist-drill biopsy with radiotherapy, vs.</td>
<td>0.6–0.7</td>
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<td>Medulloblastoma vs. sarcoma, all treatments</td>
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