CEREBELLAR ASTROCYTOMA IN CHILDHOOD

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IT IS GENERALLY appreciated that medulloblastoma, perhaps the most rapidly growing and highly malignant of all brain neoplasms, commonly involves the cerebellum and fourth ventricle of young children. Unfortunately, the gloomy outlook for this tumor has too often spread to encompass all brain tumors in childhood and overshadow the fact that cerebellar astrocytoma, a tumor at the opposite end of the scale of differentiation among gliomas and perhaps the most favorable of all intracranial neoplasms at any age, is almost equally common in young children. It is the purpose of this presentation to discuss the characteristic symptoms and findings, both clinical and laboratory, of this benign tumor of the cerebellum in children and to stress the high rate of curability when proper treatment is carried out early enough.

PATIENT MATERIAL

This report is based on a series of 34 consecutive astrocytomas of the cerebellum in children seen in our clinic since September of 1948, that is, during a period of 7 years. We have not gone back further in our records to expand the series because it seemed wiser for this purpose to discuss only results achieved by current, uniform methods.

During this period a total of 170 new intracranial tumors in children were treated; 115, or 67 per cent of these were located in the posterior fossa, that is, in the cerebellum, pons, fourth ventricle or cisterna magna. In addition to 34 astrocytomas there were 36 medulloblastomas, 14 ependymomas, 19 gliomas of the pons, 5 dermoid cysts, 1 choroid plexus papilloma, 1 tuberculoma and 5 miscellaneous neoplasms.

In summary, in the pediatric age group, over a given 7-year period, 20 per cent of all intracranial tumors and 30 per cent of posterior fossa tumors, were cerebellar astrocytomas.

RESULTS

Of these 34 consecutive children with cerebellar astrocytomas, 32 are now living and free from increased intracranial pressure. The 2 fatalities include a child who died 4 years after 2 unsuccessful attempts, at 9 months of age, to remove a mid-line tumor which extended from the cerebellum through the tentorium into the middle fossa, and a second child who died 9 months after surgical removal of the tumor as a result of unsatisfactorily treated secondary communicating hydrocephalus. There was no operative mortality among this group of patients, a total of 39 operations in 34 children. The case mortality is 5.8 per cent to date. In 30 of these patients, complete excision of the tumor was carried out at the first operation; 3 patients were operated upon in 2 stages; 1 patient had 3 operations over a 2-year period with eventual complete removal of the tumor.

In reporting a recent series of cases such as this, it is obviously impossible to state an absolute cure rate or to claim a definite proportion of complete removals of the tumor because only follow-up study of many years could make this certain. However, with this particular type of tumor, the surgeon knows exceptionally well whether or not he has removed the entire lesion. It is my judgment that in 27 of these 34 patients complete removal was definitely accomplished; in 5 patients complete removal is possible but uncertain; in only 2 patients is removal known to be incomplete. One of these two is the patient who died of remaining tumor 4 years later.

More important, perhaps, than the facts


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that 32 out of 34 patients with this type of cerebellar tumor are living, from a few months up to 7 years after surgical treatment, and that from 80 to 90 per cent of these are probably surgical cures, is the present status of the living patients after extensive surgery in the posterior fossa. We are all primarily interested, or should be, not only in survival but in useful survival. Is radical surgery justified?

Of the 32 living patients, 24 (75 per cent) are asymptomatic and have normal neurological examinations or have only a minimal degree of ataxia of one or more extremities by careful examination. The other 8 living patients are free of increased intracranial pressure but have residual neurological disturbances; 6 estimated as moderate, and 2 as severe. These disturbances include ataxia, cranial nerve palsies and diminution of vision. No patient is totally blind. With perhaps one exception, all of the living patients are happy and a joy to their families; all are at home. Here, then, is a group of children with intracranial tumors in whom the operative mortality is now virtually nil, the cure rate will probably be 80 to 90 per cent and the rate of completely normal development and function may be as high as 75 to 80 per cent. This illustrates why it is such a tragedy to temporize or to postpone treatment of these children on the assumption that a brain tumor in a child is probably malignant, the operative mortality prohibitive or the incidence of severe neurological deficit unduly excessive (Fig. 1).

DISCUSSION

Tremendous progress has been made in the technical management of cerebellar astrocytomas since they were first attacked surgically by Harvey Cushing between 40 and 50 years ago. In those days only 4 of the first 20 patients were operated upon in one stage, and only 11 of these 20 survived long enough to suggest removal was complete. The incidence of blindness and other complications was high. Results improved dramatically in Dr. Cushing’s hands and in 1931 he reported that most fatalities at that time were incident to secondary operations.1 The principal reasons for continued improvement in surgical results in such patients are: improved methods of general anesthesia and supportive care for children; improvement in methods of neurosurgical hemostasis; use of the sitting rather than the prone position during surgery (Fig. 2); selected use of pre- and postoperative constant ventricular drainage of cerebrospinal fluid. All of these maneuvers permit a higher frequency of one-stage surgical procedures. It is at the first operation that the surgeon has his best chance to enucleate this tumor completely and successfully.

An important problem connected with all surgery in the posterior fossa is the de-
Symptomatology

Whereas mass lesions in the cerebral hemispheres usually give rise to focal neurological disturbances, this is not true in the cerebellum. The symptoms or signs of slowly progressive increased intracranial pressure may be the only abnormalities and these may be so insidious in onset and varied in their manifestation as to delay recognition. The symptoms most commonly recorded in this group of patients have been headache, vomiting, unsteadiness of gait, pain in the neck and behavior disturbances; the latter being variously described as apathy, listlessness, irritability, generalized weakness or simply “he is just not himself” (Fig. 3). Any one of these complaints may be the first or the only one elicited.

In reviewing the histories of the patients in this series an unusual lag in diagnosis seemed to be present, most often for 1 of 3 reasons:

1) Because of the prominence of repeated vomiting without pain or other symptoms, the child was studied from the point of view of gastrointestinal disease,
chronic appendicitis, anorexia nervosa or poor dietary habits. The vomiting associated with cerebellar tumor is apt to be accompanied by little or no nausea or anorexia, it is usually intermittent, not necessarily forceful and often, though not always, precedes or immediately follows the first meal of the day.

Such a patient is R.P., a 7½-year-old girl, who vomited either before or after breakfast almost every morning for 4 months before hospital admission. During this time she was seen by several different physicians without a diagnosis being made. She was described as always a poor eater. She was an adopted child and a great deal of importance was placed on emotional tension in the family where there were also two natural children. It was not until the child developed a squint that an ophthalmologist was consulted who discovered bilateral papilledema. A large solid astrocytoma of the left cerebellar hemisphere was subsequently removed.

(2) The second type of history noted in these patients was that in which unsteadiness of gait was the predominant or only complaint. Such youngsters were apt to have had attention focussed on the feet and legs for some time with use of corrective shoes, physical therapy, gait training and orthopedic consultations.

Such a patient is L.S., a 3½-year-old girl, who began walking at about 14 months of age and was considered by her mother and a pediatrician from the beginning to have pronated feet. She was taken to an orthopedic surgeon and given corrective shoes which did not alter the gait. One year prior to hospital admission, at 2½ years of age, she was noted to bump into things and to fall frequently. At 3 years of age this was noted to be progressively worse and, under orthopedic supervision, she had gait-training exercises, another change in shoes and many roentgenographic studies of her lumbosacral spine and legs. Just prior to admission she developed pain in her neck and marked irritability. Roentgenograms of the skull then showed separation of the cranial sutures. Subsequently a large midline astrocytoma of the cerebellum was removed.

(3) The third type of history was usually obtained in somewhat older children or early adolescents, and consisted primarily of disturbance of behavior. This may have taken the form of simple lethargy, of indifference to surroundings, family and friends, of lack of initiative or of unwillingness to co-operate as usual. Such complaints are difficult to define and often the family cannot be sure they are real. They may lead to increased efforts at discipline or to child guidance and formal psychiatric consultation.

Such a patient is M.G., a 13-year-old boy, seen in the adolescent department with a history of “not feeling well” for 3 months. The mother stated the child was pale, didn’t want to do anything and seemed very nervous. She said he was high-strung, worried about his work and vomited frequently in the morning. She seemed over-solicitous to the physician. The boy did not think he was ill but said his food did not taste good. He complained that his mother was a poor cook and was a “private-eye” who did not let him do anything. He complained that his mother was a poor cook and was a “private-eye” who did not let him do anything. The tentative diagnosis was, “adjustment to adolescence.” Gastrointestinal roentgenograms were normal. After repeated interviews with mother and patient, a return visit in 6 months was arranged. However, in 2 months the boy was brought back with new complaints of headache, dizziness and unsteady gait. Subsequently a large cystic astrocytoma was removed from the left cerebellar hemisphere.

When headache was the principle complaint, the duration of the history was usually somewhat shorter, as might be expected. Certainly, the possibility of a resectable cerebellar tumor should be entertained and investigated in any child with one or more of the following complaints: recurrent headache; recurrent vomiting; any type of disturbance of gait; strabismus; pain in the neck; failing vision; unexplained chronic apathy, listlessness or irritability.

In this series of 34 patients, the sex distribution was essentially equal, 19 males to 15 females, and the age-range showed a predominance in the middle third of the first decade (Fig. 4). The length of the history, which in our opinion, is always only a
grossly approximate estimate of the duration of symptoms varied from less than 1 month up to over 24 months, and in itself, although averaging somewhat longer than for other more malignant types of tumor, proved of no diagnostic value (Fig. 5).

**Physical Signs**

The physical signs exhibited by children with cerebellar tumors are generally well understood, but certain features deserve emphasis. As already stated, these signs may be minimal until spinal fluid obstruction becomes extreme. If one looks for many of the so-called “cerebellar signs,” such as, nystagmus, dysmetria as determined by finger-to-nose and heel-to-knee tests, adiadochokinesia, and a positive Rhomberg test, he is apt to be falsely reassured by their absence. In many of the patients with cerebellar astrocytoma the sole evidence of ataxia is brought out by testing the gait. It is found that a wide base is maintained with a tendency to drift to one side or the other, and inability to walk on a line or turn quickly and reverse direction.

Another misconception pertains to the importance of spasticity and hyperactive reflexes. Their absence is of little or no significance. Intrinsic tumors within the cerebellum are accompanied almost uniformly by a decrease in muscular tone and by marked generalized depression of deep tendon reflex activity. These findings are usually more striking in the lower than in the upper extremities.

There is a prevalent impression that because separation of the cranial sutures in young children permits some spontaneous decompression, slowly increasing intracranial pressure does not ordinarily cause choking of the optic discs. It is interesting, therefore, to note that in this group of 34 patients with cerebellar tumors, 30 (91 per cent) showed papilledema at the time of hospital admission and almost all of these were described as chronic; that is, papilledema which had undoubtedly been present for some time. The obvious comment is: ophthalmoscopic examination should never be omitted in young children who are diagnostic problems, or put off on the ground that it requires a consultant, simply because it may require a little additional patience and practice. If children have the funduscopic examination explained to them and have it carried out so that the eye not being examined remains unobstructed and can fix
on something interesting, the examination can usually be performed with complete satisfaction.

Although these are slowly growing tumors and severe symptoms may be long in appearing, there are certain special laboratory signs which may often be helpful at an early stage. These consist essentially in evidences of chronic low-grade increase in intracranial pressure.

Röntgenograms

Whereas, in an adult there may be little or no evidence of increased pressure on plain roentgenograms of the skull, in a child visible separation of the cranial sutures is a
then the sagittal. In the series of patients with astrocytomas under study, 28 of the 34 showed separation of the cranial sutures at the time of first roentgenographic study. An early use of this examination in problems of vomiting, headache, strabismus, neck pain and chronic lethargy in childhood is recommended.

Electroencephalograms

In addition to roentgenographic and ophthalmoscopic examination a third objective test of definite, but less consistent, help has been electroencephalography; 31 of the 34 patients in this series had preoperative electroencephalograms. Only 7 of these were considered to be within normal limits. The predominant abnormality in the records was the presence of slow-wave and slow-spike activity, almost always associated with an increase in wave amplitude. This was sometimes seen throughout the tracing and at other times only paroxysmally, occurring occasionally in all leads, but always more prominent in the occipital leads and sometimes only there (Fig. 6). There is nothing diagnostic, of course, about this type of

Fig. 6. Duration of symptoms reasonably attributable to the tumor in this series of 34 children with cerebellar astrocytoma.

very sensitive sign. In posterior fossa tumors the coronal suture usually separates first, and

Fig. 7. Electroencephalogram of child with mid-line cerebellar astrocytoma showing the characteristic slowing and increase in amplitude of waves in both occipital leads.
electroencephalogram. It is really a record of increased intracranial pressure due to any subventricular obstruction to cerebrospinal fluid circulation. However, as a screening test, and as an aid in differentiating a lesion in the posterior fossa from one in the cerebral hemispheres, in the presence of increased intracranial pressure without focal signs, it has proved of definite value.

Ventriculography

The use of air contrast studies in the presence of internal hydrocephalus due to tumors obstructing the ventricular system has had a rather bad reputation because of a supposed high incidence of complications and even fatality. This is probably justified if lumbar pneumencephalography is carried out in the presence of increased intracranial pressure, or if an attempt is made during ventriculography, in the presence of marked hydrocephalus, to obtain complete replacement of fluid by air, or if ventriculography demonstrating marked obstruction of the aqueduct of Sylvius or fourth ventricle is not followed promptly by surgical relief of the obstruction. If, however, ventriculography is performed under local anesthesia using relatively small amounts of air (25 to 75 ml.), and subsequent to making the films the ventricle is tapped again to insure relief of increase in pressure before general anesthesia is induced, it is a safe procedure and often an invaluable one. In this series of 34 cerebellar astrocytomas, ventriculography was carried out in 28 patients without mishap. In every case air study was followed by immediate operation.

If ventriculography reveals normal or only slightly dilated ventricles in a suspected posterior fossa tumor, but does not show the aqueduct or fourth ventricle, it is our custom to then introduce small amounts of air (10 to 20 ml.) by the lumbar route (Fig. 7). This should never be done when primarily a cerebellar tumor is suspected in a child and never unless a Burr hole or separated sutures are available to allow immediate access to the lateral ventricles if necessary. In our experience it is rare in the presence of posterior fossa tumor that diagnosis cannot be made definitely by partial ventriculography if satisfactory films are made (Fig. 8).

These tumors are slowly growing and may reach large size before producing sufficient obstruction to spinal fluid circulation to cause symptoms of increased intracranial pressure. A common, but by no means universal, feature is the presence of a cyst containing clear, light yellow to amber colored fluid with a protein content often so high that the fluid clots on exposure to the air. In this series, 22 of the 34 tumors were partly cystic and the remaining 12 were entirely solid. The presence of a cyst, particularly a large one, is of course a great boon to the surgeon because of the additional room and improved exposure which can be obtained quickly.

One of the more unfortunate aspects of these lesions from a surgical point of view is that such a high percentage of them occur largely in the vermis; 12 of these 34 tumors were limited entirely to one or the other cerebellar hemisphere. The surgical management and postoperative courses of these patients were uniformly benign. The other 22 tumors were primarily in, or extended to, the mid-line. Encroachment directly into the walls or roof of the fourth ventricle always adds to the surgical hazards and to the incidence of postoperative cranial nerve complications.

Cerebellar astrocytomas are not sensitive to safe quantities of deep radiation therapy. They are not affected by any known chemical therapeutic agent. The treatment is surgical. Excellent results can and should be obtained whenever this is carried out before irreversible damage to the brain has occurred.

SUMMARY

Thirty-four children with cerebellar astrocytoma have been treated at Children's Medical Center in Boston during the last 7 years. There was no operative mortality. The case mortality to date is 5.8 per cent.

Thirty-two of the thirty-four children are alive and free of increased intracranial pressure. Six have moderate and 2 have severe
neurological disturbance in the form of residual ataxia and cranial nerve disturbances. The other 24 patients are leading normal lives, and show no abnormality on neurological examination.

The importance and desirability of early recognition and complete one-stage surgical removal of cerebellar astrocytomas, which represent 20 per cent of all intracranial tumors of childhood, is emphasized by the high rate of cure which can be achieved.

It is estimated that the rate of cure for this intracranial tumor should be about 90 per cent. Significant neurological disturbance should probably not persist in more than 15 to 20 per cent of these patients.

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