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Cerebellar astrocytomas: a 24-year experience

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Abstract *Introduction:* Cerebellar astrocytomas are the most benign tumors of the CNS. Seventy to eighty percent are found in children. *Methods and results:* We report on 38 children under 18 who had cerebellar astrocytoma in the posterior fossa and were treated by a multidisciplinary team in our Neurosurgical Department from January 1974 to December 1997. We included all patients in whom the histopathological diagnosis was astrocytoma, regardless of malignancy. The diagnostic methods used were pneumoventriculography, cranial X-rays, CT scan, and MRI. All patients were treated surgically. Neither radiotherapy nor chemotherapy was indicated in patients with pilocytic or fibrillary astrocytomas. A greater prevalence was observed in female (25/38; 66%) than in male (13/38; 34%) patients. Histopathological results revealed 27 (71%) pilocytic astrocytomas, 8 (21%) diffuse fibrillary astrocytomas, 1 (2%) anaplastic astrocytoma and 2 (6%) glioblastomas. These tumors were more frequently located in the right cerebellar hemisphere; increased intracranial pressure syndrome was the most frequent form of clinical presentation. Total tumor resection was obtained in 29 (83%) cases and subtotal resection in 9 (17%). In 6 (16%) cases, ventriculoperitoneal shunts were placed to control persistent hydrocephalus after tumor excision. *Conclusion:* The most frequent complication was increased ataxia. The mortality rate was 8.5%.

Keywords Cerebellar astrocytomas · Malignant astrocytomas · Hydrocephalus · Aseptic meningitis · CSF fistula · Pseudomeningocele

Introduction

Cerebellar astrocytomas are the most benign tumors of the CNS. Seventy to eighty percent are found in children [4], and they account for 28% of all pediatric brain tumors [23] and 35% of all tumors in the posterior fossa [17]. Malignant astrocytomas are uncommon in the pediatric age group [5, 6, 11, 15], the majority being found in the adult population and some in young persons in the second decade of life [12, 18, 19].

Since the arrival of CT scans and MRI, it has been possible to diagnose these tumors while their size and the extent of neurological compromise are not significant. The advances in surgical techniques on top of the possibility of earlier diagnosis have meant that treatment results are considerably improved, cure rates of up to 100% being achieved in some cases [16, 21].

Between January 1974 and December 1997, 201 children were treated in our Neurosurgical Department for tumors of the CNS; 97 (48%) tumors were located in the brain and 104 (52%) in the posterior fossa. Of the 104 posterior fossa tumors, 38 (37%) were astrocytomas and 66 (63%) were of other histological types.

In this study, we review the principal clinical aspects and the results obtained with the treatment performed.

Materials and methods

For this retrospective study we reviewed the clinical case histories of patients under 18 years of age who presented at our institution with cerebellar astrocytomas from January 1974 to December 1997.

These patients were treated by a multidisciplinary team made up of pediatricians, pediatric neurosurgeons, neuropathologists, oncologists, radiotherapists, and physical therapists. We included all patients with a histopathological diagnosis of astrocytoma, regardless of malignancy (pilocytic, fibrillary, anaplastic and glioblastoma) and histological pattern (cystic, solid and mixed [21]). The diagnostic methods used were: X-rays, pneumoventriculography, CT scan, and MRI.

All patients were treated surgically, undergoing a bilateral suboccipital craniectomy with removal of the arch of C1 and opening of the dura with a Y-shaped incision. In all cases, including those of cystic tumors, total removal of the tumor was attempted. In patients who presented with symptoms of hydrocephalic decompensation, treatment started with the placement of an external ventricular catheter, which was followed by removal of the tumor. Patients with persistent hydrocephalus after tumor removal underwent postoperative shunting.

During surgery, 20 of the patients were in a sitting position whilst the remaining patients were placed in a prone position; ultrasonic aspiration was performed when the tumor was deep seated with extension to the brain stem. Hermetic closure of the dura was attempted in all cases.

In patients with pilocytic or fibrillary astrocytomas, a strict follow-up procedure was implemented to control possible tumor recurrence. Neither radiotherapy nor chemotherapy was indicated. In cases of anaplastic astrocytomas or glioblastomas treatment was completed with radiotherapy and chemotherapy. In the patient with anaplastic astrocytoma, radiation therapy was initiated 1 week after tumor removal. A total dose of 54 Gy was administered as fractionated doses given in 30 sessions over a 6-week period, with the focus on the tumor bed only. In the patients with glioblastomas, craniospinal radiation therapy was carried out; this was started 1 week after tumor resection, with a total dose of 54 Gy in fractionated doses given in 30 sessions over a 6-week period, with a boost of 35 Gy to the tumor bed and 25 Gy to the rachis. Treatment was completed with chemotherapy.

Observations and results

No significant variation was observed with age (Table 1). However, a greater prevalence was observed in female (25; 66%) than in male (13; 34%) patients.

Histopathological results revealed that there were 27 (71%) pilocytic astrocytomas, 8 (21%) diffuse fibrillary astrocytomas, 1 (2%) anaplastic astrocytoma, and 2 (6%) glioblastomas.

The histological patterns of the low-grade astrocytomas indicated cystic tumor in 25 (72%) cases, solid tumor in 6 (16%), and mixed tumor in 4 (12%). The most frequent location of these tumors was in the right cerebellar hemisphere (16 cases; 42%), followed by the left cerebellar hemisphere (Table 2). The most frequent clinical presentation was increased intracranial pressure syndrome (Table 3).

Table 2 Location of cerebellar astrocytomas (*RCH* right cerebellar hemisphere, *LCH* left cerebellar hemisphere)

Location of cerebellar asytrocytomas	*n*	%
RCH	16	42
LCH	10	26
Vermis	9	23
RCH + left quadrigemina	1	3
Vermis + brain stem	1	3
RCH + brain stem	1	3
Total	38	100

Table 3 Clinical presentation

Clinical presentation	*n*	%
Headache	30	90
Vomiting	28	84
Papilledema	27	81
Ataxia	19	56
Dysmetria	17	53
Nystagmus	9	25
Nuchal rigidity	8	24
Lethargy	6	17
Macrocephaly	4	12
Diplopia	1	3

For diagnosis, pneumoventriculography was performed in 7 patients, X-rays in all, CT scans in 31, and MRI in 9. All X-rays showed signs of increased intracranial pressure, and in 1 case a double outline of the posterior fossa was seen on X-ray. All CT scans revealed the presence of a posterior fossa tumor, the structural pattern of it, and hydrocephalus. MRI permitted better definition of the anatomical relationships through the use of three cuts in three planes. MRI is less limited than CT scan, as it does not visualize bone.

Total resection of the tumor was obtained in 29 (83%) cases and subtotal resection in 9 (17%). In 6 (16%) cases, ventriculoperitoneal shunting was performed to control persistent hydrocephalus after tumor excision. The most frequent complication was increased ataxia, which occurred in 26 (74%) patients, followed by aseptic meningitis (Table 4).

None of the patients had symptoms of air embolism.

Pseudomeningocele and CSF fistula were treated by suturing the wound at fistula level, bed rest lying on the back though raised to 45°, acetazelamide at 40 mg kg^{-1} day^{-1}, and lumbar punctures. Six (18%) patients required postoperative shunts as they failed to respond to treatment and had progressive hydrocephalus.

The mortality rate was 8.5%. Two (5.7%) patients died as a result of tumor recurrence and 1 (2.8%) patient with a tumor located in the left cerebellar hemisphere with extension into the brain stem died of immediate postoperative complications.

Table 1 Incidence according to age

Age (years)	1	2	3	4	5	6	7	8	9	10	13	14	Total
No. of cases	5	3	5	4	2	3	5	3	5	1	1	1	38

Table 4 Complications

Complications	*n*	%
Increase in ataxia	26	74
Aseptic meningitis	18	51
Pseudomeningoceles	15	43
CSF fistula	12	34
Dysphagia	8	23
Mutism	6	17
Subdural hematoma	1	3

Discussion

The incidence of cerebellar astrocytomas by age in these series did not differ significantly from those published by other authors [9]. However, an obvious female predominance was noted (66%).

Cerebellar astrocytomas are circumscribed, cystic, solid or mixed [24]. Cystic tumors account for between 70% and 85% [8, 10, 16, 18] of astrocytomas and are generally sited in the cerebellar hemispheres, while solid cerebellar astrocytomas appear within the vermis in 90% of the cases. Between 8% and 40% [8, 18, 19, 24] of the tumors in these locations may extend into the brain stem. The most common location in our series was the right cerebellar hemisphere (44%). Seventy-two percent were cystic astrocytomas, 16% solid astrocytomas, and 12% mixed astrocytomas. Three tumors (8.3%) extended into the brain stem.

Although some authors [17, 20] consider malignant astrocytomas to be extremely rare, others report incidences of 10% [21] and 26% [19]. In our series, malignant astrocytomas represented 12% of the cases.

Leptomeningeal dissemination can occur in a benign astrocytoma, or infiltration of the neck muscles can result from tumor resection [11]. This was not observed in our series.

Primary symptoms are often insidious and nonspecific. The duration of symptoms before diagnosis varies from weeks to several years, with a mean of 5–6 months. In children with malignant astrocytomas the time-lapse is short, with an average of only a few weeks [15]. Increased cranial pressure is the most frequent form of clinical presentation [1, 6, 18, 22], since these tumors are frequently accompanied by hydrocephalus. In our series 27 (81%) of the patients presented with increased intracranial pressure. Ataxia may be difficult to assess, above all in small children [9]. A deterioration in handwriting may be the only sign of ataxia in the upper limbs. Macrocephaly may be observed up to the age of 10 years [21].

Surgery was performed on the first 20 patients with them in a sitting position. There were no serious problems, although some difficulties in accommodating the patients were experienced. There was a certain degree of inconvenience and discomfort to the surgeon, especially in the form of arm fatigue, and the assistants also had some difficulty. However, the incidence of surgical complications, such as air emboli, frontal pneumoencephalus and subdural hematomas [16], was greatly reduced. This position eliminates the need for use of the Mayfield headrest, thereby avoiding the possibility of epidural hematoma or a depressed skull fracture, which can be caused by the pins of the headrest [21].

Although it is widely accepted that removal of the mural nodule [17] is sufficient in cystic tumors, we believe that total excision of the tumor should be attempted, since no complementary examination can fully establish that the cyst wall is totally benign. In our series, total excision of the tumor was achieved in 80% of cases. In the cases of subtotal resection the patients were only followed up.

The most frequent postoperative complication was increased ataxia [21]. This complication was present in 56% of the cases in our series, and was transitory in the majority of patients.

Another frequent complication is aseptic meningitis [21] appearing in the first week after surgery. Affected patients present with headaches, fever, and nuchal rigidity. A cerebrospinal fluid sample should be sent for culture, and Gram staining should also be performed to eliminate the possibility of bacterial meningitis. This complication is caused by the presence of blood in the cerebrospinal fluid and normally disappears spontaneously within a week. Patients improve significantly with the administration of steroids [21].

Another common problem is the development of pseudomeningocele as the result of subcutaneous collections of cerebrospinal fluid (CSF). This entity is related to alterations in the reabsorption of CSF owing to the presence of blood and to the increase in protein concentration and changed cell pattern in the CSF, especially in malignant tumors. The natural progression of pseudomeningocele is variable. Some are not resolved until shunt placement is carried out. Others persist for days or even 2–3 weeks and then clear up spontaneously [21]. This complication is clearly related to the dehiscence of the operation wound and CSF fistula with the possible risk of bacterial meningitis. If pseudomeningitis is accompanied by increased intracranial pressure and ventricular dilatation, a shunting procedure is required [21].

The postoperative incidence of hydrocephalus is variable around 30% [17]. In our series the incidence was 16% (6 cases). It has been suggested that hydrocephalus can be treated by endoscopic ventriculostomy in the floor of the third ventricle [3]. This procedure can be highly effective before tumor excision but is not so effective afterwards, since permeability of the aqueduct of Sylvius and of the fourth ventricle is generally regained. As hydrocephalus is produced by a block in the circulation of CSF at the level of the subarachnoidal space or arachnoidal granulations, the placement of an extracranial shunt is mandatory [21].

Another worrying complication is mutism, which appears immediately after surgery or after a brief period of

1–3 days [2]. It is generally associated with marked ataxia in the limbs and is always associated with a midline tumor [5]. Its pathogenesis is unknown, but it is believed to be related to a lesion of the dentate nucleus of the brain stem or the inferior vermis [14]. Mutism usually resolves in a few days or months.

According to some authors, however, speech and cerebral functions never become normal again [5].

Although surgical and immediate postoperative mortality of 5–25% was reported for earlier series [7, 13], it is currently accepted that it is extremely rare. Mortality could be caused by resection of a tumor, with resulting brain stem compromise, or of a deep cerebellar nucleus, or by a shunt-related complication, such as an acute subdural hematoma, pneumoencephaly, or infection [21].

In our series the mortality was 8.5% (3/38). One patient died 3 days after surgery as a result of the resection of a deep tumor with brain stem compromise. Of the remaining two, each had a multiform glioblastoma and died as a result of progressive tumor growth, one 12 months and the other 14 months after surgery.

Conclusions

1. The incidence of cerebellar astrocytomas is more frequent in women.
2. The majority of tumors are benign (97%).
3. The most frequent location is the right cerebellar hemisphere.
4. The most common clinical presentation is with increased intracranial pressure syndrome.
5. CT scan is a highly effective method of diagnostic evaluation.
6. The MRI allows better surgical planning (management).
7. Total resection of the tumor is possible in the majority of cases.
8. Postoperative ataxia has a favorable prognosis in the pediatric age group.
9. Surgical mortality is very low.

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