CLINICOPATHOLOGICAL STUDY OF CEREBELLAR ASTROCYTOMA: REPORT OF THIRTY CASES

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ABSTRACT: AIMS: Cerebellar astrocytoma occurs more often in children and young adults than in adults. They are the most common astrocytic tumours in children, accounting for 80–85% of cerebellar astrocytomas and 60% of optic gliomas. They comprise about 33% of all posterior fossa tumours in children and represent about 25% of all paediatric tumours. The aim of this study was to study the clinicopathological features and analyze the clinical outcome, complications and prognosis of total or subtotal excision of cerebellar astrocytomas. **METHODS AND MATERIAL:** In the present study, the sex distribution was male predominance: 17 patients were male and 13 were female. The age at diagnosis varied from 4 years to 28 years. 84% of the patients were below 20 years of age.
RESULTS AND CONCLUSIONS: Histopathological examinations showed low grade astrocytoma and two cases had malignant tumors which were pilocytic and non pilocytic. Reccurence and mortality in the present study showed that 1 patient had recurrence. Out of 30 cases, 2 patients expired (6%). The cause of death was brain stem dysfunction probably due to brainstem infiltration. In conclusion, the present study may be of importance to clinicians to establish the correct diagnosis and proper therapy about cerebellar astrocytomas.
KEYWORDS: cerebellar astrocytoma, prognosis

INTRODUCTION: Astrocytomas represent approximately 22% of all brain and spinal tumors (1). Cerebellum is a common site for involvement of these tumors, which account for 5-8% of all gliomas and nearly 35-40% of all posterior fossa tumors (2). Between 70 and 80% of cerebellar astrocytomas are found in children. Few patients are less than 1 year of age or older than 40 at the time of diagnosis. There appear to be no age peaks. It is surgically curable in vast majority of cases. The questions frequently arise to why astrocytomas are benign in the cerebellum of children and yet their cerebral counterparts behave in a malignant fashion, and most of the patients don't survive beyond five years. Bergstrand in 1937 clearly voiced the opinion that these cerebellar tumors are represented as congenital malformations and not true neoplasms and thus accounting for their benign behavior (3). This controversial question was thoroughly discussed by Bucy and Gustafson in 1939 (4). In 1968, Busy and Thieman reported a long term study which confirmed the general opinion that astrocytomas of the cerebellum are the most benign and most favorable of all brain tumors. The present work has thus attempted to examine and analyze the clinical outcome, complications and prognosis of total or subtotal excision of cerebellar astrocytomas from patients of all ages seen at Mamata General Hospital, Khammam, India, between the periods from August 2010 to March 2013.

MATERIALS AND METHODS : Thirty patients with cerebellar astrocytomas have been managed by the Department of Neurosurgery at Mamata General Hospital, Khammam, India, between the period

from August 2010 to March 2013 were studied prospectively. This study comprised of clinical features, such as age group, sex, presenting symptoms, and clinical signs; along with imaging findings, surgical outcome, histological findings and recurrent at follow up. All patients underwent CT scan head, plain followed by contrast. Twenty two patients underwent ventriculoperitoneal shunt prior to surgery. All patients underwent definitive surgery through suboccipital craniectomy. Total excision was achieved in 26 cases, subtotal in 4 cases. The diagnosis was histologically confirmed by the Department of pathology, Mamata Medical College. Three patients underwent radiotherapy following surgery. Out of 30 patients, 25 were available for follow up and were evaluated for clinical features and functional outcome. Repeat CT scan was done if symptoms are suggestive of recurrence. All patients with recurrence underwent surgery. The duration of follow up varied from one month to 2 years 8 months.

RESULTS:

CLINICAL PRESENTATION: Basic clinical data of the patients were as shown in Table 1. The sex distribution was male predominance: 17 patients were male and 13 were female. The age at diagnosis varied from 4 years to 28 years. 84% of the patients were below 20 years of age. The age distribution is given in the table 2. Commonest presenting symptom was headache, which was seen in 94% of cases, followed by vomiting 89% and 2 patients presented in altered sensorium. Other symptoms were visual disturbances (blurring of vision, diplopia, loss of vision), gait disturbances, ataxia of limbs, truncal ataxia, dysphasia, dysarthria, regurgitation of food & neck pain.

PHYSICAL SIGNS: The commonest physical sign was papilloedema, which was present in 23 patients. Followed by cerebellar signs in 22 patients. Other signs were optic atrophy, visual acuity & field deficits, nystagmus, other cranial nerve disturbances, and altered sensorium (Table - 3).

RADIOGRAPHIC FEATURES:

The cerebellar astrocytoma were most commonly seen in the cerebellum (19) followed by vermis (11). In the present study the radiological examination by CT-scan showed tumor enhanced on contrast administration in 6 cases, 28 cases had hydrocephalus. The tumor was cystic with solid component in 14 cases and only 23 % (7 cases) had tumors confined to vermis, and 77% (23 cases) to hemispheres.

HISTOPATHOLOGIC FEATURES: Histopathological examinations in twenty eight patients showed low grade astrocytoma and two cases had malignant tumors. Among 28 individuals, 21 were pilocytic and 7 cases were non pilocytic. Astrocytoma was microscopically dominated by monomorphous bipolar (pioloid) cells, which lay within a rich myxoid matrix and often showed an angiocentric radiating arrangement. No Rosenthal fibers or eosinophilic granular bodies were observed which normally could be found in astrocytoma. Other changes including hemorrhage, necrosis, mitosis, nuclear atypia, lymphocytic infiltration, and vascular endothelial hyperplasia were not observed. In the present study, pathologically they were astrocytoma grade 1 (17 cases), grade II (16 cases) and grade III/IV in 2 cases.

TREATMENT : Ventriculo peritoneal shunt was performed in 22 cases (73.3%). All patients underwent definitive surgery for the tumor through sub occipital craniectomy. Total excision of the tumor was achieved in 26 cases. & subtotal in 4 cases (Table-4).

RECURRENCE AND MORTALITY: Out of 30 patients 25 were available for follow up. The duration of follow up ranged from 1 month to 2 years 8 months. At follow up 1 patient had recurrence. Out of 30 cases, 2 patients expired (6%). The cause of death was brain stem dysfunction probably due to brainstem infiltration (Table-5).

DISCUSSION: Cerebellar astrocytomas are among the most frequent gliomas of the central nervous system. They account for 8% of all gliomas and 30% of all paediatric gliomas (5). Cerebellar astrocytoma is predominantly a tumor of childhood, 75% of the tumor seen in the first two decades of life (6). Overall cerebellar astrocytomas appear to occur most often during the middle to later half of the first two decades of life and early in the second decade, although there is a second incidence peak in young adulthood (i.e at 25 years of age) (7). The youngest patient in series was 5weeks old and the oldest being 41 years (6). Levy and Elvidge reported average age of 13 years and 11 months (8). In most series males out numbered females. Grant et al (6) reported equal frequency in males and females, with no obvious racial prediction. As series of cerebellar astrocytomas are reported in the modern imaging era, age at presentation is decreasing because of earlier detection. In our series youngest patient was of 4years and the oldest was 28years 83.5% of patients were in the age group 5-20 with the peak in the age group of 10-15 years. All malignant gliomas were seen in above 20years age. In our series males were 17 (56%) & females were 13 (44%). In the series of pancalet et al (9) out of 168 patients 91 (54.2%) were boys and 77 (45.8%) were girls (10). As In the other series there was no significant difference between the number of male (56%) and female (44%). However Gessinger and Bucy reported a high incidence in female than male children (11).

As in the order series, headache is the most common symptom, which was seen in 94% of cases, followed by vomiting (89%), gait disturbance (64%) and visual disturbances (54.9%). Our observations were in the accordance with Geissinger and Bucy studies, headache occurred in 100% cases followed in frequency by vomiting and gait disturbances (11). Pencalet and co-workers observed headache was presenting symptom in 77.4% of cases, followed by vomiting (73.8%) and cerebellar syndrome (69%), papilloedema was the commonest sigh, which was seen in 68.5% followed by cerebellar signs (46.4%) and abducent nerve palsy in 22.6% two (1.2%) patients presented with complete blindness (9). In our present observation 77.5% of the patients presented with papilloedema and 13% of patients with secondary optic atrophy. Cushing reported that 22 (29%) of 76 children with cerebellar astrocytomas were blind at admission and abducent palsy was seen in 15% of children (12). In our series 1 (3.9) was blind at presentation and 8 (28.4%) had abducens palsy. Seizures are extremely rare and maybe related to intracranial hypertension.

The tumor may occur predominantly or solely in the cerebellar hemisphere or in the vermis and occasionally infiltrate the brainstem. Among 26 cases reported by Geissinger and Bucy (11), 16 were hemispherical and 10 were in the vermis. Chandra et al found that among 56 cases of cerebellar astrocytoma, 13 were hemispherical, 19 midline and 24 were extensive tumor (13). In the present study, 11 (36.6%) cases had vermian lesion and hemispheric lesion was seen in 19 (63%) cases. Among the hemispheres no significant difference was seen between left and right side.

In the present study low grade astrocytomas were seen in 27 (90%) cases of which 21 (70%) were pilocytic astrocytomas. Anaplastic astrocytomas originating primarily in the cerebellum are rare (14). The incidence of anaplastic cerebellar astrocytomas and glioblastomas is approximately 3-5% and 1-2% respectively (11, 14) not unexpectedly these malignant astrocytic cerebellar tumor

are always found in adult population with the median age of occurrence being 53 years (15). A predisposing factor to the development of such a lesion de novo is a history of previous radiation therapy for completely unrelated tumours (16). In the present study, 2 (6.6%) patient had anaplastic astrocytomas.

Although long survival period are generally reported, a surprisingly high incidence of recurrence ranging from 7-49% is evident in many series (11, 17). The extent of surgical removal is the most common important factor bearing on recurrence. The recurrence is common after subtotal removal and recurrence after total removal is rare (18). Recurrence can occur after total excision but paradoxically cure is reported even after partial excision (18). Other factors that can be associated with the tumor's tendency to recur are grossly solid tumors. According to some authors the diffuse histological variant carries a worse prognosis than the juvenile one but is not predictive of recurrence. Hayostek and colleagues found that pilocytic astrocytomas are associated with improved survival and recurrence rates (19). Austin and Alvord (1988) however showed no correlation between histological variable and tumor recurrence (17). Pencalet et al reported 9.5% recurrence (9). In the present study 1 (3.3%) patient had recurrence.

Complete surgical resection is an important prognostic factor in relation to the tumour recurrence and patient survival (20). Pencalet et al also showed that it is a sole independent factor that significantly determines the risk of tumor recurrence (9). Some authors still contend that a complete resection is not essential, eliciting several cases in which there was no tumor recurrence several years after partial removal. In our series, extent of surgical resection had influence on recurrence (25 were available for follow up, duration of follow up: 1 month to 2 years 8 months, only one patient had recurrence). Total resection has no recurrence. Brainstem infiltration and non-pilocytic varieties of astrocytomas were associated with recurrence.

In this series of Gessinger and Bucy mortality rate was 15.4% with surgical mortality of 3.8% most patients had tumor extension into the brainstem (11). In Pencalet et al series mortality rate was 4.2% (9). In present study mortality was seen in 2 (6.6%) cases out of 30 cases, these 2 cases were female, in these 2 cases tumor was present in the vermis and infiltrating brain stem in both cases. Brainstem dysfunction was commonest cause of death probably due to tumor infiltration.

CONCLUSION: Cerebellar astrocytomas are predominantly seen in children with peak incidence in the first half of the second decade. This study clearly showed that commonest presenting symptom is raised intracranial pressure. Majority of cerebellar astrocytomas are hypodense on CT scan and enhancing with contrast. Commonest location is cerebellar hemisphere. Recurrence is more common with fibrillary variety. Brainstem dysfunction is the commonest cause of death. Prognosis is good in patients with total excision. This study may be of importance to clinicians to establish the correct diagnosis and proper therapy about cerebellar astrocytomas.

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Symptoms	No. of Patients	%
Headache	28	94%
Vomiting	26	89%
Visual Disturbances	16	54%
Gait disturbances	19	62%
Ataxia of limbs	11	36%
Truncal Ataxia	4	13%
Dysphasia, dysarthria Regurgitation	4	13%
Altered sensorium	2	7%

TABLE 1: Details of clinical data of patients with cerebellar astrocytomas.

Age	No. of Patients	%
0-5	2	6.66%
6-10	5	16.66%
11-15	11	36.66%
16-20	7	23.33%
21-25	3	10.33%
26-30	2	6.66%

TABLE2: Frequency of age groups in patients with cerebellar astrocytomas.

Signs	No. of Patients	%
Papilloedema	23	77%
Cerebellar signs	22	75%
Visual Activity & field Disturbances	14	48%
Nystagmus	14	48%
Cranial nerve Disturbance	11	36%
Optic atrophy	4	13%
Altered Sensorium	2	7%

TABLE 3: Frequency of physical signs in patients with cerebellar astrocytomas.

Treatment	No. of Patients	%
Total resection	26	87.7%
Subtotal resection	4	13.3%

TABLE- 4: Treatment details of patients with cerebellar astrocytomas.

Age/ Sex	Extent of resection	B. stem Involvement	Histo-pathology	Post op Survival	Cause of Death
4/F	Subtotal	+	fibrillary	11 days	B. stem dysfunction
4/F	Subtotal	+	fibrillary	7 days	B. stem Dysfunction

TABLE -5: Recurrence and Mortality data in patients with cerebellar astrocytomas.

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