BRAIN STEM GLIOMAS

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SUMMARY :

Sixty-two patients with brain stem glioma are presented and analyzed. The most common presentation was cranial nerve palsy with cerebellar and pyramidal tract involvement. Fourty-four patients received radiation therapy. In eleven cases a histological diagnosis was made at operation. 14 cases died during follow up. Radiation therapy increases the survival time and relieves clinical symptoms.

KEY WORDS : *Brain Stem, Glioma*

Brain stem gliomas constitute 10-20 % of all central nervous system tumours of childhood. The natural history of this group of tumours is a slow progression to death (1-6). The onset of neurological symptoms and signs is frequently insidious and inreased intracranial pressure usually does not develop until late in the course of the disease. This slowly-developing clinical picture and short survival frequently make early diagnosis difficult (3,4). Brain stem gliomas usually derive from the pons, but less commonly they may originate from the mesencephalone or medulla oblongata.

The reported approximate survival period is between 4 and 15 months and the majority of patients die within the first two years in the natural course of the disease (1,8). Since it is well-established that radiation therapy prolongs the survival time, it has been a principal mode of treatment for brain stem glioma (1,2,7,9,10,11). The median survival time of patients who received radiotherapy may prolong up to 47 months and five years survival rate has been reported as 41 % (1). Although, formerly surgery on brain stem gliomas could not be performed, because of the intraaxial location of operations in this location is now increasing, especially with the use of the ultrasonic aspirator and laser techniques (1,5,7,11).

Today, assessment of the prognosis of brain stem tumours can be made according to the side of tumour on CT, characteristic CT findings of the lesion and histopathological findings. This assessment gains importance in predicting the survival of patients as well as in deciding upon the most useful therapy modes.

CLINICAL MATERIAL

This report is an analysis of 62 cases of brain stem glioma seen from 1976 through 1987. The ages of patients in the study group ranged from 9 months to 55 years with 74 % between 0 and 10 years. The were 34 males and 28 females.

Gait disturbances, headache and motor weakness were the most common symptoms (Table 1). The most common finding was cranial nerve involvement (90,3 %). Sixth and seventh nerve involvement were the most common, occurring in over 69 % of cases. Nystagmus, evidence of cerebellar disorders and pyramidal tract signs were frequent manifestations (Table 2).

In this group of 62 brain stem gliomas, 44 patients (70.9 %) received radiation therapy (Table 3); the remainder received no RT because death occurred soon after hospitalization or therapy was refused by the patient or parents. Fourteen patients (22.5 %) received adjuvant chemotherapy with RT. Eighteen patients (29.0 %) underwent surgical exploration of the posterior fossa with or without a biopsy procedure; shunting procedures were carried out in ten patients who underwent exploration. In two patients, midline cysts were evacuated from the brain stem (Table 3). Diagnostic biopsy was performed in fourteen patients. Histological evaluation was obtained in eleven. Seven patients had grade I-II astrocytomas, and four had grade III-IV astrocytomas.

Twenty-five patients were followed up; 15 (24.1 %) who had received radiation therapy alone, for periods between one month and ten years: six patients who had undergone shunting procedures came for follow up between three months and six years; and four patients from whom biopsies were taken were followed up for periods between six months and five years. Fourteen patients (22.5 %) died between two days and four years after the diagnosis was established.

DISCUSSION

Brain stem gliomas are a group of tumours which have a rapid mortal course and an incidence of nine to ten times higher in childhood than in adults. It is rare in children younger than three years old and the mean age is reported to be 7.3 years (1,4,12). In our series, 38.7 % of the patients were between six and ten years old. The classical piscture of brain stem glioma is cranial nerve involvement which apprears after complaints such as drowsiness and headache.

Symptoms and signs such as gait disturbances, diplopia, dysarthria, and motor weakness may be associated. Cranial nerve involvement at a rate of 89 % has been reported (13). Most commonly involved are the sixth and seventh cranial nerves were the sixth and seventh. Involvement of the pyramidal tract and the cerebellar connections were also noted. Pyramidal involvement 75 % and cerebellar dysfunction 80 % in different series (4, 12, 13).

In our series, the rate of cerebellar dysfunction was 51.6 % and the rate of motor deficit 50 %. Brain stem gliomas are distinct from other tumour in that they do not cause raised intracranial pressure early in the course of the disease despite all these neurological signs. Pneumoencephalography and angiography have been most commonly used diagnostic procedures for brain stem gliomas before the CT. There has, however, been a significant improvement in their diagnosis and management with the CT coming into use.

Stroink et. al., in their study, identified distinct groups of brain stem gliomas based on CT scan characteristics such as contrast enhancing, being intrinsic or extrinsic, hypo or hyperdense, with or without cyst (14). These criteria based on CT appearance can help to determine which cases will benefit from surgery and also assess the prognosis. For example, it can be said that intrinsic, isodense and contrast enhanced focal brain stem gliomas tend to be low grade tumours and several of these patients have long survival times. In some series, localization of the tumour in the brain stem is accepted as another prognostic factor. For example, upper brain stem gliomas have a longer survival period (5). Briefly, CT shows important prognostic characteristics in brain stem gliomas.

It is well-established thet radiation therapy prolongs survival time, and is the major palliative method available (1,2,15,16). Some authors report that patients treated with RT alone have an average duration of life of 27 months after the first symptoms appear, and the five-year survival incidence is 41 % (1,4). It has been reported that patients treated with RT have survived as long as 10-20 year (17). In Lassman's series, the mean survival time of patients treated with RT was 15 months, whereas in the untreated patients it was 4 months (13,18). These survival times, in Panitch's series, are 47.2 months in patients treated with RT, 15.5 months in the untreated group (12). And in the same study, it is reported that survival time correlated well with clinical improvement after TR. The patients who responded to radiotherapy survived average of 60.8 months while those who showed no response survived an average of only 5.6 months. In our series the five-year survival incidence is 12.8 %.

Bailey described the treatment of brain stem gliomas as "a pessimistic chapter in the history of neurosurgery". For a long time, most neurosurgeons suggested that it was not technically feasible to carry out a tumour excision in the brain stem. But now, brain stem gliomas can be safely biopsied and sometimes resected. Coffey and Lundford obtained diagnostic tissue from 12 patients with pontine and mesencephalic masses with no morbidity and mortality using the CT guided stereotactic technique (18). And in more recent years, subtotal or total resection has been performed using adjusts such as the laser, the ultrasonic aspirator and evoked potential monitoring. Surgery in brain stem glioma can be performed for exploration, biopsy or tumour resection. Exploration and biopsy provide additional prognostic information and valuable for planing treatment. The histopathological characteristic of the brain stem glioma is the heterogeneity of the tumour, so single site biopsy is of little value and multiple biopsies are required for diagnosis of the lesion (5,6,8).

The histological classification of pontine gliomas has frequently been discussed. Most of the tumours are low or high grade astrocytomas. In Jenkin's series, 48 percent of the patients were low grade astrocytoma, 39 percent were high grade astrocytoma (7). Lassiter's series contained 13 cases classified as astrocytoma and six as glioblastoma multiforme (10). In our series, histological examination was possible in 11 of the cases. Seven were grade I-II astrocytoma and 4 were grade III-IV astrocytoma, In three cases specimens were inadequate.

Panitch has shown a relationship between pathological diagnosis, clinical course and survival (12). In his series, the average survival time for patients with low grade astrocytoma was 32.4 months, while those with high grade astrocytoma survived on average only 6.4 months. It was seen that in Bilaniuk, Albright and Littman's series, the difference of survival time between low and high grade tumours was statistically significant (3,8,19).

In our series, 79 percent of patients received RT. The other 21 % were cases whose therapy was refused by the parents or who died before the RT. Fourteen patients died between two days and four years, Twenty-five patients have come to follow up for

periods between one month and ten years. The two-year survival incidence is 25.6 % and five-year survival incidence 12.8 %.

Table I : FREQUENCY OF SYMPTOMS

Symptoms	Number	%
Gait Disturbance	41	66.1
Motor Weakness	26	41.9
Vomiting	26	41.9
Headache	18	29.0
Squint or diplopia	14	22.5
Speech Defect	11	17.7
Dysphagia	6	9.6

Table II : FREQUENCY OF NEUROLOGICAL SİGNS ON ADMISSION

Signs	Number	%
Mental status change	14	22.5
Cranial nerve involvement	56	90.3
VI. nerve	43	69.3
VII. nerve	43	69.3
IX. nerve	14	22.5
X. nerve	14	22.5
V. nerve	8	12.9
Motor deficit	31	50.0
Sensory deficit	9	6.2
Extensor plantar response	38	61.2
Cerebellar signs	38	61.2
Nystagmus	18	29.0
Papilledema	16	25.8

Table III : TREATMENT MODES

Treatment	Number	%
Radiation Therapy	49	79.0
Chemotherapy (with RT)	14	22.5
Shunting	10	16.1
Post. Fossa Exploration	4	6.4
Biopsy	14	22.5

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