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ORIGINAL ARTICLE

Central Neurocytoma: Determining Rate of Shunting and Results after Subtotal and Total Excision

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ABSTRACT

Objective: To assess the shunting rate and the results of control measures in patients having central neurocytoma who underwent total and subtotal resection.

Study design: A cross-sectional study

Place and Duration This study was conducted in People's University of Medical and Health Sciences for Women Nawabshah from Septmeber 2016 to Septmber 2020 .

Methodology: In this study, overall 15 patients were included. Every patient had a follow-up of 2 years. The age of participants was between 13 to 49 years. The data of every patient was recorded which included the demographics (age, gender), early mortality and morbidity, presentation of clinical factors, and radiological findings. The radiological findings include features, tumor location, hydrocephalus, recurrence, and residual. A transcortical approach was used for the treatment of individuals for both, total or subtotal excision. EVD (External Ventricular Drain) was installed which was later replaced by a shunt. For confirmation of diagnosis and guidance of the follow-up, the MIV index and histopathology were used. For residual tumor and recurrence, radiosurgery of Gamma knife or adjuvant radiotherapy was used.

Results: There were 3 patients who died due to sepsis and thalamic infarction after the total and subtotal excision. There were 9 patients who had a total excision and 6 patients who had a subtotal excision. Among the 9 patients with a total excision, 3 of them showed small recurrence at 1 year and 18 months follow-up along with a high MIB index. A total of 2 of them needed a shunt. In the remaining participants,the shunt was not placed. Among the 6 patients who had a subtotal excision, 2 of these patients had an early shunt insertion. Five patients were controlled by radiotherapy while one was controlled by radiosurgery. One patient, who was controlled by radiotherapy, failed radiotherapy in his second treatment at 18 months of follow-up.

Practical implication : total excision has better results for central neurocytoma and should be considered procedure of choice

Conclusion: Central neurocytoma may have a better chance of recovery than other intraventricular tumors if treated with total excision, with a lower incidence of shunt insertion throughout its course.

Keywords: neurocytoma, shunt insertion, excision, radiotherapy, total excision

INTRODUCTION

In 1982, neurocytomas were first defined as historically distinct entities [1]. According to the World Health Organization, these are rare grade II neuronal tumors [2]. They most likely have an incidence of less than 1% [3]. Mostly, neuronal tumors develop within the ventricles as a type of cancer that grows slowly. These tumors have a better chance of recovery after surgical intervention [4]. MIB index is used to determine the behavior biologically and also helps in directing the adjuvant therapy. If the MIB index level is greater than 2 percent, it is defined as tumor recurrence and poor prognosis [5]. To differentiate neuronal tumors from other CNS neoplasms, immunohistochemistry is used. Because of its rarity, the diagnosis and treatment of this neoplasm are debatable [6].

The reports that were collected contained scattered data related to the clinical management of patients. This is why the known information about the patients with the tumor and its management is very limited. For local control and survival, the ideal therapeutic option at this stage is the safe maximal resection. It provides a good long-term prognosis. If the resection is achieved completely, the chances of recovery are far more than expected. For subtotal excision, radiation therapy is chosen. However, it is an option for those who have recurrent diseases or cannot be operated on [7]. The study's objective was to assess the shunting rate and the results of control measures in patients who were diagnosed with central neurocytoma and who underwent total and subtotal resection. Sample size is very small in our study , and need more research on this topic.

METHODOLOGY

In this study, overall 15 patients were included. Each patient was diagnosed with intraventricular central neurocytoma. Every patient had a follow-up of 2 years after the confirmation of neuronal tumor radiologically as well as pathologically. The data of every patient was recorded which included the demographics (age, gender), early mortality and morbidity, presentation of clinical factors, and radiological findings. The radiological findings include features, tumor location, hydrocephalus, recurrence, and residual. Patients were managed either with transcallosal approach or transcortical approach if needed. An external ventricular drain (EVD) was inserted that was later replaced by a shunt.

For confirmation of diagnosis and guidance of the follow-up, MIB index and histopathology were used. After the surgery, to check the extent of the removal of the tumor, MRI and CT scans were used. It was mandatory to document whether the tumor is removed totally or sub-totally. CT scans and MRI were also used in the follow-ups to check whether the tumor is recurrent, progressed, or residual. Patients who had residual tumors or showed a progression in the follow-up, adjuvant radiotherapy was provided. For patients who had recurrent or residual tumors, Gamma Knife radiosurgery was done. In the presence of tumor, the ventriculoperitoneal shunt was inserted and the patients were re-operated.

The medical records of patients included the demographics (age, gender), early mortality and morbidity, extent of tumor excision (whether total excision or subtotal excision), presentation of clinical factors, and radiological findings. The radiological findings include features related to neurocytoma, tumor location, date of progression of tumor, hydrocephalus, recurrence, and residual. If the radiotherapy was given after the immediate postoperative period, it was defined as adjuvant therapy. If the patient was observed and then treated for progression after surgery, in that case radiotherapy was defined as salvage therapy. CT scans or MRI scans were used to detect the progression of the

disease. When the tumor was not regrown or the progression was not detected on MRI, it is called local control (excluding the patient that died during the surgery). From the date of the first surgery, local control was calculated.

RESULTS

Overall, 15 patients were involved in this research which included 9 males and 6 females. The age range was between 13 to 49 years. Table No. 1 shows the common clinical symptoms of the study participants. The most common symptom was headache, which represents 56% of the total symptoms, followed by seizures, blurred vision, and unsteadiness.

Table No. 2 shows the location of the tumor. The location showed extension to the occipital horns, frontal, or third ventricle.

A total of 12 (80%) patients underwent the transcortical approach and 3 (20%) patients underwent the transcallosal approach. There were 3 patients who died due to sepsis and thalamic infarction after the total and subtotal excision. The patients who died were excluded from the follow-up research. There were 9 (60%) patients who had a total excision and 6 (40%) patients who had a subtotal excision.

Each patient was diagnosed with central neurocytoma. Overall 3 patients had an MIB Index greater than 2%, among which 2 had total excision and 1 had subtotal excision. Five patients were controlled by radiotherapy while one was controlled by radiosurgery using a Gamma Knife. For 3 patients, a ventriculoperitoneal early shunt was inserted. About 1 of them had total excision and 2 of them had subtotal excision. Among the 9 patients with a total excision, 3 of them showed small recurrence at 1 year and 18 months follow-up along with a high MIB index. Overall 1 patient in the subtotal group, who was controlled by radiotherapy, failed radiotherapy in his second treatment at 18 months follow-up when there was a progression of residual tumor. Table No. 3 shows the summary of the results.

Table 1: Clinical features of the study participants

Symptoms	%
Headache	56
Blurred Vision	32
Unsteadiness	9
Seizures	3

Table 2: location of the tumor (n=15)

Location	N	%
Left	7	46.7
Right	2	13.3
Extending to third ventricle	1	6.7
Biventricular	5	33.3

Table 3: Results and outcome of the operations

	Total excision	Subtotal excision
No. of patients	9	6
MIB >2%	3	1
Recurrence	3	1
Residual	0	4
Gamma Knife	0	1
Radiotherapy	0	5
Reoperation	0	2
VP shunt	1	2
Local Control	5	0
Failure of control	0	1

DISCUSSION

The number of studies on the treatment of central neurocytomas is very limited. Through institutional case series, retrospective case reports, and meta-analysis of institutional experiences, the neurocytomas are being guided [8]. Surgery is an option chosen as initial intervention. Surveillance cannot be defined as the primary option [9]. Although the nature of these tumors is indolent, the patients require surgical intervention rather than surveillance. These patients have symptoms such as hydrocephalus or increased intracranial pressure. The symptoms of the tumor include blurred vision, mental disturbances, pyramidal signs, and increased intracranial pressure. It is very rare that these tumors do not have any symptoms.

Our research shows that the tumor was biventricular in 33.3% of the cases and located in the left ventricle in 46.7% of the cases. According to Shin et al., the tumor was located in the left ventricle in fifty percent of the cases and it was biventricular in only 15 percent of the cases [10]. In the current study 26.7% of the cases had a high MIB level, which was defined as aggressive nature. According to Brat et al. and Soylemezoglu et al., it was found that 25 percent of the cases had a high MIB level (greater than 2%), which were defined as more aggressive [11,12]. According to Rades et al., 25 percent of these tumors had an MIB level above 2% along with seventy percent local control [13]. According to Mackenzie et al., clinical follow-up was available for all 14 patients in a study of 14 cases [14]. The ability of a central neurocytoma to proliferate was found to be a useful forecaster of clinical outcome, whereas histological atypia alone was not.

There are a number of options available for the treatment of neuronal tumors. These options include complete resection (total excision) with radiotherapy, incomplete resection (subtotal excision), complete resection, and radiotherapy with incomplete resection and Stereotactic radiosurgery (SRS). A number of prior research studies states that local control and chances of survival is better with complete resection rather than incomplete one [15,16]. After incomplete resection, patients are provided with chemotherapy and radiotherapy after the surgery.

A total of 9 patients, representing 60%, had a total excision and 6 patients, representing 40%, had a subtotal excision. After 24 months of follow-up, 5 patients were locally controlled while 4 patients had high MIB (greater than 2%) in which recurrence was shown. Our research shows that among the cases of incomplete resection, radiotherapy was effective in 4 patients but it failed in one of them. On the other hand, the Gamma knife was effective in 1 patient. Our results can be compared with prior studies who have similar results [17].

According to Rades et al., complete resection, complete resection with radiotherapy and incomplete resection with radiotherapy had better local control instead of subtotal excision [18]. The survival rates in their study were 86.1 percent (subtotal) and 99.2% (total) after a 2 year follow-up.

Leenstra et al., reported the largest institutional experience. Their research showed that thirty-five percent of their participants were provided with adjuvant radiation initially. However, one third of their participants faced a recurrence of tumor. They selected their participants on the basis of atypical neurocytomas [19].

In our research, the most effective treatment was SRS. After the total excision, SRS controlled the recurrent and subtotal resected cases completely. According to Rades et el., postoperative SRS was significant for local control. It was 87 percent effective after incomplete resection with radiotherapy, and fully effective after a five year follow-up [19,20].

Even though SRS was effective, the tumor may progress in indolent cases. In the debate over adjuvant treatment, a study of 504 published institutional reports from 91 centers was conducted in 2002, which concluded that adjuvant radiation was beneficial after incomplete resection. The debate arose because the extent of resection affects only local control rather than survival, and the impact of symptomatic recurrences is not well established in the literature.

According to Kim et al., Gamma Knife SRS is the most effective primary and secondary treatment after the operation of central neurocytomas [20]. According to our study, a total of 3 patients were provided with VP shunt. 2 patients were the ones who were provided with VP in replacement of EVD. These 2 patients were the cases of total excision. Overall1 patient was provided with VP shunt because tumor was recurrent near the foramen, before the Gamma Knife therapy. Not a lot of information is collected by us related to shunting rate. However, as it is in the

body of the ventricle, the tumor is soft, and moderately vascular, the tumor can be extracted, the foramen has a plane of cleavage from the ependyma, and the tumor is attached to the septum. The above mentioned factors could be contributing to the low incidence of postoperative hydrocephalus, as well as the ease of draining the ventricles at the foramen with minimal blood loss.

CONCLUSION

Central neurocytoma may have a better chance of recovery than other intraventricular tumors if treated with complete resection, with a lower incidence of shunt insertion throughout its course. Complete resection is much more effective and it improves local control.

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