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Large lateral intraventricular tumors – Outcome of radical surgery

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ABSTRACT

This is a retrospective analysis of 145 cases of lateral intraventricular tumors that were larger than 4 cm in their maximum dimension. The aim of surgery was radical tumor resection. During the period January 2000 to December 2019, 145 cases of lateral intraventricular tumors were treated by surgery by an interhemispheric approach. There were 101 males and 44 females. The ages of the patients ranged from 2 months to 77 years (average 29 years). Histological examination of tumors identified 73 central neurocytomas, 20 choroid plexus papillomas, 23 subependymal giant cell astrocytomas (SEGA), 5 ependymomas, 21 gliomas, 2 primitive neuroectodermal tumors (PNET/embryonal tumors) and 1 atypical teratoid rhabdoid tumor (ATRT). Nineteen patients had mild to severe hemiparesis in the immediate postoperative period. Eight patients died in the postoperative period. At a follow up of 1 year 137 patients were leading active and symptom free lives. Twenty seven patients died of their disease. Tumor recurrence or re-growth was observed in 13 patients and 2 patients needed reoperation. Surgery on large lateral intraventricular tumors can be associated with significant postoperative morbidity and mortality. Majority of tumors in this location are relatively 'low-grade' malignant tumors and when successfully treated, the long term outcome can be gratifying.

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1. Introduction

Majority of lateral intraventricular tumors are histologically 'relatively benign' in nature and grow 'slowly' to acquire a 'large' or 'massive' size prior to initiating neurological symptoms and when diagnosed. The authors discuss their 20-year experience in treating 145 cases of lateral intraventricular tumors that were more than 4 cm in their maximum dimension. The aim of surgery was 'radical' tumor resection. The general clinical behavior and the surgical strategy for various types of pathological lesions matched. The technical issues regarding surgical approach, tumor debulking and control of tumor bleeding are elaborated. Whilst radical surgical resection can be 'curative', disastrous postoperative complications were encountered and have been described for large intraventricular tumors on multiple occasions in the literature [1,2]. Despite the fact that intraventricular site is a frequent location of large intracranial tumors; there are relatively few reported surgical series in the literature [3–6]. Essentially, surgery on lateral

intraventricular tumors necessarily involves high-level understanding of nuances of neurosurgery and perfect execution of technical skills. Issues of safe brain retraction, working in narrow surgical corridor, strategy and ability of tumor debulking and control of bleeding in the depth of the surgical field are crucial technical issues. The article essentially aims at providing epidemiological information and surgical subtleties involved in the treatment of large lateral intraventricular tumors. The study evaluates the general surgical outcome of various pathological lesions located in the lateral ventricles and treated by interhemispheric surgical approach. No specific attempt has been made to assess the surgical subtleties, management options and the need for or validity of postoperative adjuvant therapy for each individual pathological entity. Literature review of described treatment format of each individual tumor is beyond the scope of the article.

2. Material and methods

During the period 2000 to 2019, 145 cases of lateral intraventricular tumors that were larger than 4 cm in their maximum dimension were surgically treated in the neurosurgery departments of the authors. All patients provided written informed consent, and all clinical tests and surgical procedures were conducted



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according to the principles of the Declaration of Helsinki. Radiological investigations included contrast-enhanced magnetic resonance imaging (MRI) in all patients. Intraventricular meningiomas, colloid cyst, arachnoid cysts, epidermoid cyst, vascular malformation, hematopoietic malignancies, metastasis and infectious lesions are not evaluated. Tumors operated by any surgical route other than interhemispheric approach have not been included in the analysis.

There were 101 males and 44 females. The age distribution has been presented in Table 1. Table 2 summarizes the presenting clinical symptoms and the clinical signs.

Essential observations on radiological imaging are summarized in Table 3.

2.1. Surgical strategy

The aim of surgery was radical tumor resection. Although subtle nuances of surgery, in particular those related to tumor consistency, vascularity, morphological aggressiveness and brain reaction varied with the histological nature of the tumor, general surgical strategy and principles matched. Unilateral interhemispheric surgical approach was used in all patients. No preoperative shunt surgery or any kind of external CSF drainage was done prior to surgery. The side of larger extension of the tumor was selected as the side of approach. In general, the scalp incision was frontoparietal, centered on the coronal suture and extended across the midline. The conventional steps of interhemispheric transcallosal surgical approach have been elaborately discussed in the literature [6–9]. Avoiding or minimizing injury to the bridging veins forms a crucial technical issue. The retraction of the brain was first carried out in the region anterior to the coronal suture in order to minimize brain retraction related damage to the more critical cortex posterior to the coronal suture. Due to the large size of the tumor and the difficulty in exposing the corpus callosum and problems associated with dissection around the stretched anterior cerebral artery branches, in 10 cases a cortical incision was taken in the 'cingulate gyrus' away from the midline and was deepened vertically to traverse the 'tapetum' and ependymal layer to expose the tumor. Tapping of the loculated and enlarged ventricle was done prior to brain retraction to assist relaxation of the brain in 8 cases. The general surgical strategy was to relatively rapidly debulk the tumor, a process that was facilitated by the soft, necrotic and cystic tumor nature and loculated collections of ventricular CSF. Whilst debulking of the tumor within a relatively narrow surgical field was possible and safe, frequently encountered 'significant' bleeding affected the progress of tumor resection. The surgical strategy was to resect the bulk of the tumor and attempt to achieve hemostasis only after the major part of the tumor was resected and the surgical field was relaxed. Large sized veins that bled rather profusely were frequently encountered. Blood vessels arising from and in the vicinity of the choroid plexus were the frequent sites of bleeding and were encountered after the tumor bulk was removed. The microscope was appropriately angled to resect the tumor located anteriorly in the frontal horn and posteriorly in the atrium and occipital/temporal horn of the lateral ventricle and limit the extent of corpus callosal incision. All attempts were made to avoid cortical handling and damage, more particularly the cerebral cortex posterior to the coronal suture and to the brain around the confines of ventricle in general and in the vicinity of foramen of Monroe in particular. Radical tumor resection was attempted in all cases. Following hemostasis the wound was closed. No external ventricular drain was placed in the postoperative period.

Table	1

	able	showing	the	age	distribution	of	the	patients.
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Age (Years)	Number of patients
Less than 1 year	3
1–10	19
11–20	27
21-30	33
31-40	28
41-50	14
51-60	16
61–70	5

2.2. Post-operative care

In general, all patients were extubated following surgery and were kept under close neurological observation. An emergency CT scan was performed if the patient was found to have a post-operative neurological deficit or if the neurological status worsened in the post-operative period. After the acquisition of a mobile CT scanner in the year 2018, post-operative CT scan was always performed after about 8 h of surgery. Re-exploration for operative site clot was dictated by the neurological status of the patient. Wherever possible, routine post-operative MRI was performed within 48 h after surgery, at 3 months of follow up and then yearly thereafter (Figs. 1–3).

3. Results

3.1. Surgical outcome

The results of histological examination of the tumor are shown in the Table 4. The post-operative outcome has been outlined in Table 5 and is detailed below.

3.2. Postoperative clinical assessment

Six patients did not regain consciousness following surgery and died in the immediate post-operative period within 48 h of surgery. Three of these patients had developed a postoperative hematoma and were reoperated for clot removal. Four of these had a central neurocytoma, 1 had choroid plexus papilloma and 1 had an ependymoma. Re-exploration of the operated surgical site was done in additional 6 patients. These patients were conscious and alert after surgery but in the early postoperative period developed either sudden (3 patients) or progressive (3 patients) worsening in the neurological condition. Three of these patients deteriorated rapidly, became unconscious and had bilateral decerebration at the time of re-exploration. All these patients were identified to have a large clot in the operative site. Following re-exploration and removal of the clot, four out of these 6 patients improved in their neurological status and 2 patients succumbed. (5 patients had been operated for a central neurocytoma and 1 for a choroid plexus papilloma). Out of these four surviving patients, three improved completely at follow up of 3 months and had no neurological deficits. One patient who had bilateral decerebration prior to reoperation, gradually recovered in about 3-4 months to a state that he was independant and was able to perform all activities of daily living.

Nineteen patients had postoperative unilateral hemispheric deficit. They were all conscious and alert after extubation. Nine of these patients had hemiparesis/hemiplegia at the time of reversal from anesthesia. Ten patients developed hemiparesis and worsening in sensorium following a generalized seizure in the early postoperative period. Hemispheric deficit was partial in 13

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Table 2

Table showing the clinical features depending on histological subtype.

Presenting complaints	Central neurocytoma (N = 73)	SEGA (N = 23)	Ependymoma (N = 5)	PNET (N = 2)	Glioma (N = 21)	Choroid plexus papilloma (N = 20)	ATRT (N = 1)
Headache, vomiting	73	23	5	2	21	20	1
Convulsions	15	5	1	-	2	-	-
Visual complaints	7	3	2	-	2	-	-
Urinary incontinence	2	-	-	-	-	-	-
Altered behavior	-	-	1	1	8	12	1
Limb weakness	3	-	-	-	1	1	-

Table 3

Radiological differentiating factors amongst common intraventricular tumors.

Radiological characteristic	Central neurocytoma (Number of patients = 73)	SEGA (Number of patients = 23)	Ependymoma (Number of patients = 5)	Choroid plexus papilloma (Number of patients = 20)
Tumor Size				
4–5 cm	19	15	3	12
5–6 cm	24	8	2	8
6–7 cm	30	-	-	-
Calcification	5	4	-	-
Multiple small cystic change	71	5	-	-
Ventricular Involvement				
Univentricular	18	21	5	20
Biventricular	55	2	-	-

patients and complete in 6 patients. Five of these patients also had aphasia. Investigation in all these patients showed a small to moderate clot in the operative tumor cavity and cerebral cortical injury/ ischemia or infarct in the surgical trajectory. None of these patients needed a re-explorative surgery. Fourteen of these patients had been operated for a central neurocytoma, 1 patient for an ependymoma and 4 patients for a choroid plexus papilloma. At follow up of 3 months after surgery, 16 out of the 19 patients with post-operative limb weakness had recovered 'significantly' or completely. The other 3 patients recovered completely at follow up of one year. One hundred and fourteen patients were conscious, alert and neurologically intact and had no focal motor deficit at 48 h following surgery.

3.3. Extent of resection

The extent of resection was gauged by MRI scans performed at 3 months follow up following surgery. One patient with a central neurocytoma was found to have residual tumor on postoperative imaging performed within 48 h of surgery. This patient was re-explored and a radical excision of the tumor was performed on the third day of first surgery. Delayed postoperative imaging after at-least 3-months of surgery showed that the tumor was resected 'totally' in 122 out of the 137 surviving patients (61 out of 67 patients with neurocytoma, 23 out of 23 patients with SEGA, 4 out of 5 patients with ependymoma, 2 out of 2 patients with PNET, 15 out of 21 patients with glioma, in the 1 patient with ATRT and in 18 out of 20 patients with choroid plexus papilloma). In 15 cases there was at-least some residual tumor. Six patients with a central neurocytoma were found to have residual tumor on delayed post-operative imaging. In one of these patients a symptomatic growth of the tumor was observed at a follow up of 2 years and the patient was re-operated. The other five patients are being clinically observed. Long term follow up of 12 years was available in two of these patients and serial follow up imaging has not shown any growth of the residual tumor.

One patient with an ependymoma had symptomatic recurrence of the tumor and was re-operated. Two patients with choroid plexus papilloma who are asymptomatic and have relatively small residual tumor are currently under observation. Twenty-seven patients (21 patients with glioma, 2 patients with PNET, 1 patient with ATRT and 2 patients with ependymoma) were subjected to post-operative radiotherapy.

3.4. Delayed follow up

At one year after surgery, 137 patients were back to their active life. Follow-up after more than 3 years of surgery was possible in 95 cases. Five patients with high-grade gliomas, one patient with AT/RT and one patient with PNET succumbed to their illness in the follow up period. One patient with PNET had a prolonged 10year survival but ultimately succumbed to complications of diabetes.

4. Discussion

Large intraventricular tumors form a discrete subset and a relatively complex neurosurgical challenge. Slow progression of these essentially 'benign' or low grade malignant tumors and accommodative capacity of the ventricles make them unique in the sense that the symptoms are subtle and long standing and the tumors are significantly large at the time of diagnosis. The patients may not have any significant neurologic symptoms even when the tumor has reached a massive size. All patients had dull and generalized headache as initial presenting symptom, which increased in intensity and forced medical advise. With tumor growth, the additional symptoms of raised intracranial pressure in the form of vomiting and visual symptoms related to papilloedema become prominent. Focal, motor or generalized seizures as presenting symptoms were seen in 21 cases (14.5%). Sensory seizures occurred in 5 patients (3.4%). Ataxia as a prominent presenting feature was seen in 9 (6.2%) patients. Owing to the nature of growth pattern, some of these tumors may be detected incidentally or with only marginal symptoms. Considering the fact that postoperative deficits can be devastating, surgical decisions have to be taken carefully and keeping the patient and relatives under informed confidence. The general consensus is that other than radical tumor resection by



Fig. 1. Images of an 18 year old male patient with a central neurocytoma. a. T1-weighted MRI shows a large mixed intensity intraventricular tumor. The lesion extends across the midline. Both occipital and frontal horns are dilated. b. Coronal image of contrast enhanced MRI showing patchy enhancement. c. T-2 weighted MRI showing the mixed intensity lesion and necrotic/cystic areas within it. d. Postoperative MRI after 6 months of surgery showing tumor resection. e. Sagittal image of MRI showing tumor resection.

surgery, there is no other better or generally accepted treatment option.

Magnetic resonance imaging scan (MRI) is the primary tool for diagnosing intraventricular tumors. It differentiates intraventricular lesions from the paraventricular tumors. It helps in defining the extent of the tumor, its physical nature and its vascularity and thereby assists the surgeon in planning the surgical approach. Asymmetric hydrocephalus with ipsilateral dilatation of the temа



Fig. 2. Images of a 10 year old male child with a subependymal giant cell astrocytoma. a. Sagittal T1-weighted MRI showing the large intraventricular tumor. b. Coronal T2weighted MRI showing the mixed attenuation lesion. c. Axial T1-weighted MRI showing the tumor extensions. d. Postoperative image showing tumor resection. e. Postoperative axial image of T1-weighted MRI showing tumor resection.

poral and occipital horns is common and frequently 'diagnostic' of the intraventricular location of the tumor. [6] Calcification within the tumor was seen in 9 (6.2%) cases. Considering that in a large majority of cases the tumors are relatively benign and slow growing, outcome of a successful surgery can be 'curative' in nature. On the other hand, considering the location in the depth, high tumor vascularity and critical tumor adjoining relationships make surgery a formidable challenge and a possibility of complications that can lead to high morbidity or death. The techniques of tumor debulking and hemostasis have to be learnt and acquired over years of experience. Surgery on intraventricular large tumors has been associated with the range of morbidity and mortality that has been reported to be up to 45%. [1,2]

Transcallosal or transcortical approaches are the two possible surgical options [8,9].

We preferred to use the transcallosal approach. Retraction of the cerebrum off the falx for interhemispheric exposure is by itself a complex procedure considering that the brain is tense due to the presence of large tumor mass and its effects. Damage to the bridging veins has been identified to result in venous congestion related brain damage. Whenever the brain was not unusually tense and could be retracted off the falx safely, classically described surgical steps of an interhemispheric transcallosal approach were adopted. However, when the brain was tense and there were potential difficulties in brain retraction a modification of the transcallosal surgical route was adopted. Following an interhemispheric exposure cortical incision was taken in the region of the cingulate gyrus and cingulum and was deepened through the lateral aspect of corpus callosum and tapetum into the enlarged ventricle. This procedure allowed a rather quick exposure of the lateral ventricle and the tumor and possibility of drainage of CSF or tumor cyst to relax the brain. Dissection in the vicinity of the anterior cerebral artery and its branches is thus avoided. Tapping of the loculated and enlarged ventricle is also a useful maneuver to drain CSF and relax



Fig. 3. Images of a 23 year old male patient with a central neurocytoma. a. Axial image of contrast enhanced T1-weighted MRI showing the large intraventricular tumor. b. FLAIR image showing the large tumor. c. Contrast enhanced T1-weighted coronal MRI showing the large tumor. d. Postoperative MRI (after 1 year of surgery) showing tumor resection. e. Coronal MRI showing tumor resection.

the brain in selected situation. Such a procedure was in general avoided and was used only when the CSF filled ventricular loculation was large and retraction of the brain off the falx was wrought with significant possibility of cortical damage. A relatively hurried tumor debulking was performed; as it was observed that coagulation at each step to obtain a bloodless operative field was unnecessary. During surgery, identifying and maintaining a plane of demarcation between the tumor and the ependyma, choroid plexus and the surrounding white matter particularly around the region of foramen Monroe is of importance. Loss of the plane will essentially result in bleeding and injury to vital neural structures in the vicinity. The incidence of neurological deficits is high in such instances. Surgery has to be directed towards radical or total tumor resection as incomplete or partial tumor resection has been associated with tumor bleeding related issues. The ventricular cavity, particularly the regions of atrium and occipital horn have to be

Table 4

fable showing the	histological	diagnosis	of the	patients.
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Histological Diagnosis	Number of patients
Central Neurocytoma	73
SEGA	23
Choroid plexus papilloma	20
Ependymoma	5
Glioma	21
Pilocytic astrocytoma	9
Grade II astrocytoma	7
Garde IV astrocytoma	5
PNET	2
ATRT	1

Table 5

Table showing	the su	rgical out	come of	the j	patients.
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Post-operative Outcome	Number of patients	Percentage
	N = 145	
Immediate post-operative period		
No deficits	114	78.6%
Death	8	5.5%
Hemipareis	13	8.9%
Hemiplegia	6	4.1%
Aphasia	5	3.4%
Re-explored (for clot)	9	6.2%
Outcome at 3 months		
No deficits	133	91.7%
Persistent hemiparesis	4	2.8%
Outcome at 1 year		
No deficits	137	94.5%

appropriately explored by angulation of the microscope to confirm the radicality of tumor resection.

Recently, few authors have advocated the use of endoscope for resection of intraventricular tumors [10,11]. This modality of treatment was not used in our series.

Histological examination revealed 73 (50.3%) central neurocytomas, 20 (13.7%) choroid plexus papilloma, 5 (3.4%) ependymoma, 23 (15.8%) subependymal giant cell tumors, and 16 (11%) astrocytomas. In 8 (5.5%) cases, the tumor was 'malignant'; glioblastoma in 5 cases, primitive neuroectodermal tumor (PNET) in 2 cases and atypical teratoid rhabdoid tumor (ATRT) in 1 case. All these 8 patients received post-operative adjuvant radiotherapy. The need for radiation therapy and effect on overall long-term survival is a debated subject for central neurocytomas.

In a multicenter study with seventy-one patients, larger tumor volume, incomplete surgery, and a mitotic count >3 per 10 highpower fields were all predictors of a higher risk of recurrence [12]. However, despite many patients having tumors over 4 cm in size, overall survival was excellent (89% for all at five years), and particularly favorable for those who have a complete resection (96%). Sixty-one (91%) out of 67 patients with central neurocytoma in our series had a complete tumor resection and had a favorable outcome. Radiation was not given in any patient. Recently, some authors have used gamma knife as an upfront treatment after a biopsy or partial removal for giant central neurocytoma and noted reduction in the tumor volume. [13,14] In a meta-analysis of 438 patients with central neurocytoma, the tumor control rate at 10 years was 74% after gross total resection and 35% after subtotal resection [15]. The 10-year survival durations for total and subtotal resections were 99% and 82%, respectively. [15,16] Surgery remains the best option to achieve these goals.

There were 23 patients with SEGA. All the patients had associated tuberous sclerosis. Radical excision was achieved in all the patients without any morbidity or mortality. Our experience suggests that surgery is relatively straightforward and safe for these tumors even when they have attained large sizes and can be performed with low morbidity. The tumor in such cases is relatively avascular and better defined. Twenty patients had choroid plexus papillomas, and a majority of the patients were in the young age group. A gross total excision was achieved with a good overall outcome.

Intraventricular tumor surgery is wrought with significant risk of complications like hemiparesis, aphasia and risk of sudden deterioration due to cerebral venous infarction, ischemia related to compromise of anterior cerebral artery or post-operative hematoma. Patients operated with intraventricular tumors need vigilant monitoring following surgery to look for any worsening in consciousness. The indication of re-exploration for post-operative hematoma evacuation depends entirely on the clinical neurological status of the patient. However, re-exploration is generally favored if the size of the post-operative hematoma is equal to or more than the original size of the tumor. Some authors have identified cognitive impairment following interhemispheric surgical approach. However, psychological assessments and higher function evaluation was not diligently pursued in the present retrospective analysis.

A higher incidence of post-operative morbidity was seen in patients with central neurocytoma followed by choroid plexus papilloma in our series. This is primarily related to the large size and vascular nature of these two tumors. Nineteen patients (13%) had varying degrees of post-operative deficits and six patients (4%) needed a re-exploration for clot evacuation. Twenty-three of these patients had improved at follow up. None of the patients in our series required any CSF diversion procedure in the early or late post-operative period.

5. Conclusion

Surgery on large intraventricular tumors can be a challenging surgical endeavor. Successful surgical resection is associated with a long-term 'cure' from the disease.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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