Surgical considerations in the microsurgical management of lateral ventricle meningiomas: a report of 15 cases and review of literature

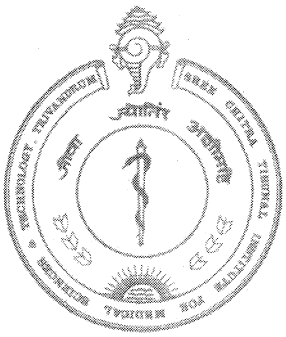
Submitted for MCh Neurosurgery

By

Dr. Bimal
September 2009

Department of Neurosurgery
Sree Chitra Tirunal Institute for Medical Sciences & Technology
Thiruvananthapuram – 695011

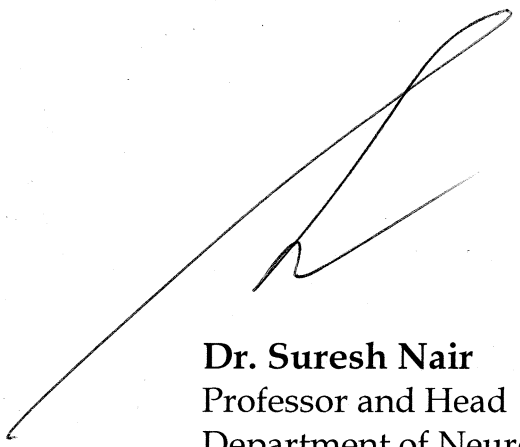
Surgical considerations in the microsurgical management of lateral ventricle meningiomas : a report of 15 cases and review of literature



Submitted by	:	**Dr. Bimal**
Programme	:	**MCh Neurosurgery**
Month & Year of submission	:	**October 2009**

CERTIFICATE

This is to certify that the thesis entitled "**Surgical considerations in the microsurgical management of lateral ventricle meningiomas : surgical issues : a report of 15 cases and review of literature**" is a bonafide work of Dr. Bimal and was conducted in the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram under my guidance and supervision.



Dr. Suresh Nair
Professor and Head
Department of Neurosurgery
SCTIMST, Thiruvananthapuram

DECLARATION

This thesis titled "**Surgical considerations in the microsurgical management of lateral ventricle meningiomas : surgical issues a report of 15 cases and review of literature**" is a consolidated report based on a bonafide study done by me during January 2007 to October 2009 under the Department of Neurosurgery, Sree Chitra Tirunal Institute for Medical Sciences & Technology, Thiruvananthapuram.

This thesis is submitted to SCTIMST in partial fulfillment of rules and regulations of MCh Neurosurgery examination.

Dr. Bimal
Department of Neurosurgery,
SCTIMST,
Thiruvananthapuram.

ACKNOWLEDGEMENT

The greatest force, the **Almighty God** is all powerful. He was with me always guiding and showing me the path of wisdom.

The guidance of **Dr. Suresh Nair**, Professor and Head of the Department of Neurosurgery, has been invaluable and I am extremely grateful and indebted for his contributions and suggestions, which were a valuable help during the entire work. He will always be a constant source of inspiration to me.

I owe a deep sense of gratitude to **Dr. Ravi Mohan Rao** for his invaluable advice, encouragement & guidance, without which this work would not have been possible.

The critical remarks and suggestions of **Dr. Girish Menon**, helped me in achieving a high standard of work.

I would also like to thank **Dr. Mathew Abraham, Dr. Easwer H V, Dr. Krishnakumar K, Dr. Gopalakrishnan C.V & Dr. Vikas** for their constant encouragement & support.

I would like to acknowledge the help that my colleagues, senior and junior, extended to me during the course of the study.

Dr. Bimal

CONTENTS

Part I

Part II

INTRODUCTION

Meningiomas arising within the ventricles without an obvious meningeal source are rare and constitute 0.5 to 5% of all intracranial meningiomas and 13-30% of all intraventricular tumours(2, 3, 7-10, 16, 27, 28, 32, 33, 42). The first description of an intraventricular meningioma was by Shah in 1854(32). In 1965, Delandsheer(9) summarized 175 cases of lateral ventricular meningiomas reported in literature until then, a data which was later updated by Crisculo(7) (400 cases) in 1986 and Nakamura(33) (532 cases) in 2003. These meningiomas pose considerable surgical challenge.

Meningiomas arising in the ventricular system are rare, present late when the tumour has grown to a considerable size and involve the dominant hemisphere more often. Surgical access almost always involves violation of normal eloquent cortex. They are vascular tumours, but vascular control is late to obtain during surgery. For all these reasons, they are tumors for which it is difficult to achieve the perfect surgical result: complete removal without complications or new neurologic morbidity. With a thorough understanding of the anatomy of structures around the ventricle, selection of the proper surgical approach, and use of modern neurosurgical techniques, good surgical results are possible. This study reviews some of the issues. We share our experience with fifteen such tumours and briefly discuss important clinical and technical considerations for the surgery of intraventricular meningiomas.

REVIEW OF LITERATURE

Epidemiology

Origin & anatomy

Clinical Features

Radiologic diagnosis

Pathology

Management

Recurrence

Surgical considerations

Prognosis

Epidemiology

Meningiomas are the second most common primary brain tumor in adults(8). In a surgical series reported by Cushing and Eisenhardt (N = 295)(8), intraventricular meningiomas accounted for only 1.3% of the total. Guidetti and Delfini(16) found that over a 38-year period, only 22 (1.5%) of 1451 meningiomas were intraventricular: 20 in the lateral ventricles, 2 in the fourth ventricle, and none in the third ventricle. In an earlier extensive review of the published literature up to 1986, Criscuolo and Symon(7) identified 400 intraventricular meningiomas and categorized their location. Eighty percent of intraventricular meningiomas occur in the lateral ventricles, 15% in the posterior third ventricle, and 5% in the fourth ventricle (Table 1). When a meningioma does occur in the lateral ventricle, it is more often on the left than on the right and more than 90% are located in the atrium. Within the third ventricle, meningiomas are more often posterior than anterior(32). The incidence of ventricular meningiomas is higher in pediatric patients.

Table 1 Common sites for intraventricular meningioma

Ventricle	Site	Frequency	Blood supply
Lateral atrium	>80%(L > R)	**80**	AChA/PLChA
Third	Posterior	**15**	PMChA/PLChA
Fourth	Midline	**5**	PICA

Abbreviations: AChA, anterior choroidal artery; L,left; PICA, posterior inferior cerebellar artery; PLChA, posterior lateral choroidal artery; PMChA, posterior medial choroidal artery; R, right.

Origin and anatomy

Meningiomas arise within the ventricle from the choroid plexus or from the tela choroidea within the ventricular system. As pointed out by Cushing and Eisenhardt(8), meningiomas within the ventricle tend to assume the shape of the ventricle in which they lay. The atrium is the most common location for lateral ventricular meningiomas. The atrium is formed by the junction of the temporal horn anteriorly and inferiorly, the occipital horn posteriorly, and the posterior body of the lateral ventricle anteriorly and superiorly. The floor of the atrium is formed by the hippocampus, the medial wall by the splenium of the corpus callosum, and the roof and lateral wall by the splenium and tapetum of the corpus callosum(32). Fibers of the geniculocalcarine tract run lateral and inferior to the atrium and account for visual symptoms seen with larger tumors (Fig. 1, 2). Meningiomas of the fourth ventricle arise from the choroid or the inferior tela choroidea(8).

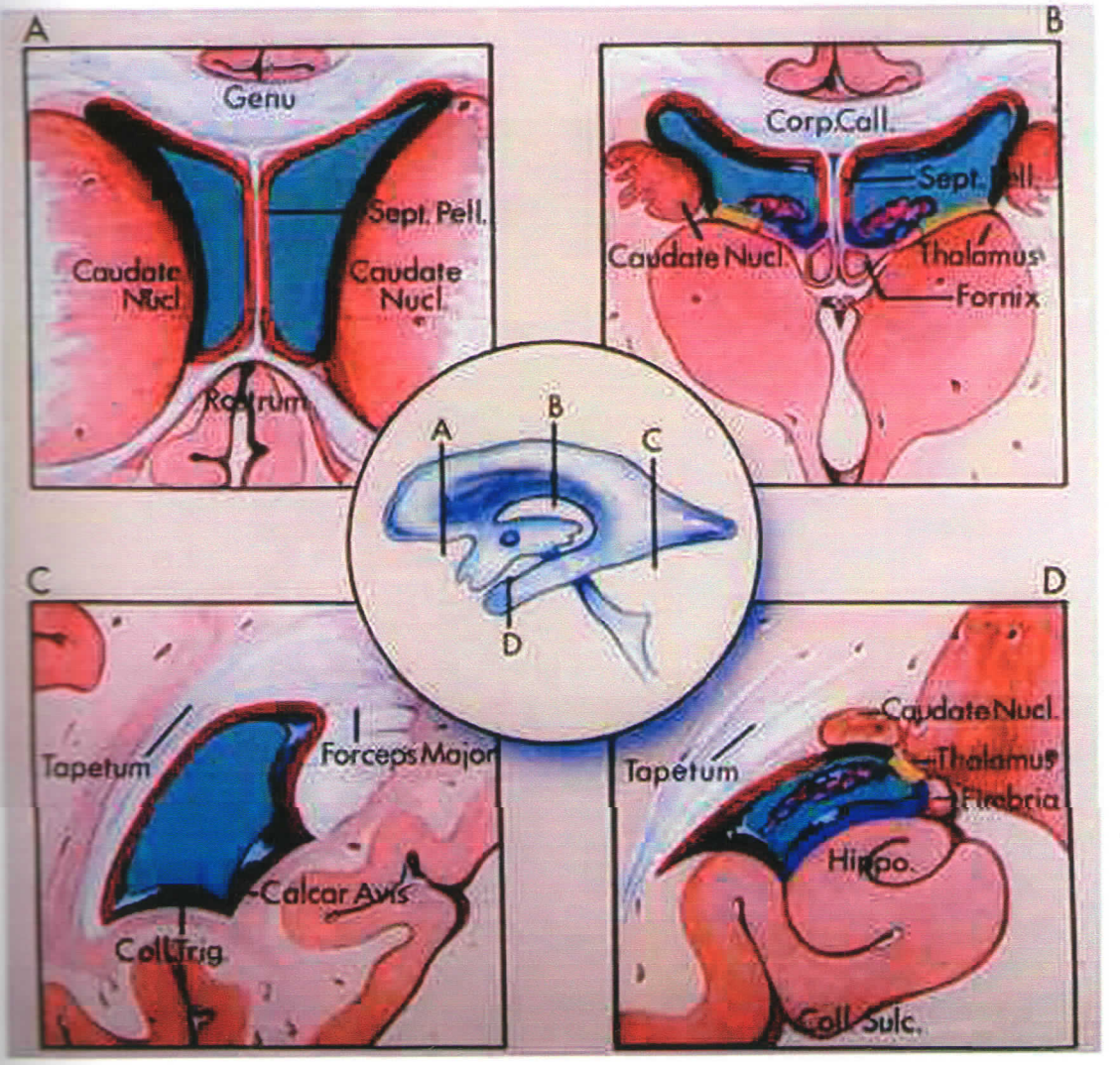


Figure 1 Relations of the lateral ventricle

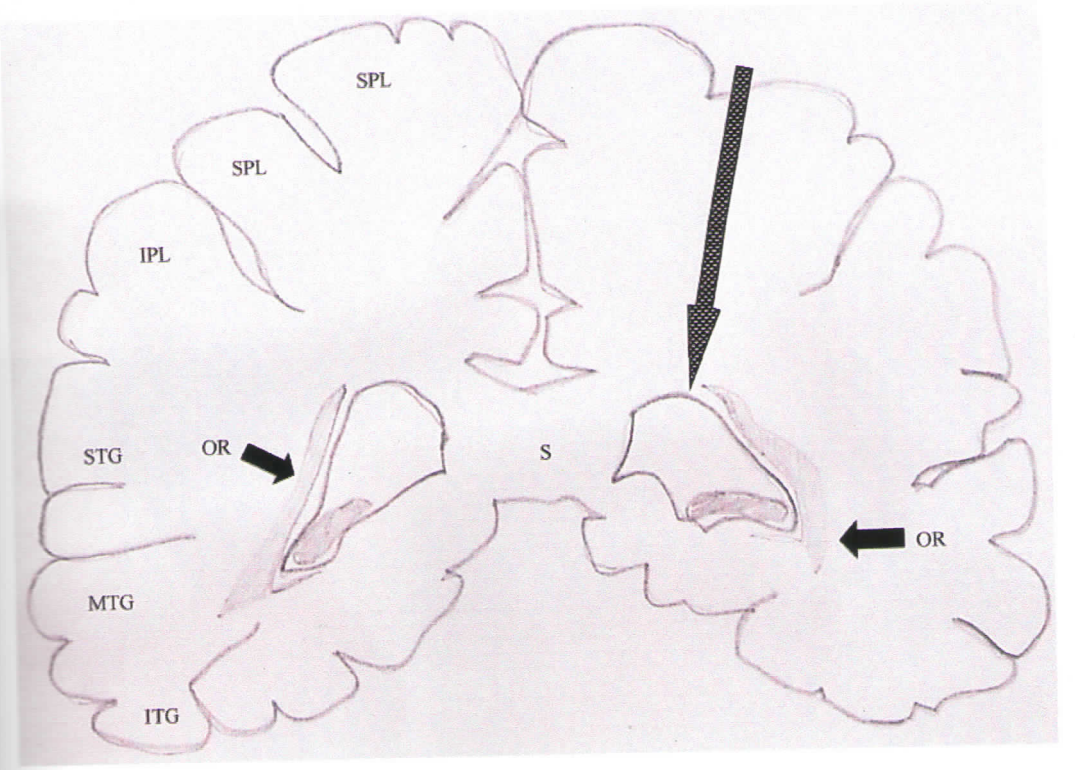


Figure 2 Posterior coronal section of hemispheres. Note position of optic radiations lateral and inferior to atrium. Large arrow on left indicates trajectory with posterior parietal-occipital approach. SPL, superior parietal lobule; IPL, inferior parietal lobule

Tumors of the posterior third ventricle are thought to arise from the tela of the velum interpositum, the space between the two layers of the tela choroidea in the roof of the third ventricle that carry the posterior medial choroidal arteries and internal cerebral veins(32). Tumors in this location are to be distinguished from meningiomas arising from the falcotentorial junction immediately behind the third ventricle.

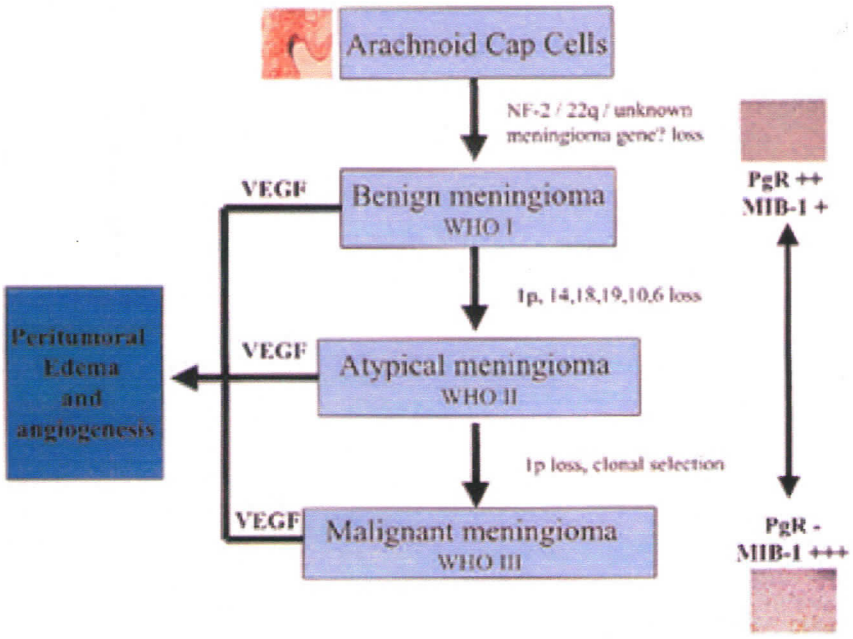


Figure 3 Proposed schema of meningioma tumour progression

Genetics and Molecular biology

The association between NF-2 and meningioma is well known, and they may share common mechanisms of pathogenesis. Thirty to eighty percent of sporadic meningiomas and nearly all neurofibromatosis-related meningiomas have mutations in the NF-2 gene located in chromosome band 22q12, that result in mutations in the protein *merlin*. Chromosomal banding techniques have identified chromosome subband 22q12.3-qter, which is near the NF-2 gene but is believed to represent a separate and distinct locus in meningioma formation. The genetic factors involved in the tumorigenesis of meningiomas are currently a subject of investigation to inform screening and prediction of risk for tumor progression to atypical or anaplastic disease. In Perry's

series(32, 42), sporadic and NF2-associated paediatric meningiomas were histopathologically similar with the exception that brain invasion was nearly exclusive to the sporadic tumours, a difference that reached statistical significance. Besides NF, Gorlin syndrome also known as multiple basal cell carcinomas is another familial tumour condition with autosomal dominant inheritance with an association with meningiomas(36).

Loss of expression of another tumour suppressor gene, DAL-1, which is located in 18p11.3, has been found in 30-70% of meningiomas and is thought to play a role in both early tumourigenesis and meningioma evolution. Other tumour suppressor genes implicated in the development or progression of meningiomas are SMARCB2 (22q11.2), p53 (17p), and CDKN2B (9p21). The fact that malignant and atypical meningiomas tend to have more chromosomal aberrations than benign tumours do, suggests progressive loss of tumour suppressors and potential activation of oncogenes. Some of the genes implicated in meningioma oncogenesis are c-*sis*, C-*myc*, Ha-*ras*, K-*ras*, c-*fos*, c-*erb*B and S6*k*. A variety of other chromosomal aberrations have been implicated in the formation and progression of meningiomas including losses on 1p, 2p, 6q, 10q and 14q and gains on 1q, 9q, 12q, 15q, 7q and 20. Alteration on chromosomes 1, 10, and 14 and reactivation of the telomerase subunit hTERT seem to be practically important in the progression of more biologically aggressive meningiomas. Radiation induced meningiomas have been shown to express genetic alterations that are different than those of sporadic meningiomas. In particular, there are fewer losses of genetic

material on chromosome 22 and more losses on chromosomes 1p, 6q, 9q, 18q and 19q.

Gonadal steroid hormones and receptors

Estrogen receptors have been reported in 0-94% of meningiomas and progesterone receptors in 40-100%. Recent studies using modern experimental and laboratory techniques have revealed minimal amounts of functional estrogen receptor. This finding is supported by the generally disappointing results of anti-estrogen agents (Tamoxifen and Mepitiostane) in treating meningiomas. Most investigators have identified high levels of progesterone receptors in meningiomas, and the presence of these receptors has correlated with less aggressive tumour biology, more favourable prognosis and a lower incidence of recurrence. Antiprogesterone agents used to treat meningiomas have yielded varied results; the most recent phase-III double-blind, randomized, placebo-controlled trial of mifepristone reported no significant benefit(15). Though one might not expect paediatric meningiomas to be hormonally driven, progesterone receptor (PR) is expressed with similar frequency, regardless of age at presentation. There is a roughly inverse association between PR expression and tumour grade in meningiomas of children and adults alike(30).

Androgen receptors are found in meningiomas with about the same frequency as progesterone receptors and are expressed in 69% of males and 31% of females. Testosterone stimulates in vitro meningioma cell growth, and it has

been speculated that androgen receptors may help modulate progesterone receptor activity.

Other receptors and Growth Factors

Using polymerase chain reaction analysis. Carrol et al (1996)(5) detected D_1 receptor mRNA in meningiomas, particularly in females, as well as D_2 receptor mRNA and prolactin receptor mRNA, but the functional importance of these findings is unclear. Somatostatin receptors, particularly type 2a (hsst2a) receptors have also been reported at high levels in meningiomas. There have been a few reports of success using somatostatin analogues to treat meningiomas, but the role of somatostatin receptors in tumour progression or growth is still unclear. Growth hormone receptor mRNA is ubiquitiously expressed in meningiomas. Growth hormone receptor blockade by pegvisomant has been shown to result in decreased growth rates of primary meningioma cell cultures and reduced tumour growth and regression in an in vivo animal model.

Westphal and Hermann(43) (1986) discovered functionally intact epidermal growth factor (EGF) receptors, a product of the oncogene c-*erb,* and reported increased DNA synthesis after EGF treatment of meningioma cell cultures. Weisman et al (1986) noted a modulatory effect on this receptor by platelet derived growth factor (PDGF) and revealed near maximal levels of DNA synthesis in meningioma cell cultures when PDGF and EGF were added together(3, 32).

The finding of c-erb/EGF receptor expression in meningiomas prompted searches for other oncogene receptor- mitogen systems. Using Northern-blot analysis, Maxwell et al 1990(31) demonstrated that meningiomas express both the c-sys/ PDGF-2 proto-oncogene and the PDGF receptor (PDGFR) gene. Further studies revealed that PDGF-β is expressed in meningiomas, that PDGF-BB increases c-fos expression in meningioma cell cultures, and that over expression of PDGF- β and PDGF-BB is associated with higher grade and proliferative activity in meningiomas. These results support the concept of PDGFR activation by an autocrine-paracrine loop and the idea that PDGFR activation contributes to tumour cell proliferation or malignant transformation. Vascular endothelial growth factor (VEGF) levels are associated with increased angiogenesis, edema, and frequency of recurrence in meningiomas. Fibroblast growth factor and insulin-like growth factor 1 have also been identified in meningiomas and implicated in tumour progression. Many of the growth factor receptors (PDGFR, EGFR and VEGFR) are protein tyrosine-kinase receptors that activated ras and associated intracellular cascades, which mediate cellular proliferation, differentiation and transformation.

Clinical Features

Meningiomas of the lateral ventricles present primarily with signs of increased intracranial pressure. Headache, nausea/vomiting, and disturbance of vision are seen in 40% to 80% of patients (Table 2)(9). Visual symptoms most often relate to impaired vision from papilledema but can also include

visual field deficits from large atrial tumors. Motor, sensory, and speech disturbances are also seen, and seizures were the presenting symptom in 27% of patients for one series(9). Symptoms of impaired memory may relate to dilatation of a trapped temporal horn or to direct compression on the hippocampal formation in the floor of the atrium with larger tumors. Tumors of the third and fourth ventricles usually present with symptoms of hydrocephalus as a result of obstruction of cerebrospinal fluid (CSF) flow. Parinaud's syndrome with impaired upgaze and pupillary light reflexes may be seen with posterior third ventricular masses(32).

Table 2 Common symptoms and signs for atrial meningiomas

Symptoms	Frequency
Headaches	80%
Nausea/vomiting	40%
Seizures	35%
Speech disturbance	30%
Motor	25%
Mental disturbance	20%
Visual disturbance	20%
Sensory	15%
Signs	
Papilledema	60%
Visual field deficit	50%
Motor disturbance	50%
Dysphasia	40%
Sensory disturbance	20%

From Guidetti B, Delfini R. Lateral and fourth ventricle meningiomas: In: Al-Mefty O, editor. Meningiomas. New York: Raven Press; 1991. p. 569–81;

Radiologic diagnosis

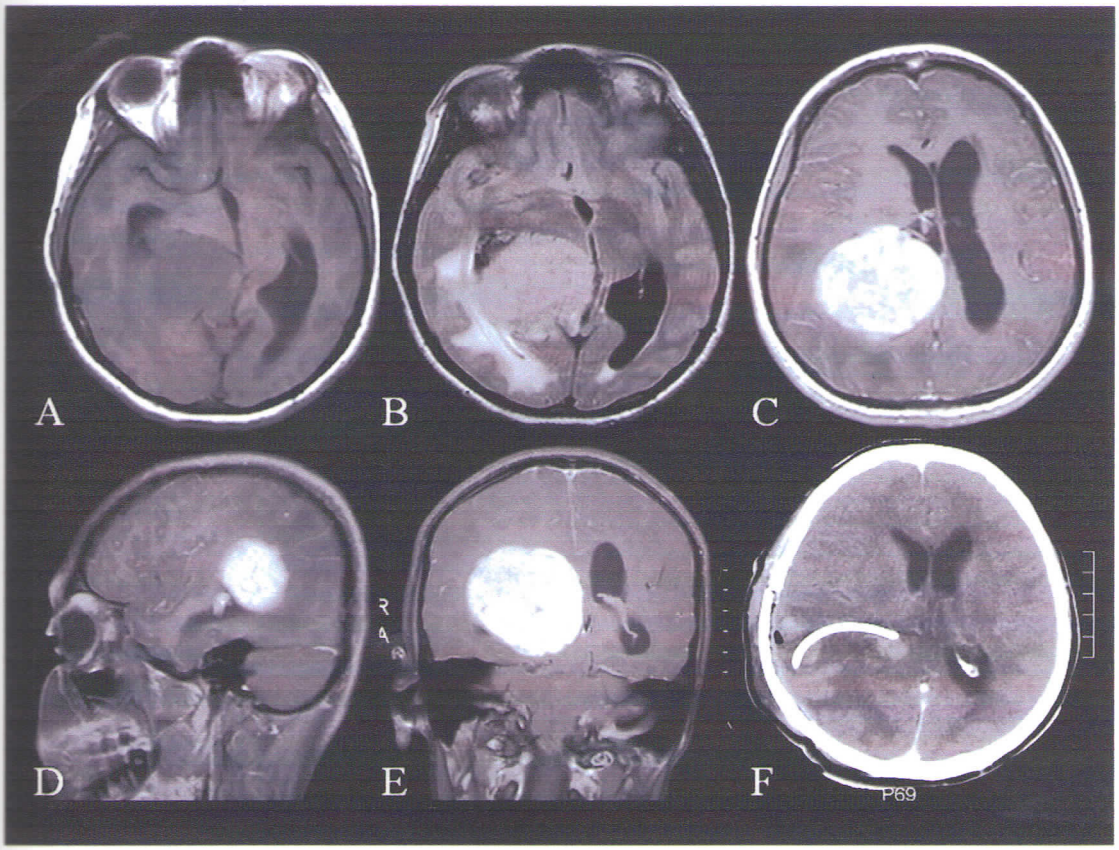


Figure 3 A- E : preoperative MR images of a patient with an intraventricular meningioma located in the right trigone. A. T1 axial image showing a well circumscribed isointense intraventricular lesion; B the same lesion inhomogenously hyperintense on flair sequence ;C Axial contrast; D sagittal contrast; E coronal contrast showing brilliant contrast enhancement F Post operative CT scan showing total tumour scission with an intraventricular catheter drain insitu

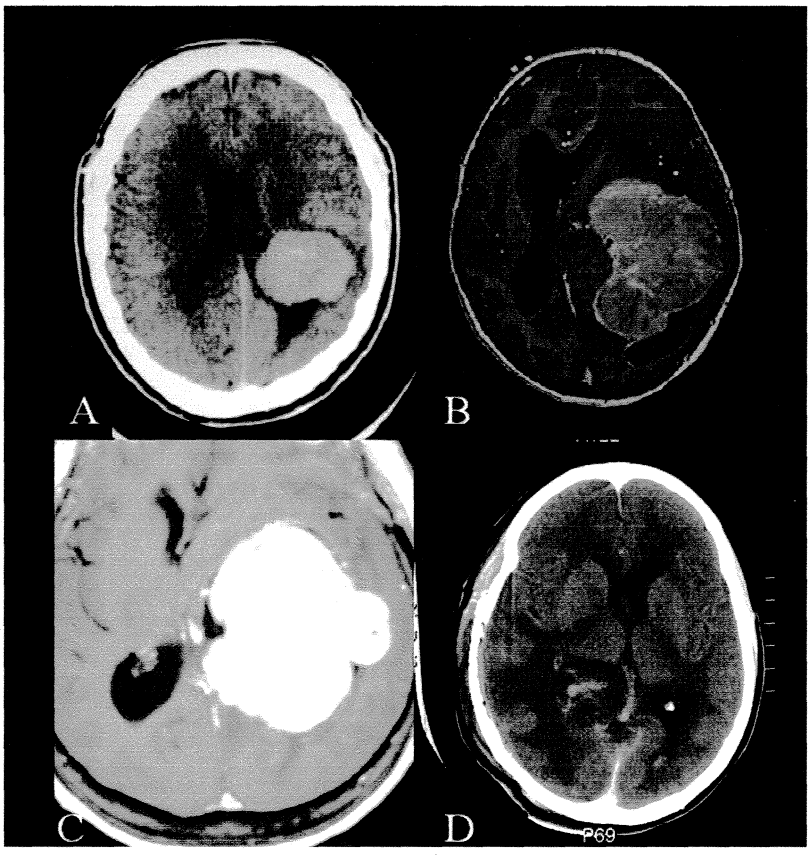


Figure 4:A- D pre an post operative images of a patient with a left trigonal meningioma. A. Axial contrast image showing uniform contrast enhancement B Axial MR images showing an iso intense lobulated lesion C. Axial contrast MR sequence showing intense contrast enhancement D. Postoperative CT scan showing total tumour excision

On Computerised tomography scans, meningiomas in the lateral ventricle are slightly hyperdense, may have small areas of calcification, and usually show homogeneous enhancement(3). With larger tumors, obstruction of the temporal horn, which contains the choroid plexus, results in dilatation of this portion of the ventricle and low density in the brain surrounding the atrium, which is partially related to transependymal flow of CSF and partially related to tumor-associated vasogenic edema (Fig. 3). MRI reveals superior anatomic detail compared with CT scans, with meningiomas being iso- or hypointense

on T1-weighted images and T2-weighted images(2). On T1- weighted postcontrast images, there is uniform contrast enhancement. Thin-cut axial spoiled gradient recall (SPGR) images are routinely used for image-guided surgical systems, and these images can also provide the opportunity for creating two-dimensional (2D) and three-dimensional (3D) venograms to assist the surgeon with the preoperative selection of surgical approaches(42). MRI also offers the ability to perform spectroscopy and blood volume time intensity maps, which, alone or together, may increase the certainty of the radiologic diagnosis. A high alanine-to-creatinine ratio has been reported as a relatively specific MR spectroscopy finding for meningiomas(32). Cerebral angiography is rarely used these days, because intraventricular meningiomas can rarely be embolized. Angiography can confirm the predominant blood supply (Table 1) and the position of prominent parasagittal draining veins(32). Catheter cannulation of choroidal arteries is difficult, however, and the target for occlusion is distal in the vascular territory. Thus, in selecting an operative approach, the surgeon may need to take into consideration the potential advantage of occluding the arterial blood supply to large tumors early in the operation.

The Magnetic Resonance Imaging (MRI) characteristics of meningiomas are generally consistent. On T1 weighted images, 60-90% of meningiomas are isointense and the remainder are mildly hypointense compared to grey matter. On T2 weighted images, 30-45% of meningiomas have increased signal intensity and approximately 50% are isointense compared to grey matter. Their typical extraparenchymal location heightens the

neuroradiologist's ability to diagnose these tumours. There is increased interest in using MRI characteristics to subtype meningioma tissue before surgery. The results of these studies have been variable. Some have reported 75-96% accuracy whereas others have found no correlation. The MRI characteristics that allowed accurate preoperative identification of meningioma subtypes were confined to findings on T2 weighted images. Specifically, meningiothelial and angioblastic variants had higher signal intensity on T2 weighted sequences than fibroblastic and transitional meningiomas did. The amount of cerebral edema associated with meningiomas was also found to be greater in meningiothelial or angioblastic variants. Finally, a high signal intensity on T2 weighted images has been correlated with microscopic hypervascularity and soft tumour consistency.

Contrast enhanced MRI provides the most sensitive and specific means of detecting meningiomas. Most meningiomas enhance intensely and homogenously with intravenous contrast material, and in 10% cases, small additional meningiomas are encountered that are missed on un-enhanced MR images. Contrast enhancement of the dura mater extending away from the margins of the mass is typical of meningiomas, although this pattern can be seen with other dural based lesions. This dural tail can represent either tumour extension or reactive change, and its resection is important to reduce the risk of recurrence. Post operative enhanced MRI has also been found to be sensitive and specific in detecting residual or recurrent meningiomas. Thick and nodular enhancement has a high correlation with recurrent or residual neoplasm.

In vivo, MR spectroscopy is an evolving area of study. Compared with the MR spectra of normal brain, the typical MR spectra for meningiomas reveal a markedly increased choline peak and reduced N-acetyl aspartate and phosphocreatine/ creatine peaks. An additional peak present in some meningiomas at 1.47ppm has been attributed to alanine.

The diagnosis of various lesions can based on the site of occurrence and age group of involvement.

Common radiographic differential diagnoses for intraventricular meningioma

Site	Child	Adult
Atrium	CPP Ependymoma	Metastasis Ependymoma
Posterior third ventricle	Germinoma Pineocytoma blastoma Astrocytoma Teratoma	Pineocytoma Astrocytoma Metastasis
Fourth ventricle	CPP Ependymoma Astrocytoma Medulloblastoma	Ependymoma CPP Hemangioblastoma Medulloblastoma

Pathology

Microscopically meningiomas have a varied but characteristic histopathologic appearance. This diversity forms the basis for their pathologic classification. The system based on the 2000 WHO classification which associates histopathology with information on recurrence and aggressiveness is used in this study. There are three grades of meningiomas. Grade I meningiomas are associated with a low risk of recurrence and aggressive growth, whereas

Grade II and III meningiomas have a greater likelihood of one or both of these characteristics.

Grade I meningiomas:

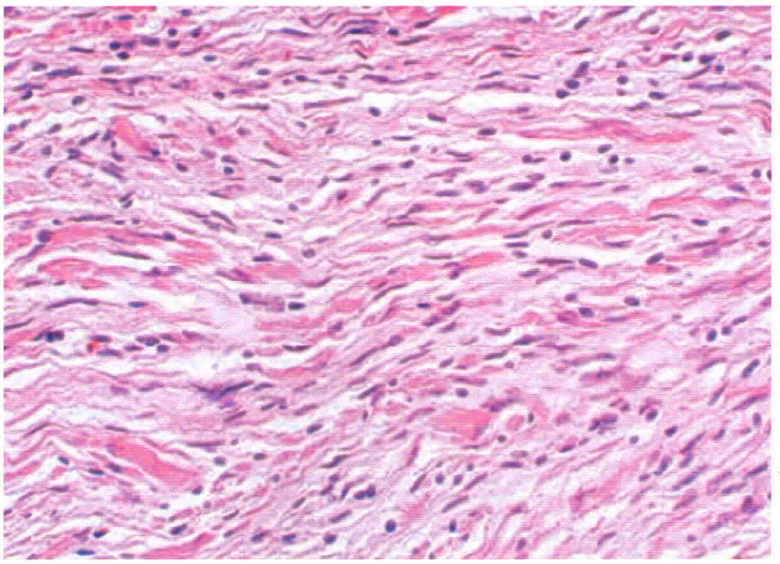


Figure 4 Fibroblastic meningioma with parallel fascicles of fibroblastic cells

Of the nine subtypes of Grade I meningiomas, the three most common are meningiothelial, fibrous and transitional. Although it is important that these and the other subtypes are recognized, the prognostic significance of each one is unclear. But they are currently considered equivalent.

Grade II meningiomas:

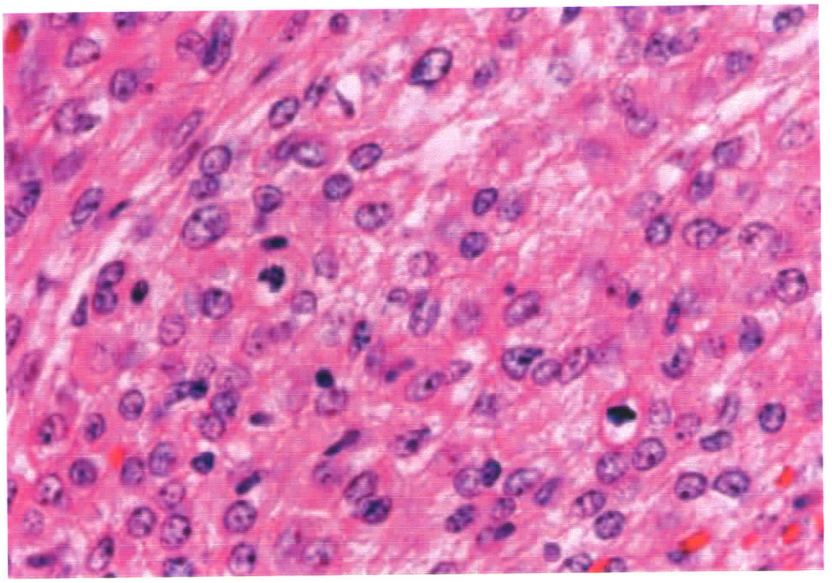


Figure 5 Atypical meningioma with multiple mitosis

Apart from brain invasion and metastatic spread which define malignancy, certain features of Grade II meningiomas that may be seen by light microscopy suggest increased tumour aggressiveness and and increased recurrence rate. Among these atypical features, are loss of architectural pattern, high cellularity, increased number of mitotic figures (>4 mitoses per 20hpf), necrosis, prominent nucleoli and nuclear pleomorphism. The three subtypes of grade II meningiomas are atypical, chordoid and clear-cell type.

Grade III meningiomas:

The diagnosis of a grade III (malignant) meningioma traditionally requires histologic evidence of brain invasion or distant metastasis which in most cases is accompanied by further evidence of biologic aggressiveness such as cellular sheeting, nuclear pleomorphism, increased cellularity and mitoses

(>20mitoses per 20hpf), and necrosis. When dissemination occurs, the most common location for metastasis are the lungs and pleura, abdominal viscera (especially the liver), lymphnodes and bones. Patients with meningiomas associated with frank malignancy are reported to have only a 2-year median survival duration. The three subtypes of grade III meningiomas are anaplastic, papillary and rhabdoid.

In the series of Guidetti and Delfini(16), 81% of the lateral ventricle meningiomas were fibroblastic. Meningotheliomatous and psammomatous variants reported by others as case reports are less common. All three types are classified as grade I tumors in the World Health Organization scheme, with a low risk of recurrence and nonaggressive clinical behavior(27, 32). The pathologic entities encountered in the lateral ventricle also include choroid plexus papilloma, ependymoma, and metastases. Posterior third ventricular tumors that may mimic meningiomas radiographically include pineocytoma and teratoma, whereas choroid plexus papilloma, ependymoma, and hemangioblastoma should be considered in the fourth ventricle.

Management

The treatment of a meningioma depends primarily on the size and location of the tumour, the age of the patient, the associated symptoms and neurologic deficits. The mainstay of treatment is surgical resection, although small, asymptomatic, incidental meningiomas can typically be managed with observation and serial imaging. After surgery, re-imaging is typically performed at 6 month intervals initially, which may be extended to longer

intervals if there are no radiographic signs of tumour growth and the patient remains asymptomatic. Treatment should be initiated when symptoms arise or tumour growth is documented. Critical parameters that affect the ease of surgical removal include the tumour's location, size and consistency, vascular and neural involvement and in case of recurrence, prior surgery or radiotherapy. New and innovative approaches have been devised to reach and widely expose meningiomas in any location. Furthermore, a greater appreciation of risk factors for and patterns of tumour recurrence has changed surgical planning and goals. The surgical goal now is to decrease the incidence of recurrence by resecting all of the neoplasm and all involved dura mater, soft tissue and bone. However, the tumour size and location and the involvement of adjacent structures may not allow all meningiomas to be completely resected in this manner.

Observation

Intraventricular meningiomas discovered incidentally, those that are not causing obstruction of CSF flow or hydrocephalus, and those not associated with vasogenic edema should be observed(12, 32). Mcdermot(32) usually follows patients with two successive 3-month scans; if the tumor is stable, the interval is increased to 6 months for 2 years and then to once a year. It is important to compare follow-up scans with the original scan and not the one immediately preceding it, because smaller changes in size may not be appreciated. Anterior-posterior, lateral, and vertical dimensions should be recorded. Documented tumor growth on serial scans in a medically fit

individual or development of symptoms related to secondary tumor effects, such as hydrocephalus or vasogenic edema, may warrant surgical intervention. Certainly, for a nondominant right atrial meningioma with symptomatic trapping of the temporal horn, surgical removal of the tumor is preferred over shunting and radiotherapy. In contrast, a slowly growing meningioma in the left atrium of a 70- year-old patient that is not causing hydrocephalus or vasogenic edema may be considered for radiosurgical treatment.

Surgery

Surgery is the gold standard of treatment for growing symptomatic meningiomas in patients who are candidates for general anesthesia. Because most of these tumors are benign and complete excision of lateral ventricle tumors can be accomplished, surgical cures can be achieved. Potential side effects of the various surgical approaches need to be considered and reviewed with the patient(28).

Radiosurgery

Radiosurgery is an effective form of treatment for selected meningiomas, including intraventricular meningiomas. Reports on the tumor control rates achieved with radiosurgery of 85% to 98% cannot be overlooked during the informed consent process(23). A size limit of 3 cm as used in other brain locations is not acceptable for intraventricular tumors of the atrium, third ventricle, or fourth ventricle Mcdermott(32) usually limits tumors to 1.5 to 2.0

cm depending on associated vasogenic edema. Of the 13 patients treated at Mcdermott's institution, 4 have had radiosurgery for their intraventricular meningioma. In 2 patients, the treatment was used for residual or recurrent disease, and in 2 patients, it was used as the primary form of therapy. Of these latter 2 patients, 1 developed symptomatic subependymal radiation toxicity despite a low marginal dose of 12 Gy. In spite of the fact that the tumor decreased in size, obstructive hydrocephalus with a trapped temporal horn developed and surgical removal was performed 20 months later. Mcdermot reports seeing exaggerated radiation toxicity in the subependymal periventricular region as well as with fractionated radiation therapy; thus, even "noninvasive" treatments should be recommended only to those who are not candidates for, or refuse, microsurgical removal(32).

Radiotherapy

Although there have been no randomized, controlled or prospective studies with long-term followup conducted to evaluate the efficacy of radiotherapy in treating meningiomas, the use of external beam irradiation has become an important part of the management of these tumours, particularly as adjuvant treatment for patients after subtotal tumour resection. In a prospective analysis of 140 patients with benign meningiomas treated by subtotal resection plus adjuvant radiotherapy over 23 years, Goldsmith et al 1994(32) reported 5 and 10-yr progression-free survival rates of 89 and 77 % respectively. In patients treated using CT planning (after 1980), the 5-yr progression-free survival rate was 98%. Recently, Soyuer et al (38) (2004)

compared 92 patients with WHO grade I benign cerebral meningiomas who underwent gross total resection, subtotal resection plus adjuvant radiotherapy or subtotal resection plus delayed radiotherapy.

At a median followup of 7.7yrs, the 5-yr progression-free survival rates were 77%, 91% and 38% respectively. The overall 5 and 10-yr survival rates were not statistically different among the three groups or from the age-adjusted expected survival rate. Thus delaying radiotherapy until tumour recurrence without compromising overall patient survival is possible and may spare the patient from the potential toxicity of radiation. The data do not permit determination of which strategy is optimal. For meningiomas that are considered inoperable because of their location, poor patient health, patient refusal of surgery, external beam radiotherapy would seem beneficial for aggressive (atypical or anaplastic) tumours, but very little information exists to support this theory.

Stereotactic irradiation in the form of radiosurgery or conformal, fractionated or intensity modulated radiotherapy has increasingly been used to treat meningiomas with improved efficacy and diminishing untoward effects. Stereotactic irradiation uses various forms of energy, the most common of which are photons from Cobalt-60 gamma-ray sources (gamma-knife) or linear accelerators (LINAC) and heavy particles (protons) from cyclotrons. Radiosurgery provides effective tumour control of small meningiomas. Kondziolka et a(24) 1999 observed a 93% tumour control rate in patients whose meningiomas were treated by gamma-knife radiosurgery and a 61%

incidence of tumour shrinkage in 99 patients who were followed for 5-10yrs. The incidence of new neurological deficits in this group of patients was 5%. In a recent retrospective study, Pollock et al 2003(34) reported that gamma-knife radiosurgery of small or medium sized beingn meningiomas, provided progression-free survival rates equivalent to that of complete surgical resection after a mean follow-up of 64months. Gamma-knife radiosurgery has also been shown to be an effective treatment for difficult to resect cavernous sinus meningiomas. Lee et al 2002(26), reported an actuarial tumour control rate of 93% at five years for benign cavernous sinus meningiomas; adverse effects of radiation were experienced by 6.7% of patients.

LINAC based radiosurgery also offers effective control of small meningiomas. A recent study of 43 patients who underwent LINAC based radiosurgery for skull-base meningiomas, reported a 7-year local control rate of 89.7%(6). This value correlated with the 5-yr control rate of 89% and a complication rate of 55 in a previous study of 127 patients. Spiegelmann et al(39) 2002 reported that both the 3- and 7-yr actuarial tumour growth control rates were 97% for cavernous sinus meningiomas treated by LINAC based radiosurgery. They also reported a low incidence of lonterm cranial neuropathies.

Despite the promising results of stereotactic irradiation, there are some limitations and uncertainities with this modality. The targeted tumour is limited to 35-40mm because this is the size at which the tumour can receive a single dose of appropriate strength with a 1% risk of radiation necrosis. However, the increased availability and use of fractionated delivery of stereotactic

irradiation have overcome this size limitation. Alheit et al(1) 1999 reported a 1-yr progression-free survival rate of 100% in 24 patients who underwent fractionated stereotactically guided conformal radiotherapy for meningiomas. Seven of fifteen patients who had neurologic deficits before treatment improved, and two patients experienced early side-effects (one facial palsy and one Addisonian state). Other recent studies have reported benefit from stereotactic conformal radiotherapy for atypical and malignant meningiomas and for large cavernous sinus meningiomas.

Intensity modulated radiotherapy delivers fractionaled, conformal radiotherapy more effectively than traditional techniques to tumours with complex shapes. Initial reports have shown that this method is effective in treating meningiomas and controlling tumour growth and carries a low risk of side-effects.

Adjuvant radiotherapy appears to be beneficial after incomplete excision of meningiomas in adults, but it is rather risky to use radiotherapy for benign and partially excised cerebral lesions during childhood. Re-operation is thought to be better than adjuvant therapy. Tumour behaviour following resection was difficult to predict and paediatric patients with histologically benign meningiomas deserve careful and extended clinical follow-up(12, 17). Atypical meningiomas are known to have long survival and benign ones are known to recur fast .

Fractionated 3D conformal radiotherapy offers effective tumor control for residual or recurrent meningiomas in a variety of intracranial locations(12).

Although there is no reason to believe that the results should be any different for intraventricular meningiomas, none of the four patients at Mcdermots institution who required additional therapy other than initial surgery (ie, reoperation, radiosurgery) was treated with this method. Although the conformality of present-day techniques limits the volume of normal tissue irradiated, intraventricular meningiomas are such discrete targets that surgery or radiosurgery has been recommended instead.

Adjuvant chemotherapy

Little information is available regarding the efficacy of traditional antineoplastic agents against benign or malignant meningiomas. Adjuvant chemotherapy (intravenous or intra-arterial cis-platinum, dacarbazine or doxorubicin) against malignant meningiomas and for recurrent benign or atypical meningiomas has been administered to a small number of paients but has generally been unsuccessful despite its effectiveness against other soft tissue tumours. Hydroxyurea has been shown to arrest meningioma cells in the S-phase of cell cycle and to induce apoptosis in vitro. Although a similar beneficial effect has been seen in a small subgroup of patients with recurrent and unresectable meningiomas, subsequent studies have shown little, if any, benefit. Interferon-α has been reported to be effective in prolonging the time to recurrence in a small group of patients with aggressive meningiomas and to have a lower toxicity than traditional chemotherapeutic agents. The South west Oncology group used Tamoxifen to treat 19 patients with unresectable or refractory meningiomas and observed tumour progression in 10 patients,

temporary stabilization of the disease process in 6 patients, and a partial or minor response in 3 patients(12, 14). A recent phase III, double-blind, randomized study of Mifepristone did not show any benefit(15).

Recurrence

The completeness of the tumour resection is the primary factor influencing the meningioma recurrence rate. Stafford et al(40) found a 25% recurrence rate at 10years in patients who had undergone a gross total tumour resection and a 61% recurrence rate in those who had undergone partial resection. Jääskeläinen (Nov 1986)(18) found an overall recurrence rate at 20years of 19%. Multivariate analysis showed that strong risk factors for recurrence included no coagulation of dural origin, invasion of bone and soft consistency of the tumour. The recurrence rate at 20years was 11% for patients with none of these risk factors, 15-24% for one risk factor and 34-56% for two risk factors. In a second study from the same group, the diagnosis of atypical and anaplastic meningioma carried an increased risk of recurrence of 38 and 78% at 5years respectively. The fact that cumulative relative survival rates (that is the observed to expected survival rates) at 1, 5, 10 and 15 years were 83%, 79%, 74% and 71% respectively, indicated increased mortality in patients with meningiomas. Using multivariate analysis, Stafford et al 1998(40) found that age younger than 40years, male sex, incomplete surgical resection, four or more mitotic figures per ten high power fields were associated with a decreased progression-free survival rate. Other factors that have been implicated in the recurrence of meningiomas include mitosis, focal necrosis,

brain invasion, syncitial tumours, hypervascularity, haemosiderin deposition, sheets of tumour cells, prominent nucleoli, nuclear pleomorphism and elevated proliferation index.

The use of Ki-67 labelling to develop a proliferation index is a common immunocytochemical technique for predicting a tumour's biologic aggressiveness and potential for recurrence. Labeling indicates averaging 1%, 5.5% and 12% have been identified for benign, atypical and anaplastic meningiomas respectively. The median MIB-1labelling index for pediatric meningiomas without histological atypia did not differ from that for adult meningiomas without atypia, in a study of 14 paediatric meningiomas by David Sandberg et al(37), suggesting that the more aggressive clinical features of meningiomas in children may be attributable to factors other than the rate of cellular proliferation.

Other markers of proliferation currently being investigated are progesterone receptors, topoisomerase II α, telomerase, transforming growth factors, mitosin, survivin, and other apoptosis related proteins. Positron emission tomography of glucose utilization have also been used to assess a tumour's biologic aggressiveness and potential for recurrence.

Surgical considerations

Evaluation of the patient for surgery involves consideration of patient factors, such as age, medical conditions, and neurologic status, and tumor characteristics, such as relation to symptoms and signs, growth rate, and

resectability(32). The routine evaluations for patients and specific discussion of the approaches for intraventricular meningiomas are reviewed below.

Preoperative studies

MRI without and with contrast is the basic imaging study needed. For all supratentorial approaches to the lateral ventricles, we obtain volumetric, thin-cut, T1-weighted images for use with image-guided surgical systems. MR venography can also be obtained to look at collapsed vertex views or 3D reformats. Cerebral angiography is not routinely done, because the blood supply to tumors in various locations is known and embolization is not possible without significant risk. For patients having a transcallosal procedure or transcortical superior parietal lobule approach in the dominant hemisphere, the author always obtains preoperative neuropsychologic testing as a baseline. This can be repeated 3 months after surgery and helps with quantitating patient status for return to work and other disability issues. It can also be used to document improvement in function for patients who undergo successful resection of large tumors or those associated with hydrocephalus. If a superior parietal occipital approach is selected, most anesthesiologists want the patient to have a echocardiogram done to rule out a patent foramen ovale and a potential right-to-left shunt. Humphrey visual fields should be tested in all patients with tumors of the atrium.

General intraoperative measures

Patients undergoing transcortical approaches receive anticonvulsants for 1 week around the operation, beginning the morning of surgery. Intravenous fosphenytoin or phenytoin is used to load those patients who are not already on medications. Standard measures to reduce intracranial pressure and improve blood rheology are used. Mean arterial blood pressure should be kept in the normal range, and hyperventilation should be avoided throughout the case. Depending on the tumor location and hemisphere, every attempt is made to identify and interrupt the blood supply to these tumors early on.

Specific surgical approaches

Common surgical approaches by tumor site are outlined in Table 4.

Site	Approach	Patient position
Atrium	1. Middle temporal gyrus (nondominant)	1. Left lateral
	2. Superior parietal lobule (dominant)	2. Supine or lateral
	3. Contralateral interhemispheric, transcallosal (dominant)	3. Lateral, tumor side up
	4. Ipsilateral interhemispheric transcallosal	4. Supine
Posterior third ventricle	1. Infratentorial supracerebellar	1. Prone or semisitting
	2. Occipital transtentorial	2. Prone, approach side down
Fourth ventricle	Midline suboccipital	Prone or concord

Middle temporal gyrus approach

- _ Patient position: semilateral
- _ Head position: extended on neck, tilted 20^0downward, rotated 90^0 to opposite side,parallel to floor

The middle temporal gyrus approach (Fig. 7) is best suited for meningiomas of the atrium of the lateral ventricle of the nondominant hemisphere. A variety of skin incisions can be used, including U-shaped incisions, reverse question mark incisions, or curvilinear incisions with posterior hockey stick extension coming off at right angles posteriorly(16, 32). A transsulcal approach in the posterior third of the temporal lobe minimizes tissue disruption, and image-guided systems can be used to define the precise trajectory to the ventricle. The advantage of this approach is the ability to pick up the anterior choroidal artery within the temporal horn and eliminate the predominant blood supply before tumor resection starts. The choroid plexus can be followed back to the tumor; internal debulking is then used, followed by capsular dissection of the tumor from the walls and floor of the ventricle. The surgeon should be mindful of visual fibers that first pass over the roof of the temporal horn and then swing back lateral and inferior to the atrium in the periventricular white matter; thus, horizontal subcortical dissection planes should be used to minimize the risk of tract disruption. After removal of the tumor, an external ventricular catheter should be left to drain CSF until returns are clear.

Superior parietal occipital approach

- _ Patient position: supine with back up 15^0 or semilateral
- _ Head position: neck flexed on chest, head slightly flexed on neck, no rotation; or laterally flexed on neck tilted 10^0 up, rotated 90^0 to opposite side

The superior parietal occipital approach can be selected for either the left or right side but is most often employed for dominant hemisphere tumors(3, 11). The author prefers to have the patient supine, with the back elevated so that the patient is in a slouching position, with the neck flexed on the chest and the head flexed on the neck. Most anesthesiologists want to exclude a patent foramen ovale and right-to-left shunt before surgery as noted previously. After pin fixation and registration, the image-guided system is used to help identify the trajectory to the ventricle and the midline. A horseshoe-shaped incision is fashioned, and the brain is exposed to the parietal-occipital junction. A posterior parietal sulcus is used for the dissection, with maintenance of the plane using self-retaining retractors. The corridor to the tumor with this approach is long, and if the tumor is firm and not that vascular, this is one of the few instances where the laser is of particular utility. One disadvantage of this approach is that some of the blood supply (anterior choroidal artery) to the tumor is not identified until late in the procedure. Internal debulking followed by marginal dissection is again employed. Care should be taken not to extend the white matter dissection too far lateral to the atrium for fear of damaging visual pathway fibers.

Interhemispheric transcallosal

Ipsilateral approach

- _ Patient position: semisitting or prone
- _ Head position: neck flexed on chest with head flexed on neck or neck neutral with head flexed on neck

Contralateral approach

- _ Patient position: full lateral, tumor side up
- _ Head position: neck neutral, head slightly flexed on neck, laterally flexed upward toward tumor

The original description of the ipsilateral transcallosal approach was provided by Kempe and Blaylock in 1976(21). The patient can be positioned semi-sitting or prone for a parietal occipital craniotomy. The bone flap should cross the midline, and the interhemispheric fissure should be opened widely. Jun and Nutik(20) described a slightly more vertical angle than along the tentorium so as to preserve part of the splenium. The posterior callosum is split for 2 cm, leaving the posterior part of the splenium intact. The tumor is debulked and removed piecemeal.

Mcdermot(32) uses a contralateral transcallosal approach with success for several cases in the dominant hemisphere (see Fig. 2; Fig. 4). The patient is positioned so that the retracted right hemisphere is toward the floor; thus, retraction is assisted by gravity. A posterior interhemispheric approach is

taken through a large parietal craniotomy crossing midline, preserving parasagittal draining veins. Once the interhemispheric fissure is split along the length of the exposure, the image-guided system is used to define the most appropriate callosotomy, which is not more than 2 cm in length. The inferior two thirds of the falx are incised to allow for gentle retraction of the medial aspect of the left hemisphere and, later, the callosum. The tumor can be removed piecemeal using the long microsurgery set for the supracerebellar approach. At the completion of tumor removal, an external ventricular drain is left in place for several days

Infratentorial supracerebellar

- _ Patient position: semisitting or prone
- _ Head position: neck flexed on chest with head flexed on neck or neck neutral with head flexed on neck

The approaches to meningiomas of the posterior third ventricle are similar to those for pineal region tumors(32). Because meningiomas in this location are so rare, this approach is not discussed in great detail. The choice of the approach generally depends on the patient's body habitus and the angle of the tentorium on sagittal MRI. For obese patients, those with a short neck, or those with a steeply angled up tentorium, the semisitting position is preferred after preoperative cardiac screening. In our experience, massive air embolism is rare. The author uses a torcular craniotomy, exposing the transverse sinuses bilaterally and the torcular with a bipartite bone flap. The posterior

fossa craniotomy below the transverse sinus is done first, followed by the second flap exposing the sinuses from above. The rest of the dissection is as described elsewhere(19).

Occipital transtentorial

- _ Patient position: prone
- _ Head position: neck neutral with head flexed on neck or head flexed and rotated downward toward floor

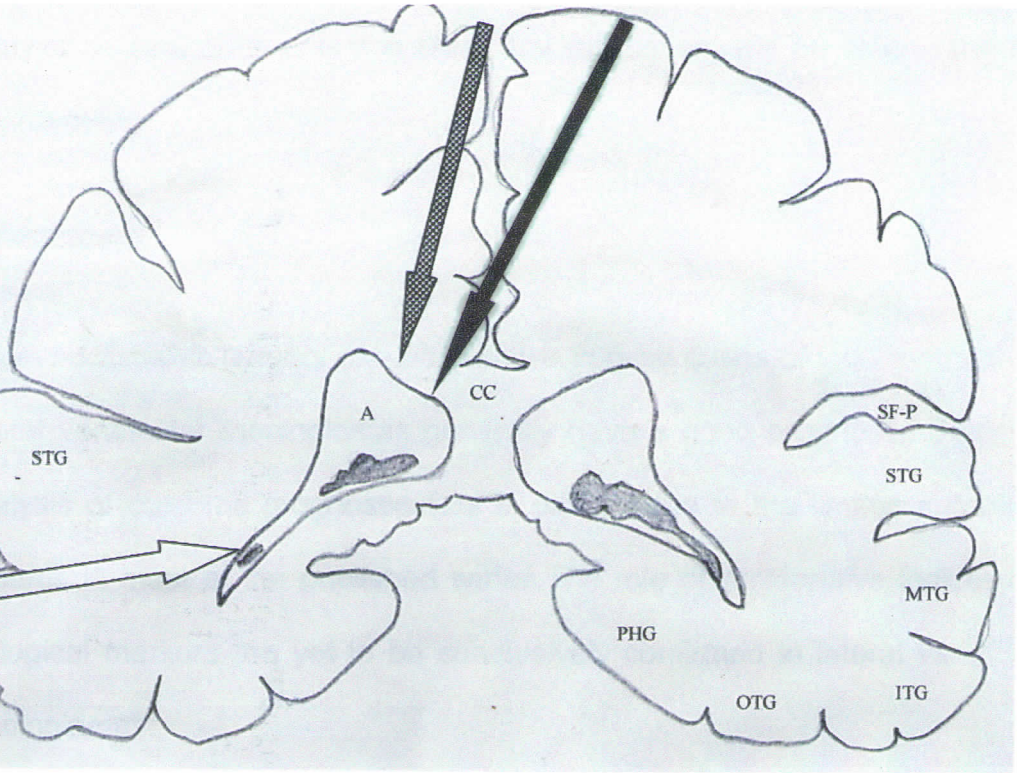
This is another approach for pineal region tumors(32). For the prone position, the craniotomy is on the side of the sinus toward the floor so that the retracted occipital lobe is assisted by gravity. The dura is opened in a U-shaped fashion based laterally so that the brain is protected from a sharp dural edge.

Midline Suboccipital

- _ Patient position: prone
- _ Head position: neck extended on chest with head flexed on neck

Again, meningiomas of the fourth ventricle are rare, but a standard surgical approach is used(9). A midline incision extending into the occipital region is used so that pericranium can be harvested for dural repair at the end. Once the occipital bone is removed and the dura and cisterna magna are opened, the cerebellomedullary fissure is dissected to gain access to the ventricle, thus avoiding splitting the inferior vermis. The fissure is developed by dissecting the lateral aspect of the tonsil and incising the tela choroidea along

posterior lateral margin of the floor of the fourth ventricle to the level of the ral recess(29). The choroidal branch of the posterior inferior choroidal ry is the main supply and should be seen entering the tumor in the roof of ventricle.



ıre 6 Coronal section through atria, anterior cut to Fig. 1. White arrow marks middle mporal transsulcal or middle temporal gyrus approach. Ipsilateral interhemispheric nscallosal approach (shaded arrow) and contralateral interhemispheric transcallosal approach

operative care

is not routinely restricted in these patients so as to reduce the chance of cal venous thrombosis. Low-molecular-weight heparin is started 24 hours

the posterior lateral margin of the floor of the fourth ventricle to the level of the lateral recess(29). The choroidal branch of the posterior inferior choroidal artery is the main supply and should be seen entering the tumor in the roof of the ventricle.

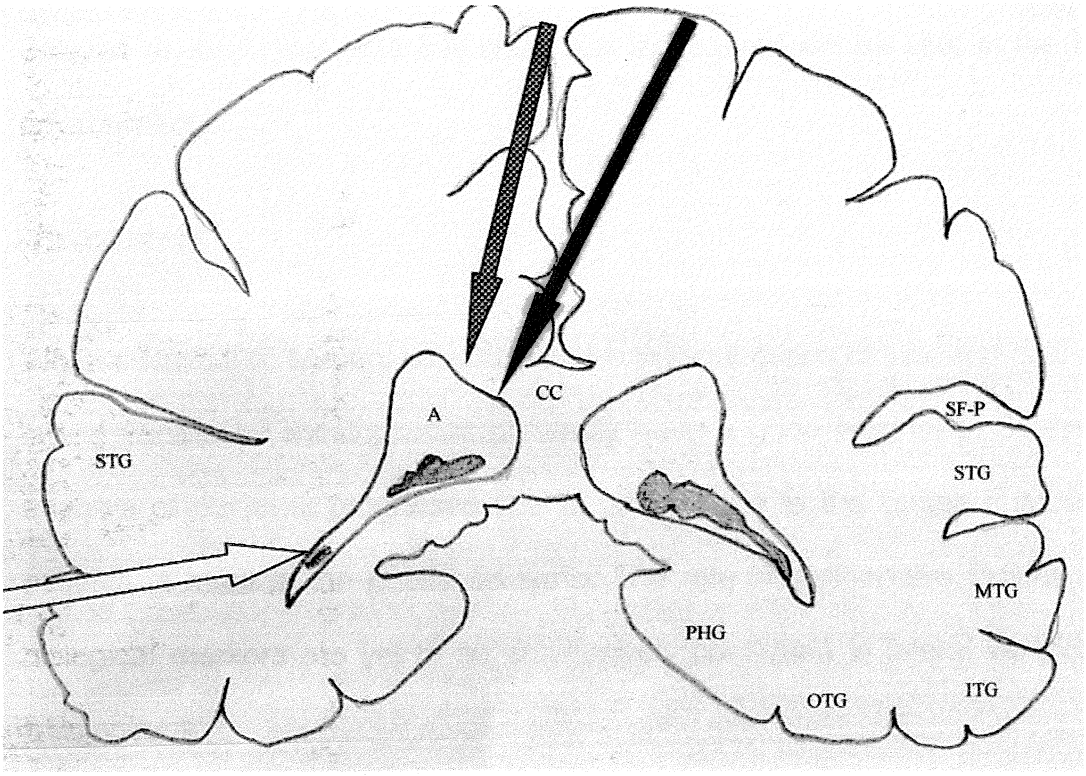


Figure 6 Coronal section through atria, anterior cut to Fig. 1. White arrow marks middle temporal transsulcal or middle temporal gyrus approach. Ipsilateral interhemispheric transcallosal approach (shaded arrow) and contralateral interhemispheric transcallosal approach

Postoperative care

Fluid is not routinely restricted in these patients so as to reduce the chance of cortical venous thrombosis. Low-molecular-weight heparin is started 24 hours

after surgery and is continued for 5 days to reduce the chance of deep venous thrombosis in the extremities. Excessive drainage of CSF should be avoided so as to reduce the chance of subdural hematoma formation. Conversely, adequate clearance of blood and debris from the ventricle reduces the chance of hydrocephalus. Patients can be followed with CT scans to assess ventricle size and debris. Postoperative MRI should be delayed several days until it is clear that the patient will be safe in the MRI environment.

Prognosis

When successful, surgery can be curative in most cases of total removal and lateral ventricular meningiomas generally have a good prognosis. Statistical analysis of outcome prognosticators is difficult due to the limited number of patients in most of the published series.The role of proliferative indices and biological markers are yet to be conclusively confirmed in lateral ventricular meningiomas.

AIM OF THE STUDY

Lateral ventricular meningiomas are rare tumours that pose considerable surgical challenge. Fifteen patients with lateral ventricular meningiomas were surgically managed in our institute in the last ten years. This study attempts to analyse the important clinical features of these tumours and review technical considerations in surgery for lateral ventricular meningiomas.

MATERIALS & METHODS

Patient Population

Between January 1998 and Dec 2007 , 15 patients with lateral ventricular menibgiomas were surgically managed at Sree Chitra Tirunal Institute of Medical Sciences and Technology, Thiruvananthapuram. Their hospital records including follow-up notes and imaging studies were retrospectively reviewed.

Preoperative Examination

The preoperative workup included a detailed neurological examination to establish baseline characteristics. Imaging studies included high-resolution CT and MR imaging depending on what was available at that time. If the internal carotid arteries or the vertebrobasilar system were involved by the tumor, a cerebral arteriogram was acquired and collateral blood flow was assessed through cross-compression studies.

Radical Excision

The patients underwent surgery in which modern techniques were used; the procedures were performed to accomplish total resection of the tumor. A variety of approaches were used depending on the location and size of the tumor. The adequacy of resection was noted based on the surgeon's observation and findings on the postoperative MR and/ or CT studies. Post operative outcome of each patient was recorded using the Glasgow outcome score. The patients underwent regular follow-up and clinical examination at each visit. The follow-up period was defined as the period extending from

surgery to the most recent clinical visit or patient contact. Patients with followup period less than six months were considered as lost to followup. Recurrent tumor was judged according to findings on imaging studies. All tumors were evaluated by a neuropathologist, and the diagnosis of meningioma was confirmed and classified in accordance with WHO classification of central nervous system tumours, 2000.

Table 1 Glasgow outcome score

Score	Rating	Definition
5	Good Recovery	Resumption of normal life despite minor deficits
4	Moderate Disability	Disabled but independent. Can work in sheltered setting
3	Severe Disability	Conscious but disabled. Dependent for daily support
2	Persistent vegetative	Minimal responsiveness
1	Death	Non survival

Data analysis

Data was analyzed using SPSS software.

RESULTS & ANALYSIS

Fifteen patients with intra lateral ventricular meningiomas were operated in the period January 1998 to July 2007. The mean age was 40.6 years (Range 14yrs to 75yrs). Lateral ventricular meningiomas constituted 1.33% of the total number of meningiomas operated during the above period. *Patient characteristics:* The study group included 10 females and 5 males showing a clear female preponderance. Raised intracranial pressure was the predominant symptom at presentation (10/15, 66%). Other presenting symptoms include visual field deficits (6/15; 40%), contralateral motor deficits (5/15; 33.3%) and seizures (4/15; 26.6%). One of the patients had an acute presentation suggestive of a bleed which was confirmed on CT scan examination. Two of our patients presented with recurrence of tumours earlier operated at other centers. On examination at presentation ten patients had papilloedema. Six patients had evidence of contralateral homonymous hemianopia. The salient clinical details are summarized in Table 2. None of them had associated neuorofibromatosis (NF).

Table 1 Symptoms and signs of lateral ventricular meningiomas in our series of 15 patients

Symptoms	**Number**	**%**
Raised ICP Headaches	10	66.6
Visual disturbance	06	40
Motor deficits	05	33.3
Seizures	04	26.16
Mental disturbance	02	13.3
Signs		
Papilledema	10	66.6
Visual field deficit	06	40
Contralateral motor weakness	05	33.3

Imaging findings:

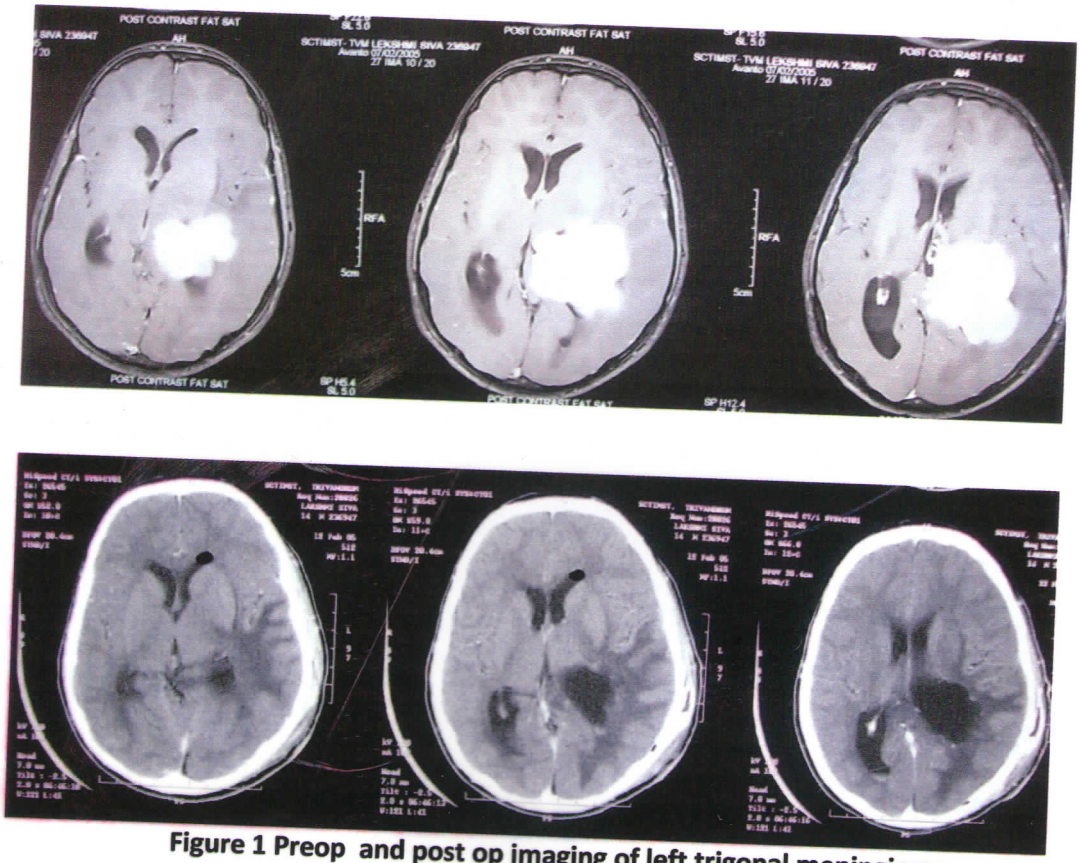


Figure 1 Preop and post op imaging of left trigonal meningioma

The tumour was on the right side in seven patients and on the left side in eight patients. Calcification was seen in five and one patient had evidence of bleed. The lesion was located in the trigone of the lateral ventricle in 13 patients and in the body of the ventricle in two patients. Computerised tomography was done in all the patients , but MRI could not be done in two patients due to financial constraints. The computerized tomography(CT) and magnetic resonance imaging(MRI) features were similar to other meningiomas except for the dural attachment. Majority (11/15, 73.3%) of the

Imaging findings:

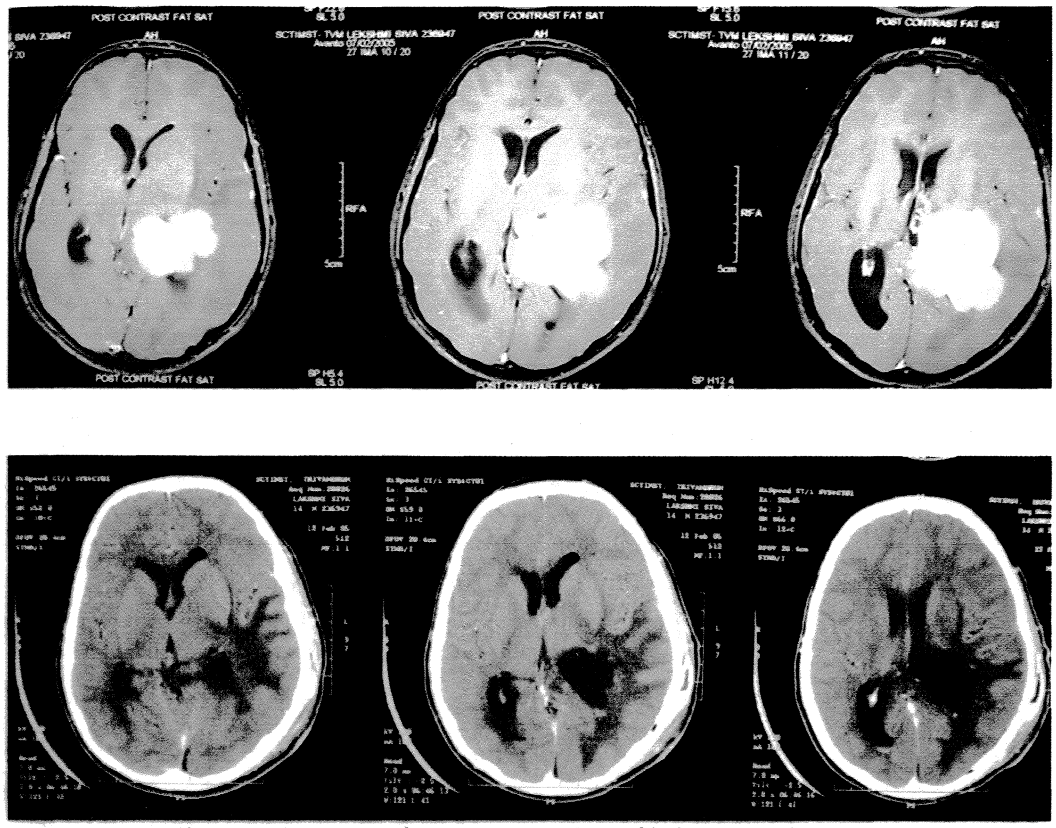


Figure 1 Preop and post op imaging of left trigonal meningioma

The tumour was on the right side in seven patients and on the left side in eight patients. Calcification was seen in five and one patient had evidence of bleed. The lesion was located in the trigone of the lateral ventricle in 13 patients and in the body of the ventricle in two patients. Computerised tomography was done in all the patients , but MRI could not be done in two patients due to financial constraints. The computerized tomography(CT) and magnetic resonance imaging(MRI) features were similar to other meningiomas except for the dural attachment. Majority (11/15, 73.3%) of the

tumours were excised through a parietoccipital approach. The middle temporal gyrus route was used in four patients with significant temporal horn extension. In both the cases with involvement of the body of the lateral ventricle , the tumour was predominantly on the non dominant side and was extending quite posteriorly. We preferred a transcortical over a transcallosal approach as we felt that the risks would be less compared to the risks associated with an extensive callosal sectioning. All the patients except two underwent macroscopic gross total resections which were confirmed on postoperative CT scans. One of the patients who presented with recurrence twice had evidence of residual lesion in the first postoperative scan. She underwent serial imaging and had to be reoperated again two years later as the tumour was seen to be increasing in size. She underwent radiotherapy after the third surgery. In another patient part of the tumour attached to the choroids plexus had to be left behind due to profuse bleeding. The residual tumour has remained static on serial imaging even after three years. Our protocol is to subject patients to serial imaging at six months follow up, and then at two years and thereafter as and when they recruit new symptoms. Majority of the meningiomas were Grade I (14/15) but one was an atypical variant .Both the recurrences occurred in patients with fibroblastic variant (Grade 1) of meningioma. Complications include fresh motor deficits in 3, contralateral homonymous hemianopia in 2, refractory seizures in 2 and dysphasia in one. Two patients operated through the superior parietal lobule approach and one operated through the middle temporal gyrus route had transient contralateral limb weakness which improved before discharge. One

patient was readmitted few months after surgery due to symptoms caused by a trapped temporal horn which required an endoscopic fenestration.

Follow up ranged from 1 year to 8years. Of the five patients with preoperative contralateral limb weakness, three improved and two remained static. Of the six patients who presented with hemianopia, two improved, two remained static and two worsened. Worsening of vision was due to onset of secondary optic atrophy in one patient and injury to the optic radiations in the other patient who was operated through a middle temporal gyrus approach. We had no operative mortality in our series. At the time of last follow up, ten patients were in GOS 5, two were in GOS 4 and three in GOS 3. Of the two patients with recurrent tumours , one was in GOS 5 and other patient was in GOS 3.

DISCUSSION

Intraventricular meningiomas are thought to arise from the mesenchymal stroma of the choroid plexus or from rests of arachnoid cell nest tissue seen within the choroids(16). In the third ventricle they arise from the tela of the velum interpositum, and in the fourth ventricle from the choroids plexus or interior tela choroidea(3). Two patterns of extension of intraventricular meningiomas have been described; one where the tumour arises from the choroid plexus and grows within the ventricle and the other where it arises from the tela choroidea and grows partly within the ventricle and partly into the surrounding brain. Intraventricular meningiomas are most often seen in the ventricular trigone, followed by the atrium, temporal horn, third and fourth ventricle in that order. This pattern is reflected in the available data on intraventricular meningioma summarized by Nakamura(33) where, of the total 532 intraventricular meningiomas, 413 (77.6%) were in the lateral ventricle, 83 (15.6%) in the third ventricle and 35(6.57%) in the fourth ventricle. In our series of 15 lateral intraventricular meningiomas, 13 were located in the trigone, while two were in the body of the ventricle. For reasons unknown meningiomas appear to be more common on the left side in most of the reported series except in one series(3, 22, 27). In the current series the meningiomas were almost equally distributed, eight being on the left side and seven on the left.

As seen in meningiomas in general, a female preponderance has been reported in intraventricular meningiomas. On an average a 2:1 female preference is observed as was seen in our series as well(10, 27). A few authors have however, reported no female predilection(33) and some(3) have

even reported a male preponderance. Intraventricular meningiomas are rare in the pediatric population and the mean age of occurrence is 20-50yrs for lateral ventricular tumours, 28.7yrs for third ventricular tumours and 35.7yrs for fourth ventricular tumours(11, 16, 30). However, among pediatric meningiomas a higher incidence of intraventricular location has been reported 4–17%(12, 23). The mean age of our study group was 40.6 years which included 3 childhood meningiomas (7.3%)

Intraventricular meningiomas distend the ventricle they occupy and due to this compensation, symptoms appear late and tumours are quite large by the time of presentation. They most often present with raised intracranial pressure symptoms but may also present with symptoms secondary to pressure on surrounding structures. Cushing and Eisenhardt(8) described five clinical features- of lateral ventricle tumors: 1) pressure symptoms with ipsilateral headache; 2) contralateral macula splitting homonymous hemianopia; 3) contralateral sensorimotor paresis, more marked sensory involvement, and numbness over trigeminal distribution; 4) cerebellar involvement in more than half of patients; and 5) paralexia, worsened by surgical intervention in left-sided tumors. Delandsheer(9) in his review of 175 cases from the literature, summarized that patients with lateral ventricular meningiomas often present with a clinical syndrome, suggesting both a posterior cranial fossa lesion and a cerebral hemisphere lesion with intermittent and paroxysmal nature of some symptoms. As seen in other series, raised intracranial tension was the most common presentation in our series (10/15). Incidence of visual symptoms can range from 25 % to 70%(3, 10). Visual deficits are attributable to stretching of

the optic radiations by large atrial tumours and to concentric field cuts secondary to papilledema. Six patients, in our series had contralateral homonymous hemianopia on presentation and four had concentric field cuts due to papilloedema. Two of our patients presented with symptoms of impaired memory probably related to chronic hydrocephalus or to direct compression on the hippocampal formation in the floor of the atrium by large tumors. Owing to their predominant intraventricular location seizures as a presenting symptom are unusual(3). However four of our patients (26.6%) presented with seizures, an incidence similar to Guidettis report(27%)(16). Two of our patients with seizures had recurrent tumours and seizures in these patients could be related to the earlier surgery. None of our patients had presented with the cerebellar symptoms or paralexias as described by Cushing(8) or Delandsheer(9). Tumors of the third and fourth ventricles usually present with symptoms of obstructive hydrocephalus. Anterior third ventricular tumours may present with hypothalamic and endocrine abnormalities and similarly posterior third ventricle lesions may manifest with Parinaud's syndrome with impaired upgaze and pupillary light reflexes. Romeike B(35) and .Bosnjak R(4) suggest that among all meningiomas , intraventricular meningiomas have an increased tendency to bleed and they may present with intratumoural and intraventricular bleed as was observed in one of our patients.

Radiologically intraventricular meningiomas resemble meningiomas at other locations except for the wide dural base(4, 11, 19, 22, 30). On CT scan they appear as a globular well defined, well enhancing mass lesions [16,17,18].

Calcification is not uncommon and can be as high as 47% (22). Calcification was seen in 33% (5 cases) of our cases. Lesions in the trigone and the atrium may cause loculated enlargement of the occipital and temporal horn. On MR they appear as isointense to hypointense on T1, isointense to hyperintense on T2. Although it is difficult to distinguish the histological type on MR, MCdermot(32) suggest that fibroblastic meningiomas, the most common pathologic subtype of meningioma within the ventricle, are usually prominently hypointense on T2-weighted images. On MR spectroscopy, a high alanine creatine ratio with high levels of alanine and low levels of creatine is characteristic of meningiomas(19, 29). Angiogram may reveal a prominent anterior or posterior choroidal artery with a diffuse tumour blush especially in large tumours. The blood supply of tumours in the atrium is from feeders from the both anterior and posterior choroidal ,in the temporal horn from anterior choroidal and in the body from posterior choroidal artery. Third ventricle tumours derive their supply from the medial posterior choroidal and tumours in the 4^{th} ventricle get supply from the PICA and precentral cerebellar branches of superior cerebellar artery. Intraventricular meningiomas are generally benign and most often are either fibroblastic or meningothliomatous variants. Anaplastic and malignant varieties have been reported but generally are rare(10, 32). The only patient with an atypical variant in our series did well after surgery and on last follow up scan done 3 years after surgery had no evidence of recurrence.

The main differential diagnosis of a tumour located in the trigone includes a choroids plexus papilloma in patients under the age of 10yrs, ependymoma,

oligodendroglioma in patients between 10 and 40 years of age, metastasis and lymphoma after the fourth decade of life. Choroid plexus papilloma can be distinguished by their frond like projections and generalized enlargement of all ventricles unlike a loculated ventriculomegaly seen in meningiomas. Glial tumours lack the characteristic enhancement pattern of meningiomas . MR spectroscopy and DWI(diffusion weighted images) help to differentiate metastasis and lymphoma to some extent(19, 29).

Although total surgical excision should be the treatment goal, observation : does have a role in select patients(21). We agree with Mcdermot's(32) policy of watchful expectancy in the elderly with left side lesion. The slow rate of growth and generally benign nature suggest that their natural history is likely to be better than the risk associated with surgery in patients with high surgical risk.

It was Macdowell who performed the first documented excision of a left trigonal tumour in 1881(25, 41). Different surgical approaches have been advocated thereafter, but the decision on where to place the incision must depend on the actual neurological deficit, the surgical difficulty of the approach, the relative risk of additional deficits, and the experience of the surgeon. For lesions in the frontal horn a frontal approach as advocated by Busch would suffice. Olivecrona proposed a posterior middle temporal gyrus approach which was popularized by many surgeons (7, 20, 21, 25, 26, 41). Cramer and later Fornari(11)-promoted a posterior parieto occipital approach which currently appears to be the favourite of most surgeons. Kempe and

Blaylco- advocated a posterior transcallosal to reach the trigone especially for tumours extending to both sides , an approach associated with a risk of disconnection syndrome(20, 25). Yasargil used his parietooccipital interhemispheric approach for parasplenial lesions including meningiomas and AVMs(44). Image guidance helps in making precise cortical incision minimizing damage to eloquent fibres. CUSA helps in piece meal tumour excision minimizing the risk associated with enbloc excision. Use of a neuroendoscope helps in locating the feeders, in ensuring completeness of excision and in clearing intraventricular clots after tumour excision. Meng Liu(27) and MCdermot(32) have suggested use of post operative external ventricular drainage for 48 hrs helps to reduce the risk of intraventricular clot formation which is a useful surgical adjunct in our series.

Preoperative investigations should include a detailed neuropsychological evaluation and an accurate visual field charting. Preoperative embolisation is difficult as the choroidal arteries are difficult to cannulate, and carry a risk of spasm and subsequent infarction. For eloquent hemisphere lesions in the trigone we prefer the superior parietal lobule approach. Parietoccipital approach (superior parietal lobule) especially for left side lesions through a high cortical posterior parietal sulcus is a safe approach with little risk of speech or motor deficits. Disadvantages include the depth, late control of vascularity, increased risk of seizure and risk of injury to optic radiations. Two of our patients developed fresh contralateral homonymous hemianopia following surgery .The reported incidence of post operative visual deficits can be as high as 20-60% which can be minimized by correct cortical incision as

the optic radiations run inferolaterally to the ventricles(33). The middle temporal gyrus approach is reserved for tumours with significant temporal horn extension, especially in the nondominant hemisphere. The advantage of this approach is the ability to pick up the anterior choroidal artery within the temporal horn and eliminate the predominant blood supply before tumor resection starts. The risk of visual deficits can be minimized by using horizontal subcortical dissection planes as the visual fibers first pass over the roof of the temporal horn and then swing back lateral and inferior to the atrium in the periventricular white matter. Language function may be compromised however by thus approach especially in the dominant hemisphere. Moreover large tumours reaching into the occipital horn and attached to the ependyma maybe difficult to remove. In such cases a two stage approach may be required combining the transcallosal approach as suggested by Couillard. In both, the superior parietal and the middle temporal gyrus approach, excessive retraction on the motor fibres can result in contralateral limb weakness . In our series two patients operated through the superior parietal lobule approach and one operated through the middle temporal gyrus route had transient contralateral limb weakness which improved before discharge. Similarly postoperative seizures is a potential complication in both the approaches and Fornari et a(11) have reported a 29% incidence of seizures via a parieto occipital approach. Two patients in our series developed refractory seizures and are on multiple antiepileptic drugs. Mcdermot(32) have used the contralateral transcallosal approach with success for several cases in the dominant hemisphere and for tumours in the middle body with bilateral

extension. Advantages of this approach include, excellent exposure of the trigone and posterior third ventricle, low risk of seizures, low risk of speech and visual deterioration and early access to posterior choroidal artery. Disadvantages include risk of disconnection syndrome and this route is contraindicated in patients with right homonymous hemianopia because splitting of the callosum may result in alexia with agraphia.

Surgical outcome is favorable in majority of the patients. Raised intracranial pressure symptoms, visual and motor deficits generally improve once the mass effect is reduced. Visual improvement also depends on the extent and duration of insult to the visual fibres. One of our patients with florid papilloedema went into secondary optic atrophy in spite of total tumour excision. Recurrence is almost always associated with subtotal excision. Recurrent tumours are best managed by repeat surgery as the role of radiosurgery is doubtful. We had three recurrences, one of the tumours recurring twice. Mortality rates have dropped from 40% in some early series to 0% in almost all recent series(2, 3, 10, 27, 28, 33, 42). We had no operative mortality. Mortality is more with intoto excision, therefore piecemeal excision using CUSA is recommended prior to capsule dissection.

Lee, Pollock(26, 34) have reported control rates as high as 85-98% with radiosurgery and Goldsmith(13) has reported similar observations with 3D conformational radiotherapy. The role of radiosurgery is however doubtful and probably confined to small lesions in patients with comorbidites contraindicating surgery. McDermott(32) has warned about the risk of

exaggerated radiation toxicity in the subependymal region even with low dose and have suggested restricting radiosurgery to lesions less than 1.5 to 2 cms

CONCLUSION

- Intraventricular meningiomas are rare tumours that pose difficult surgical challenge. Surgical approaches need to be tailored depending on patient and tumor factors.
- Total surgical excision through a superior parietal lobule is possible in most of the trigonal tumours.
- Middle temporal gyrus approach is an option for tumours in the in the dominant hemisphere with significant temporal horn extension .

BIBLIOGRAPHY

1. Alheit H, Saran FH, Warrington AP, Rosenberg I, Perks J, Jalali R, Shepherd S, Beardmore C, Baumert B, Brada M: Stereotactically guided conformal radiotherapy for meningiomas. **Radiother Oncol** 50:145-150, 1999.

2. Bertalanffy A, Roessler K, Koperek O, Gelpi E, Prayer D, Neuner M, Knosp E: Intraventricular meningiomas: a report of 16 cases. **Neurosurg Rev** 29:30-35, 2006.

3. Bhatoe HS, Singh P, Dutta V: Intraventricular meningiomas: a clinicopathological study and review. **Neurosurg Focus** 20:E9, 2006.

4. Bosnjak R, Derham C, Popovic M, Ravnik J: Spontaneous intracranial meningioma bleeding: clinicopathological features and outcome. **J Neurosurg** 103:473-484, 2005.

5. Carroll RS, Schrell UM, Zhang J, Dashner K, Nomikos P, Fahlbusch R, Black PM: Dopamine D1, dopamine D2, and prolactin receptor messenger ribonucleic acid expression by the polymerase chain reaction in human meningiomas. **Neurosurgery** 38:367-375, 1996.

6. Chuang CC, Chang CN, Tsang NM, Wei KC, Tseng CK, Chang JT, Pai PC: Linear accelerator-based radiosurgery in the management of skull base meningiomas. **J Neurooncol** 66:241-249, 2004.

7. Criscuolo GR, Symon L: Intraventricular meningioma. A review of 10 cases of the National Hospital, Queen Square (1974-1985) with reference to the literature. **Acta Neurochir (Wien)** 83:83-91, 1986.

8. Cushing H, Eisenhardt L: Meningiomas. Their Classification,Regional Behavior, Life History and Surgical End Results. Springfield IL,, Charles C Thomas, 1938, pp 139-149.

9. Delandsheer JM: [Meningiomas of the Lateral Ventricle.]. **Neurochirurgie** 11:3-83, 1965.

10. Erman T, GÃ¶Ã§er AI, Erdogan S, Boyar BI, Hacyakupoglu S, Zorludemir S: Intraventricular Meningiomas: A Review of the Literature and Report of 8 Cases. **Neurosurgery Quarterly** 14:154-160, 2004.

11. Fornari M, Savoiardo M, Morello G, Solero CL: Meningiomas of the lateral ventricles. Neuroradiological and surgical considerations in 18 cases. **J Neurosurg** 54:64-74, 1981.

12. Germano IM, Edwards MS, Davis RL, Schiffer D: Intracranial meningiomas of the first two decades of life. **J Neurosurg** 80:447-453, 1994.

13. Goldsmith BJ, Wara WM, Wilson CB, Larson DA: Postoperative irradiation for subtotally resected meningiomas. A retrospective analysis of 140 patients treated from 1967 to 1990. **J Neurosurg** 80:195-201, 1994.

14. Goodwin JW, Crowley J, Eyre HJ, Stafford B, Jaeckle KA, Townsend JJ: A phase II evaluation of tamoxifen in unresectable or refractory meningiomas: a Southwest Oncology Group study. **J Neurooncol** 15:75-77, 1993.

15. Grunberg SM, Weiss MH, Russell CA, Spitz IM, Ahmadi J, Sadun A, Sitruk-Ware R: Long-term administration of mifepristone (RU486): clinical tolerance during extended treatment of meningioma. **Cancer Invest** 24:727-733, 2006.

16. Guidetti B, Delfini R: Lateral and fourth ventricle meningiomas, in Al Mefty O (ed): *Meningiomas*, New York: Raven Press, 1991, pp 569-587.

17. Herz DA, Shapiro K, Shulman K: Intracranial meningiomas of infancy, childhood and adolescence. Review of the literature and addition of 9 case reports. **Childs Brain** 7:43-56, 1980.

18. Jaaskelainen J: Seemingly complete removal of histologically benign intracranial meningioma: late recurrence rate and factors predicting recurrence in 657 patients. A multivariate analysis. **Surg Neurol** 26:461-469, 1986.

19. Jelinek J, Smirniotopoulos JG, Parisi JE, Kanzer M: Lateral ventricular neoplasms of the brain: differential diagnosis based on clinical, CT, and MR findings. **AJR Am J Roentgenol** 155:365-372, 1990.

20. Jun CL, Nutik SL: Surgical approaches to intraventricular meningiomas of the trigone. **Neurosurgery** 16:416-420, 1985.

21. Kempe LG, Blaylock R: Lateral-trigonal intraventricular tumors. A new operative approach. **Acta Neurochir (Wien)** 35:233-242, 1976.

22. Kendall B, Reider-Grosswasser I, Valentine A: Diagnosis of masses presenting within the ventricles on computed tomography. **Neuroradiology** 25:11-22, 1983.

23. Kloc W, Imielinski BL, Wasilewski W, Stempniewicz M, Jende P, Karwacki Z: Meningiomas of the lateral ventricles of the brain in children. **Childs Nerv Syst** 14:350-353, 1998.

24. Kondziolka D, Levy EI, Niranjan A, Flickinger JC, Lunsford LD: Long-term outcomes after meningioma radiosurgery: physician and patient perspectives. **J Neurosurg** 91:44-50, 1999.

25. Konovalov A N, Filatov Y M, Belousowa O B: Intraventricular meningiomas, in Schmidek H H (ed): *Meningiomas and Their Surgical Management.* New York, Saunders W B, 1991, pp 364-374.

26. Lee JY, Niranjan A, McInerney J, Kondziolka D, Flickinger JC, Lunsford LD: Stereotactic radiosurgery providing long-term tumor control of cavernous sinus meningiomas. **J Neurosurg** 97:65-72, 2002.

27. Liu M, Wei Y, Liu Y, Zhu S, Li X: Intraventricular meninigiomas: a report of 25 cases. **Neurosurg Rev** 29:36-40, 2006.

28. Lyngdoh B T, Giri P J, Behari S, Banerji D, Chhabra D K, Jain V K: Intraventricular meningiomas: a surgical challenge. **J Clin Neurosci** 14:442-448, 2007.

29. Majos C, Cucurella G, Aguilera C, Coll S, Pons LC: Intraventricular meningiomas: MR imaging and MR spectroscopic findings in two cases. **AJNR Am J Neuroradiol** 20:882-885, 1999.

30. Mani RL, Hedgcock MW, Mass SI, Gilmor RL, Enzmann DR, Eisenberg RL: Radiographic diagnosis of meningioma of the lateral ventricle. Review of 22 cases. **J Neurosurg** 49:249-255, 1978.

31. Maxwell M, Galanopoulos T, Hedley-Whyte ET, Black PM, Antoniades HN: Human meningiomas co-express platelet-derived growth factor (PDGF) and PDGF-receptor genes and their protein products. **Int J Cancer** 46:16-21, 1990.

32. McDermott M W: Intraventricular meningiomas.: *Neurosurg Clin N Am* 2003, pp 559-569.

33. Nakamura M, Roser F, Bundschuh O, Vorkapic P, Samii M: Intraventricular meningiomas: a review of 16 cases with reference to the literature. **Surg Neurol** 59:491-503; discussion 503-494, 2003.

34. Pollock BE: Stereotactic radiosurgery for intracranial meningiomas: indications and results. **Neurosurg Focus** 14:e4, 2003.

35. Romeike BF, Joellenbeck B, Skalej M, Scherlach C, Kirches E, Mawrin C: Intraventricular meningioma with fatal haemorrhage: clinical and autopsy findings. **Clin Neurol Neurosurg** 109:884-887, 2007.

36. Rushing EJ, Olsen C, Mena H, Rueda ME, Lee YS, Keating RF, Packer RJ, Santi M: Central nervous system meningiomas in the first two decades of life: a clinicopathological analysis of 87 patients. **J Neurosurg** 103:489-495, 2005.

37. Sandberg DI, Edgar MA, Resch L, Rutka JT, Becker LE, Souweidane MM: MIB-1 staining index of pediatric meningiomas. **Neurosurgery** 48:590-595; discussion 595-597, 2001.

38. Soyuer S, Chang EL, Selek U, Shi W, Maor MH, DeMonte F: Radiotherapy after surgery for benign cerebral meningioma. **Radiother Oncol** 71:85-90, 2004.

39. Spiegelmann R, Nissim O, Menhel J, Alezra D, Pfeffer MR: Linear accelerator radiosurgery for meningiomas in and around the cavernous sinus. **Neurosurgery** 51:1373-1379; discussion 1379-1380, 2002.

40. Stafford SL, Perry A, Suman VJ, Meyer FB, Scheithauer BW, Lohse CM, Shaw EG: Primarily resected meningiomas: outcome and prognostic factors in 581 Mayo Clinic patients, 1978 through 1988. **Mayo Clin Proc** 73:936-942, 1998.

41. Tew J M, Larson J J Jr: Intraventricular meningiomas, in Kaye A H, Black P M L (eds): *Operative Neurosurgery.* London, Churchill Livingstone, 2000, pp 575-585.

42. Wang X, Cai BW, You C, He M: Microsurgical management of lateral ventricular meningiomas: a report of 51 cases. **Minim Invasive Neurosurg** 50:346-349, 2007.

43. Westphal M, Herrmann HD: Epidermal growth factor--receptors on cultured human meningioma cells. **Acta Neurochir (Wien)** 83:62-66, 1986.

44. Yasargil MG, Abdulrauf SI: Surgery of intraventricular tumors. **Neurosurgery** 62:1029-1040; discussion 1040-1021, 2008.