Our Excursion with Surgical Conduct for Craniopharyngioma

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ARTICLE INFO
Article History
Submitted: 25 August, 2021
Accepted: 15 October, 2021
Published: 8 February 2022

Source of support: None
Conflict of Interest: None

ABSTRACT

Background: Craniopharyngioma, an epithelial tumor believed to arise from the remnants of Rathke's pouch, portray approximately 1.2%-4.4% of all intracranial tumors. Due to its inherent domain in the skull base and its liaison with indispensable neurovascular structure, it has still remained an intimidating contest despite an improved dexterity among the neurosurgeons and refinement in neurosurgical gadgetry.Surgical corridor to come down in favor for is elected by the position of optic chiasm, extension of tumor and development of ACoM or ACA (A1).

Methods: A retrospective series study was conducted at Annapurna Neurological Institute and Allied Sciences between January 2016 and August 2021. A total of 20 patients who underwent surgery for histopathologically proven craniopharyngioma was enrolled. Majority of the surgery was performed via a pterional approach while two cases were addressed with a supra ciliary approach and one with a trans nasal trans septal transsphenoidal approach.

Result: The age of presentation among our study group ranged from 5 years to 46 years with a mean age of 28.2 years.The most frequent mode of presentation was headache associated with visual disturbances (visual acuity and visual field). Histopathological analysis disclosed an admantinomatous variant 15 cases and a papillary type in 5 cases. The use of Ommaya reservoir in few selective cases was escorted by the cystic ingredient of the lesion. An endeavor to aggressive surgical approach was accomplished, amidst, excision of tumor was subtotal in 9 %, near total in 36 % and gross total in 45 %. Diabetes insipidus was seen in 36%. We had to endure a case of mortality, who lamentably had a massive sub-arachnoid hemorrhage with a colossal PCA territory infarction on arrival. Post operatively, the mass effect evolved on account of PCA territory infarction.

Conclusion: The magnitude of resection of the tumor is affected by the extension of tumor, consistency of the lesion and position of optic chiasm. Even with an extensive resection, the prospect of recurrence is inordinately high.

Keywords: Craniopharyngioma; Outcome; Resection.

INTRODUCTION

Presumed to emerge from the fetal remnant of Rathke's pouch, craniopharyngioma is a benign entity, embracing a mere 1.2%-4.4% of allintra cranial neoplasm. Despite an exponential breakthrough in the neurosurgical providence, it has still remained an intriguingdefiance owing to its alliance with paramount neurovascular structures, including the pituitary stalk, hypothalamus, optical apparatus and crucial angioarchitecture within its vicinity. In spite of being a benign variant, it is associated with a soaring rate of recurrence.[1] A divergent array of surgical strategies have come forth depending on the location of the tumor and its relation with optic chiasm. The pterional, orbito zygomatic, trans basal sub frontal, front basal interhemispheric and transnasal transsphenoidal approaches are to name a few.[2–8] We herein attempt to narrate our ordeal in the surgical management of craniopharyngioma.

METHODS

This study was reviewed and approved by the medical

ethics committee of Annapurna Neurological Institute and Allied Sciences. The Institutional Review Board approved a waiver of consent for the collection of data as part of the routine clinical care and quality control. A retrospective series study was conducted between January 2016 and August 2021. A total of 20 patients who underwent craniotomy and excisionof craniopharyngioma were enrolled. Most of them were addressed via pterional approach except two required supraciliary approach and one required transsphenoidal approach. The preoperative and postoperative clinical reports, radiological and laboratory findings including the operative videos were reviewed. The ages of the patients ranged from 5 to 46 years being a mean age of 28.2. There were 14 males and 6 females ensuing male preponderance. The commonest mode of presentation was headache. Visual disturbance was encountered in 7 patients. Two of the patient had a recurrence.

Preoperative Evaluation

Foregoing surgery, all patients underwent a thorough neuropsychological, ophthalmological, endocrinological, and radiological assessment. Ten patients complained of headache at the time of presentation. Visual deterioration was detected in 7 patients. The principle diagnostic imaging modality in this series was MRI with and without enhancement.Tumor size and volume was estimated from MR images by measuring the maximum anteroposterior, vertical, and horizontal diameters. The extent of intratumoral calcification was determined using supplemental CT scans. Endocrinological assay was executed beforehand and following surgery gauging the basic levels of serum luteinizing hormone, follicle-stimulating hormone, free triiodothyronine, free thyroxine, thyroid-stimulating hormone, growth hormone, cortisol, adrenocorticotropic hormone, and prolactin. Diabetes Insipidus was diagnosed before and after surgery based on the sodium level and the presence of hypotonic polyuria.

The benchmark to the extent of resection was categorized. A Gross-total resection (GTR) was defined as no residual enhancing lesion or residual calcification. Near-total resection (NTR) was defined as residual enhancing lesion or calcification < 0.5 cm3. Partial resection (PR) was defined as residual enhancing lesion or calcification ≥ 0.5 cm3.

Operative approaches

The trail to the surgical corridor leading to these suprasellar lesions are diverse. Nonetheless, we have opted the pterional approach in most of our cases. However, in 2 case supraciliary approach was chosen

RESULTS

Tumor characteristics

Among 20 patients undergoing surgery, based on the Among 20 patients undergoing surgery, based on the anatomic sub classification of craniopharyngioma,17 (85%) were retro chiasmatic type and 3(15%) as the prechiasmatic type.

Resection outcomes

Gross –total resection (GTR) was achieved in 14 patients, sub-total resection (STR) in 2 patient and near-total resection (NTR) in 4 patients. (Table 1) In 1 patient, pituitary stalk was inside the tumor and was not separately visualized, therefore pituitary stalk was sacrificed.

Ommaya reservoir

The decision to install Ommaya reservoir was guided by the predominant cystic component of the tumor in order to get to grips with recollection in the future. Out of 20 cases, Ommaya reservoir was placed in 4 cases (Table 1).

Endocrinological status

Antecedent scrutiny of the anterior pituitary function depicted normal in 9 patients, partial hypopituitarism in 8, pan hypopituitarism in 1, hyperprolactinemia in 7 and low cortisol in 10 of them. Even so, none of the patients had diabetes insipidus. Nonetheless,transient DI was seen in 12 patients post operatively.(Table 1)

Tumor recurrence

All the patients were followed up as per the protocol with repeat MRI of brain with and without gadolinium contrast enhancement. The mean age of follow up was 6 years.One of our patients was afflicted with recurrence within 15 months post operatively regardless of gross total resection achieved in him.A practice of standard consultation with clinical oncologist is imperative in our institution. As counseled, two of our patients underwent radiotherapy.

Complications

Fortuitously, we had no intra operative mortality in our case series. However, an early postoperative mortality was recorded in 3. An anticipated hydrocephalus was encountered in other 3 patients which was intervened with release of the cystic content and deployment of Ommaya reservoir. Hydrocephalus secondary to hemorrhage accompanied by intraventricular extension was encountered in 1 patient in whom insertion of external ventricular drain was performed. He succumbed into a sequelae of ventriculitis. An utmost attempt with all probable measures including regular irrigation with an antibiotic solution was done to forestall the provocative aftermath. Neurocognitive deterioration was tape in 1 case.

Table 1: Demographics and Clinical Characteristics of Participants

Patient Characteristics	Frequency	Percentage
Age Group		
5-15 years	4	20%
16-25 years	5	25%
26-35 years	3	15%
36-45 Years	7	35%
46-55 years	1	5%
Symptoms		
Headache with Visual Disorientation	13	65%
Headache Only	1	5%
Loss of Concious Only	1	5%
Headache with LOC	1	5%
Amenorrhoea with Visual Disorientation	1	5%
Ammenorrhoes Only	1	5%
Visual Disturbance, Weight Loss, Erectile Dysfunction	1	5%
Seizure	1	5%
Hormonal Status		
Normal	10	50%
Hyperprolactenemia with Hypocorticism	5	25%
Hyperprolactenemia	2	10%
Hypocoticism	3	15%
Extent of resection		
NTR	3	15%
GTR	14	70%
STR	3	15%
Ommaya Reservoir		
Yes	4	20%
No	16	80%
Early Postoperative status		
Diabetes Incipidus	9	45%
Diabetes Incipidus with Panhypopitituarism	3	15%
Diabetes Incipidus with ICH,Hydrocephalus & Venticulitis	1	5%
Hypocorticism	1	5%
Uneventful	5	25%

Illustrative cases

Case 1

History and Examination

A 21 -year- old-male presented to our institution with the chief complaints of headache for 1 month. It was gradual in onset, mild in intensity, more over the bi frontal region without any aggravating factors and relieved by simple analgesics. He had no issues with visual acuity and field of vision.

CT scan of head was advised which revealed a mass in the supra sellar area associated with calcification and hydrocephalus. Plain and contrast enhanced Magnetic resonance imaging (MRI) was performed that showed an extra axial multicystic peripherally enhancing lesion in the suprasellar region with smooth peripheral wall with enhancing infundibular extension with mass effect and a resultant hydrocephalus(Fig1A&B). The optic chiasm was prefixed type i.e., located anterior to the tumor and pituitary stalk and short optic nerve. Hormonal evaluation disclosed a deranged prolactin level increased to 921 uIU/l. Rest of the hormonal and biochemical parameters were within normal limit.

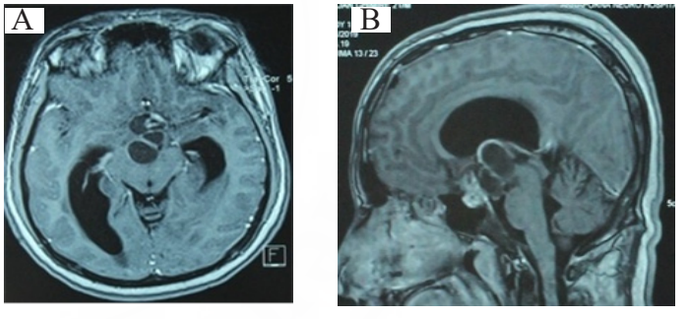

Figure 1: Preoperative axial (A) and sagittal (B) contrast enhanced MRI showing multicystic peripherally enhancing extra axial lesion in the suprasellar region with smooth peripheral wall with enhancing infundibular extension associated with mass effect and hydrocephalus.

Operation and postoperative course

A standard Pterional craniotomy was undertaken from the left side. The carotico - optic triangle was visualized. The optico-carotid cistern was opened and a prefixed variant of the tumor was encountered(Fig.2 A,B & C). Piecemeal extraction of the tumor was performed and gross total resection of the tumor was achieved.

Histopathological evaluation was performed and a diagnosis of Adamantinomatous Craniopharyngioma, WHO grade I was made (Fig.2D).

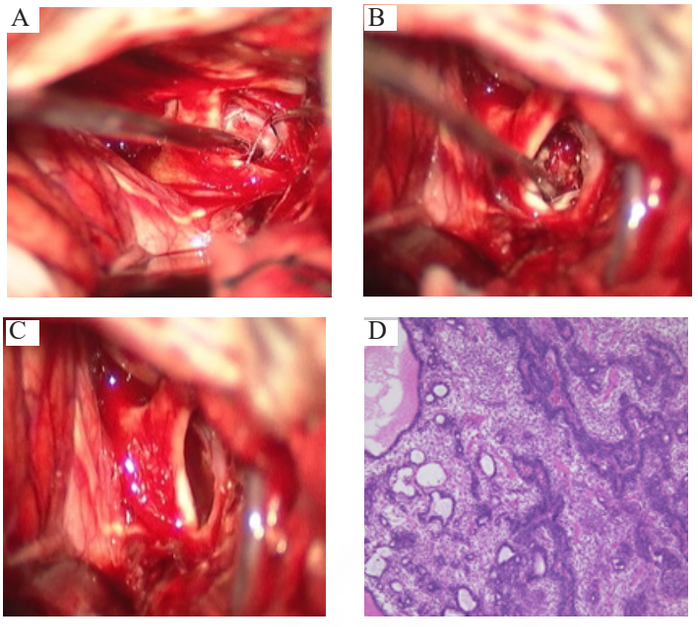

Figure 2: Frontal lobe retraction showing narrow prechiasmatic space (A),tumor was removed through optico-carotid space (B) & (C). Histopathology (D)

Case 2

History and Examination

This 40–year-old-male presented with headache for 1 year. He had blurring of vision on left eye. MRI revealed well-circumscribed solid,cystic,lobulated sellar/suprasellar lesion with maximum diameter of 27 mm(Fig.3 A,B,C&D). The optic tract was pushed by the tumor superiorly. CT head showed supra sellar mass with calcification and hydrocephalus. The optic chiasma was postfixed type i.e., optic chiasm is displaced posteriorly and superiorly.

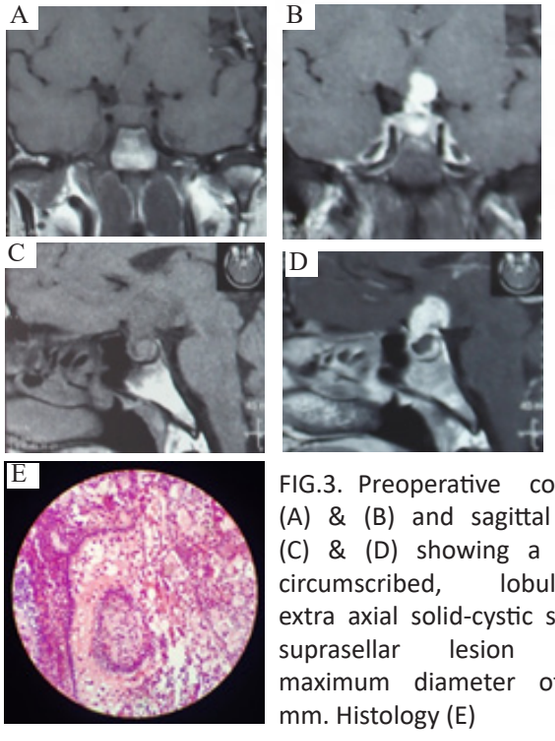

FIG.3. Preoperative coronal (A) & (B) and sagittal MRI (C) & (D) showing a well-circumscribed, lobulated, extra axial solid-cystic sellar-suprasellar lesion with maximum diameter of 27 mm. Histology (E)

Operation and postoperative course

An unaccustomed supraorbital keyhole approach was used to drill bony ridge over the orbital roof so as to flatten the frontal base. The dura was opened in a semicircular fashion. The arachnoid of the carotid cistern, the sylvian fissure, and suprasellar cisterns were opened for CSF drainage in an effort to create enough room for brain retraction and surgical manipulation. Tumor was removed in piecemeal. Gross total excision of tumor achieved. Histopathological analysis revealed a diagnosis of Papillary type Craniopharyngioma, WHO grade I (Fig.3E).

Case 3

History and Examination

This 30-year- old male presented to our out-patient department with the complaints of blurring of vision on the right eye for one and half year. It was progressively worsening in nature followed by decrease in vision of the affected eye affecting his field of vision. Moreover,he also complained of loss of body hair for 1 year and weight loss of 8 kg in the last year. It was associated with loss of libido and erectile dysfunction for same duration. On visual perimetry, bilateral homonymous hemianopia was recorded. Plain and contrast enhanced MRI of brain was advised which revealed an extra axial, sellar supra sellar cystic lesion with a maximum diameter of 45 mm (Fig.4 A,B&C). The optic chiasm was pushed by the tumor superiorly.

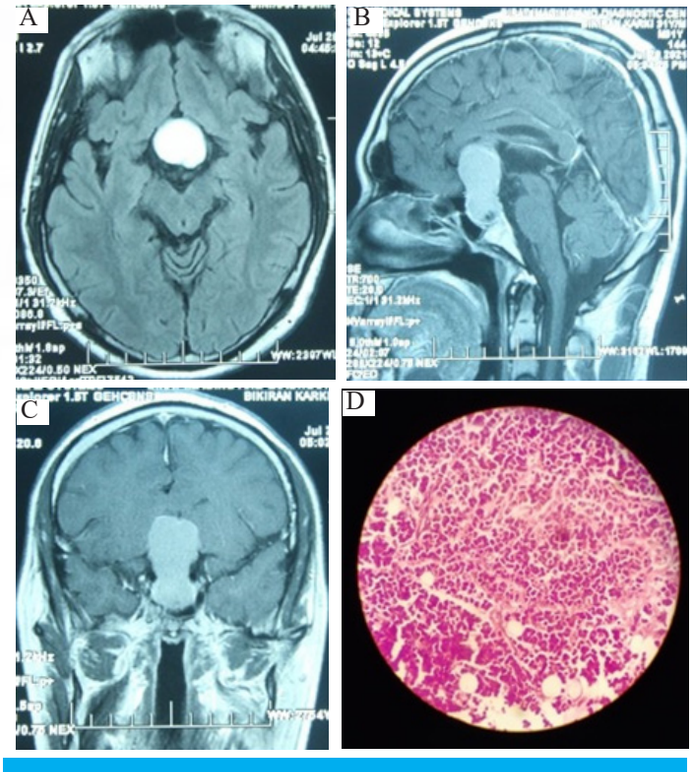

Figure 4: Preoperative MRI showing cystic lesion in sellar-suprasellar lesion with a maximum diameter of 45mm (A,B & C). The optic chiasm was pushed superiorly by the tumor histology (D).

Operation and postoperative course

Taking into consideration of the cystic consistency and whereabouts of the lesion, a trans nasal trans septal trans sphenoidal approach was plotted. With he patient in supine position and head fixed using a 3 pin Mayfield neurosurgical skull clamp, the right nostril was approached and amucocutaneous incision was made. A bivalve speculum is placed, vomer identified and was removed using a chisel and hammer.The ostia of the sphenoid sinus was encountered and the sinus was opened using Kerrison's rongeur. A thorough exenteration of sphenoid mucosa was performed.Neuronavigation guided trajectory was followed to reach the floor of the sella turcica and confirmed by the anatomic emblem visualized under microscope. The sellar floor is then removed with to expose the dura mater.Dural incision was made. A gush of cystic fluid was aspirated and the capsule was excised in piecemeal and a gross total resection was achieved with adequate decompression of the optic apparatus. Histopathological analysis revealed a diagnosis of Adamantinomatous Craniopharyngioma, WHO grade I (Fig.4D).

DISCUSSION

A benign paradigm of neoplasm, presumed to arise from the embryonal remnant of Rathke's cleft, craniopharyngioma entail an approximately 2%-5% of all primary brain tumors indulging an incidence of 0.13 cases per 100,000 people every year. Remarkably, they are found to have a bi-modal age of presentation among children aged 5-14 and adults aged 55-65 years.[9] Despite an escalating refinement in neurosurgical armory, resection of this pathology is lamentably not unsolicited with a higher rate of mortality, morbidity and recurrence. Underpinned by the location, their alliance with the pituitary stalk, or their whereabouts along the vertical hypophyseal axis, a myriad of skull base approaches have emerged over time to clinch entrance to the sur-gical corridor leading to craniopharyngioma, each with its own benefits and detriments.

The pterional (Frontotemporal) approach is the customary, yet a credible approach for the surgical treatment of craniopharyngioma involving primarily the suprasellar cistern.[10] The supremacy of this approach is accredited by the shortest distance to the suprasellar region of all the trans cranial approaches. Considering the exquisite corridor furnished by this approach, it is prudent to maneuver craniopharyngioma of the intrasellar, suprasellar, prechiasmatic, and retrochiasmatic regions. It is judicious to opt to the pterional approach for a pre fixed chiasm, owing to the fact that the tumor can be resected beneath the chiasm. Nonetheless, a pitfall to this approach is the slender outlook of the contralateral opticocarotid triangle and the contralateral retro carotid space. On top of that, when the lamina terminalis is accessed through a pterional craniotomy, the oblique trajectory of this passage makes it strenuous to envision the posterior part of the third ventricle, especially the ipsilateral wall of the hypothalamus.[11] This places the columns of fornix, supraoptic nuclei, organ vasculosum, and tuber cinereum at peril for retraction injury or injury to perforators.

The Fronto Temporo Orbito Zygomatic Approach (FT-OZ) is fundamentally a pterional craniotomy, with simultaneous removal of the supraorbital rim, zygomatic arch, or both.[12,13] It is applicablefor resecting craniopharyngioma with significant suprasellar extension in view of the fact that it offers a revamped inferior-to-superior ("looking-up") view to the hypothalamic and suprasellar region. Furthermore, it is exceptionally effective in approaching these lesions that have notable superior appendage into the third ventricle. By detaching the orbital rim and lateral sphenoid region, the obstructing bone that typically limits adequate exposure of the superior third ventricle through the lamina terminalis or underneath the A1 portion of the anterior cerebral artery is evaded.[14,15] Even so, the hindrance of this approach is an observation and dissection of the contralateral upper side of the tumor in the hypothalamus and preserving the pituitary stalk.

The minimally invasive, transsphenoidal resection is advocated for craniopharyngiomas that engage both sellar and suprasellar regions, especially if the sella turcica is enlarged.[16,17] Hence, it is considerate for intrasellar and subdiaphragmatic genre of craniopharyngiomas. An intrasellar location with enlargement of the sella turcica and a rounded suprasellar extension indicates a subdiaphragmatic craniopharyngioma.[3] Suprasellar extensions of tumors may be readily removed with the transsphenoidal approach, given that they are primarily cystic and not solid. A heterogeneous pathology with a blend of both solid and cystic ingredients in the intrasellar portion can be removed with the transsphenoidal approach. However, this approach may not be convenient for cases in which there is remarkable lateral extension. Such are the cases where a frontotemporal or a combined approach may be inevitable. As a general rule, we would rather not employ the transsphenoidal approach as the primary one if the sphenoid sinus is not favorably pneumatized.

The transsphenoidal approach endeavor a mid-line exposure, sanctioning a dissection in the space around the optic nerves, with a steer clear of brain retraction and some of the auxiliarydownside of transcranial surgery.[18]

Unfortunately, the propensity for CSF leakage was slightly soaring for craniopharyngioma removal compared with other standard transsphenoidal procedures.[19] Over and above that, the likelihood of visual injury was found to be reduced when compared with craniotomies performed to treat similar lesions.[10] Craniopharyngiomas involving the sella turcica are particularly amenable to the transsphenoidal approach taking into account the cystic and friable complexion of this ailment.[19] Furthermore, craniopharyngiomas located within this region do not percolate through surrounding structures, making tumor debulking and capsule dissection from the optic chiasm, hypothalamus, and pituitary stalk accomplishable.[19] This regional hallmark is in stark contrast to those infundibular craniopharyngiomas that are on numerous occasion calcified and those that are intraventricular are almost always solid.[20]

The supraorbital keyhole approach affords distinct advantages compared to the standard craniotomies.[21] The intracranial optical horizon broadens with increasing distance from the keyhole which is the fundamental basis of the keyhole concept.[22] Foremost, there is minimal brain exposure to the air and accidental surgical trauma. Moreover, the brain retraction is negligible or absent, which significantly decreases the technique affiliated surgical anguish and shortens the hospitalization. The nerve and vascular supply to the temporal and frontal muscles are taken care of with an excellent cosmetic result. The surgical procedure is remarkably facilitated compared to the standard techniques. The duration of craniotomy and closure is shortened significantly to about 30 minutes respectively.

Lately, endoscopic endonasal surgery has been revolutionized as the preferred surgical intervention to manage craniopharyngioma as it offers a safe, direct, and wide surgical view to the pituitary stalk or diaphragm from where the tumor emanates.[23,24] However, the tumor with pre-fixed optic chiasm has narrow access and poor visibility. Furthermore, there is an additional risk of mutilation to the hypothalamus.[25] and the A Com penetrating branches. Besides, traction has also high risk of damage to the posterior communicating artery and its perforators.

By dint of the anatomical layout and its alliance with indispensable neurovascular structures within its vicinity, including the hypothalamus and optical apparatus, resection of Craniopharyngioma is associated with either higher rates of mortality and recurrence or a lower rate of resection despite a significant glimpse of stride in the dexterity of neurosurgical skills in the modern day.[2,26]

The rate of radical resection of craniopharyngioma is found to be in a range of 40%-90%, with a retrochiasmatic location, larger size, calcification> 10% with an extension into the third ventricle. Over and above that, the recurrence was reported to be significant prognostic factors that negatively affect the extent of resection.[5] In our series, GTR was achieved in 14 patients, STR in 2 patients and NTR in 4 patients.

CONCLUSION

Craniopharyngioma, despite being a rare entity, we are bound to come across this pathology sooner or later. A diverse array of surgical approaches has been evolved over time but the preference is guided by the chronicles of the neoplasm defined by its locus and its kinship with the neurovascular structures within its proximity. A thorough scrutiny of the neuropsychological, clinical, endocrinological and imaging parameters are mandatory to achieve a convenient aftermath. The extent of resection of the tumor is sanctioned by the dimension and construct of tumor, its affiliation with contiguous neurovascular architecture. An aggressive tussle in the midst of surgical endeavor to set sights on complete resection of tumor is liable to falter into a repugnant after-effect. Complication rate is high if we aggressively attempt to completely resect tumor.

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