### 1.1 Left Medial Temporal Gangliogioma with Refractory Temporal Lobe Epilepsy – Lesionectomy with Selective Amygdalo-hippocampectomy

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#### Overview

Brain tumors associated with epilepsy are often located in the cortex or at the grey–white matter junction. When the lesion is located in the temporal lobe, its direct or indirect effects on the hippocampus may cause seizures. The lesion location may interfere with the cortical afferent and efferent connections and lead to relative deafferentation of a certain cortical area that has intrinsic epileptogenicity. Small hemorrhages in and around the tumors may also cause the occurrence of hemosiderin deposits, which are highly epileptogenic. These hemosiderin deposits may also be responsible for seizures in some patients. It has been shown with the intracranial electroencephalography (EEG) recording that approximately one-half of patients with neocortical temporal tumors have independent epileptogenic areas in ipsilateral mesial structures. It is well known that the hippocampus is a very epileptogenic structure and may even serve as a focus for secondary epileptogenesis. Therefore, significant attention has been given to the mesial temporal structures like the hippocampus and amygdala in patients suffering from temporal lobe epilepsy (TLE) even if the lesion is located in the temporal neocortex. Resection of these mesial temporal structures along with the lesion may be needed to obtain a seizure-free outcome. The indications for resection of adjacent mesial temporal structures in patients with lesional TLE are highly controversial. If the tumor directly involves the mesial temporal structures, then the surgical decision is relatively straightforward. If the mesial temporal structures are not directly in the lesional zone, however, then the risks and benefits of the resection of mesial temporal structures should be assessed carefully.

The surgical outcome in tumor-related pediatric patients with TLE is much better than in some patients with other lesional epilepsy (such as cortical dysplasia). In a large study, the best seizure control rate was seen in children with TLE secondary to a neoplasm (an 88 to 92% Engel class I and II outcome). The complete resection of tumor is the most significant factor in obtaining a seizure-free outcome. Luyken et al., noted that a seizure-free outcome was the maximum in patients harbouring a gangliogioma or an oligodendroglioma (> 90%) compared with those with a pilocytic or a grade II astrocytoma (61% and 66%, respectively). Minkin et al., stressed the significance of the procedure of amygdalohippocampectomy in the treatment of temporal dysembryoplastic neuroepithelial tumors (DNETs) and recommended an extensive presurgical evaluation in this patient group.
Anatomical Principles

Fig. 1 The coronal schematic diagram showing the anatomy of the medial temporal lobe.

Fig. 2 The operative view of the surgical anatomy of the medial temporal lobe. It is important to locate several anatomical landmarks and structures before proceeding to resect the mesial temporal structures. The hippocampus, fimbria, lateral ventricular sulcus, collateral eminence, choroid plexus, choroidal fissure, inferior choroidal point, and amygdala need to be fully exposed and can be distinctly recognized at this stage. The hippocampus lies over the parahippocampal gyrus and has a short, wide head that continues with a gradually narrowing body and tail. The tail makes a backward–upward turn at the trigonal level around the posterior cerebral peduncle. The anterior portion of the hippocampal head blends into the posterior uncus and amygdala. The hippocampus can be easily recognized between the collateral eminence and the choroidal fissure. The medial border of the hippocampus is lined by the choroid plexus over the choroidal fissure and the choroidal point at the most anterior part. If the choroidal plexus is lifted gently upward and medially, the choroidal fissure and fimbria are fully exposed. Retraction of the choroid plexus laterally over the hippocampus exposes the stria terminalis. When the anterior end of the choroid plexus is pulled backward, the velum terminale and the choroidal point at the tip of the posterior uncus can be visualized. The anterior choroidal artery (AChA) runs across the ambient and crural cisterns near the choroid plexus. It pierces the arachnoidal plane to supply the choroid plexus at the inferior choroidal point by giving rise to numerous branches. The anterior fimbria and stria terminalis join to form the velum terminale and create the anterior border of the choroidal fissure where the inferior choroidal point is also located. The fimbria is a narrow and flat band covering the medial border of the hippocampus. It is located just above the dentate gyrus and continues as fimbria of the fornix posteriorly. The temporal horn should be fully unroofed to expose the most anterior part of the temporal horn that includes the bulging amygdala, posterior uncus, amygdala–hippocampal junction, and posteriorly the head and body of the hippocampus. The uncal recess is a distinct landmark that separates the head of the hippocampus from the amygdala. The optic radiation is segregated into three bundles around the lateral ventricle: the upper, central, and inferior. The inferior or ventral bundle, known as the Meyer’s loop, travels around the temporal horn (Figure 1). This bundle makes a wide anterior and lateral loop around the temporal horn of the lateral ventricle before curving around the posterior atrium to reach the occipital cortex. These inferior fibers pass into the uncinate region of the temporal lobe and are constituents of the uncinate fascicle located at the limen insula or temporal stem.
Surgical Steps

Fig. 3  An eight-year old boy presented with a semiology of frequent loss of awareness and oral automatism suggestive of complex partial seizures since the age of one year. He also had on-and-off mild bifrontal headache for the same duration. Initially, the seizure frequency was restricted to 2-3 episodes per month and then it increased to 2-3 episodes per week. These seizures were refractory despite the administration of adequate doses of levetiracetam and phenytoin. There was no significant event in the perinatal period and his family history was negative for this type of disease. He had no complaints suggestive of an intracranial space occupying lesion or raised intracranial pressure. Complete physical and neurological examination, including higher mental functions, revealed no abnormality. Scalp EEG recording showed epileptic spikes originating from the left centro-temporal region. Video EEG confirmed the finding and the seizure focus was localised and lateralised to the left mesial temporal lobe. Magnetic resonance imaging (MRI) revealed a T1-weighted hypointense (a) and non-contrast enhancing (b) lesion in the left medial temporal lobe involving the hippocampus, left fusiform gyrus as well as the left parahippocampal gyrus, located in the floor of the temporal horn, that was relatively well-defined and was hyperintense on T2-weighted image, which was suggestive of a low grade glioma (c). It extended posteriorly up to the occipital horn. Post-operative computed tomographic (CT) scan image showed a gross total excision of the lesion (d). As the lesion was medial to the collateral sulcus, a lesionectomy with selective amygdalo-hippocampectomy via the superior temporal sulcus approach was planned. Gross total lesionectomy was done. Histopathology of the lesion was a ganglioglioma grade 1 and biopsy of the hippocampus was normal. There were no postoperative language and memory deficits. Both antiepileptic drugs were continued in the post-operative period with the plan of tapering the medication after 6 months according to our prevailing anti-epileptic medication protocol. The patient remained seizure-free till the last follow-up visit at 4 months after surgery.
The patient is placed in a supine position with his head turned to the contralateral side by approximately 60 degrees and fixed in a Sugita head-frame. The neck is slightly extended by lowering the vertex approximately 15 degrees downwards to make the zygoma and the malar prominence the most prominent point. The occiput is tilted slightly toward the ipsilateral shoulder. This head position places the base of the temporal fossa perpendicular to the horizontal plane. The lateral surface of the temporal lobe will be in a horizontal position, and the entire long axis of the hippocampus will be exposed well to the surgeon with this approach. Thus, this head position creates a good alignment of the medial structures to the surgeon's eyeline and provides an excellent exposure to the uncus-amygdala complex, the whole length of the hippocampus, and the lateral-basal temporal neocortex. The classical left frontotemporal craniotomy is performed. A smoothly curved, question mark-shaped scalp incision is drawn starting just above the zygoma and approximately 10 mm anterior to the tragus, usually posterior to the location of the palpated superficial temporal artery to ensure that the flap is well vascularised by maintaining the artery within it. Three burr-holes are placed: The first one just posterior to the junction between the zygomatic process of the frontal bone and the anterior limit of the superior temporal line, also known as the key burr-hole (as it is the key to the opening of the frontal and temporal fossae, and if required, the orbit); the second burr-hole is placed just above the zygoma; and, the third burr-hole is placed on the superior temporal line approximately 4 to 5 cm posterior to the key burr-hole. A free bone flap is removed. Further bone removal is needed along the floor of the temporal fossa down to the root of the zygoma and towards the apical region of the temporal fossa. This will provide a comfortable access to the infero-basal neocortical region and the temporal pole during the resection. Then the dura is opened in a C-shaped manner, starting from the keyhole site in the frontal region and ending at the temporal pole, thus following the edges of the craniotomy.

At this stage, the exposed area in the surgical field includes the full extent of the Sylvian fissure/superficial middle cerebral vein, the superior and middle temporal gyri, and the upper part of the inferior temporal gyrus. The temporal horn is approached via the superior temporal sulcal approach.
Fig. 6 The temporal horn was found at a depth of approximately 3cm in the line of the superior temporal sulcus. If the surgeon passes the estimated distance and the temporal horn is not in sight, the best strategy is to redirect the dissection. The two most common reasons for not being able to find the ventricle are either the placement of the entry point of the corticectomy too anteriorly; or, due to the direction of the dissection being either too medial or too lateral. At this stage, the appropriate strategy is to redirect the dissection towards the floor of the middle fossa but not towards its medial aspect. The dissection is then deepened toward the floor of the middle fossa until the grey matter is encountered on the adjacent occipitotemporal (or fusiform) gyrus. Then the dissection is redirected again, this time medially into the white matter until the temporal horn is entered, which is confirmed by the egress of cerebrospinal fluid through it.

Fig. 7 Inside the temporal horn of the lateral ventricle was the well defined, greyish-white tumor tissue involving the lateral part of the hippocampus. The collateral eminence was also involved by the tumor and could not be defined separately from it. The tumor was extending posteriorly in the fusiform gyrus, parallel to the tail of the hippocampus, curving to the region of the brainstem. The tumor was soft and suckable in consistency and minimally vascular.

Fig. 8 With the help of the ultrasonic suction aspirator, the tumor was excised in a piecemeal manner until gross-total excision occurred, leaving the hippocampus intact. The intraoperative squash histology revealed the lesion to be an intermediate grade glioma. The hippocampus, fimbriae, lateral ventricular sulcus, collateral eminence, choroid plexus, choroidal fissure, inferior choroidal point, and amygdala were fully exposed and distinctly recognized.
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Fig. 9 The anterior and medial dissection of the hippocampus involves locating the intraventricular landmarks and then resecting the mesial temporal structures. The latter starts with an incision on the lateral ventricular sulcus that is the demarcation line between the collateral eminence and hippocampus. In this patient, however, the lateral ventricular sulcus was infiltrated by the tumor and could not be defined. After tumor excision, therefore, the medial hippocampal dissection was started from the inferior choroidal point.

Fig. 10 The dissection of the hippocampus is continued medially toward the tentorial edge until the piamater along the medial border of the parahippocampus and the hippocampal sulcus encountered. The hippocampal sulcus is a very critical landmark in this procedure and should be fully visualized. It separates the hippocampus proper from the subiculum. The subiculum constitutes the most medial part of the parahippocampus bulging into the middle incisural space.

Fig. 11 At this stage, the hippocampus is further retracted laterally with the suction, and the hippocampal sulcus is exposed as a two-layered pial folding with several tiny arteries running between the pial layers. The hippocampal arteries and arising arterioles (Uchimura arteries) are located within the hippocampal sulcus. These thin hippocampal arteries mostly form a group of 2–6 thin vessels arising from the anterior choroidal artery and the P2 segment of the posterior cerebral artery close to the free edge of the tentorium. After gaining a satisfactory exposure of the hippocampal sulcus, the hippocampal arterioles are coagulated with a fine-tipped bipolar forceps and divided one-by-one using microscissors. The head of the hippocampus is completely dissected subpially from the underlying pia and lifted upwards and mobilized posteriorly. This way, the whole hippocampus and the underlying part of the parahippocampus are dissected back to the hippocampal tail.
The tail of the hippocampus is resected with bipolar coagulation at its upward turn behind the quadrigeminal plate. The hippocampus is then removed en-bloc. The final step of the procedure is the resection of the amygdala while emptying the content of the anterior uncus. During this stage of the procedure, using strictly a subpial dissection and showing the utmost respect to the pial barriers, are critical steps to protect the underlying vasculature, the third nerve, and the cerebral peduncle. The ultrasonic aspirator in a low setting and a microsuction are very useful tools to resect the uncal content. After completing the resection of the uncus and the anterior basal amygdala, the cerebral peduncle and the third nerve can be seen under the intact pia. Although the anterior and basal borders of amygdala are very well defined, there are no dorsomedial anatomical boundaries of the amygdala. Therefore, it is more challenging to define the dorsomedial resection borders of the amygdala. The M1 segment of the middle cerebral artery, which can be seen subpially, corresponds to the anterior–superior border of the amygdala. The line extending from the anterior tip of the temporal horn to the angle of the artery at the limen insula makes the anterior–superior border of the resection line of amygdala. At this stage, the tentorial edge, third nerve, internal cerebral artery (ICA), posterior cerebral artery (PCA), lateral edge of the midbrain between the cerebral peduncles, and tectum can be seen under the pia in the ambient and crural cisterns.

After ensuring hemostasis, the surgical cavity is filled with warm saline and the dura is closed in a watertight fashion with 4–0 vicryl sutures. The bone flap is replaced with microplates, and the temporal muscle and its fascia as well as the galea aponeurotica and the skin are closed.
Complication Avoidance

Avoiding Accidental Entry into the Insula

One has to be extremely careful during the initial dissection so as not to go above the temporal horn in a middle temporal gyrus approach. This avoids an accidental entry into the insula.

Avoiding Injury to the Anterior Choroidal Artery or the P2 Segment of the Posterior Cerebral Artery

Extreme care should be practised while dissecting the taenia fimbriae from the choroidal fissure, to avoid an inadvertent anterior choroidal artery or a P2 segment of the posterior cerebral artery injury. Coagulation or injury to the medial piamater and any small vessel especially in posterior relation to the uncus should be avoided as occasionally this may also lead to an inadvertent injury to the anterior choroidal artery.

Avoiding Visual Field Defects

The temporal horn is opened anterolaterally, preferably in front of the region of the choroidal point, to avoid injury to the Meyer’s loop.

Tips and Pearls

• Various surgical techniques to identify the temporal horn include the following:
  ➢ The temporal horn starts approximately 3 cm behind the temporal tip, and the average distance between the surface of the superior temporal gyrus and the ventricle is approximately 31 to 34 mm. The subcortical dissection should aim for the temporal horn at a point on the superior temporal sulcus, approximately 3.5 cm behind the tip of the temporal pole.
  ➢ Alternatively, the temporal horn can be found after completing the resection of the anterolateral temporal lobe, without initially attempting to locate the temporal horn. In this case, the uncus is located first by following the tentorial edge anteromedially. When removal of the uncus is completed, its posterior segment will open up and expose the tip of the temporal horn automatically.
  ➢ Use of a neuronavigation system to assist in the localization of the temporal horn is an option.
  ➢ The temporal horn can be located just 10–12 mm dorsal to the collateral sulcus.
  ➢ After complete excision of the temporal neocortex, the white matter dissection at the central part of the middle temporal gyrus will also open the temporal horn.
• Keeping the suction and/or the ultrasonic aspirator settings low during the subpial dissection is an essential component of the procedure as pial preservation is of utmost importance.
• Hippocampal end arteries are coagulated and divided as close to the hippocampus as possible.

Suggested Readings