Successful Resection of Hypothalamic Hamartoma with Intractable Gelastic Seizures - by Transcallosal Subchoroidal Approach

Krishnapundha Bunyaratavej MD*,**, Chaichon Locharernkul MD*, Supatporn Tepmonkol MD*, Sukalaya Lerdlum MD*, Shanop Shuangshoti MD*, Surachai Khaoropham MD**

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* Chulalongkorn Comprehensive Epilepsy Program (CCEP) under the Patronage of Professor Doctor HRH Princess Chulabhorn
** Division of Neurosurgery, Department of Surgery, King Chulalongkorn Memorial Hospital

A 19-year old female with intractable gelastic seizures was found to have 0.7 x 1.8 x 1.8 cm elliptical mass on the floor of the third ventricle. The signal intensity on the Magnetic Resonance Imaging (MRI) was consistent with the Hypothalamic Hamartoma (HH). Ictal EEG demonstrated rhythmic 7 Hz waves over Fp2, F4, and C4 with spreading to the right temporal region and then bilaterally. Ictal Single Photon Emission Computerized Tomography (SPECT) showed hyperperfusion at hypothalamic and medial frontopolar regions. The patient underwent surgical resection using Trans Callosal Subchoroidal Approach (TCSA) to the third ventricle. Pathological finding confirmed the diagnosis of hypothalamic hamartoma. Following the operation, she has been seizure free up to 12 months. Thereafter, provoked seizures seldom occurred and there has been improvement in her memory, emotional control and independence. This appears to be the first report of this surgical approach for HH, which is less likely to disturb memory function compared to previously described interforniceal approach.

Keywords: Hypothalamic hamartoma, Gelastic seizure, Transcallosal, Subchoroidal, SPECT, Epilepsy

Case Report

A 19-year-old Thai female was first noted to have 1-minute duration laughing at the age of 2 months. Since then, she has had several spontaneous laughing spells on a daily basis. At the age of 4 years, she started
having episodes of unmotivated laughter followed by Generalized Tonic Clonic (GTC) seizures lasting 3-5 minutes. She had 2-3 attacks per year. At 9 years of age, the laughing episodes were occasionally followed by unresponsiveness, blank staring. She had daily hand automatism lasting 1-2 minutes. Her birth history, growth, and development were unremarkable. She had normal intellectual function and behavior. Physical examination showed no abnormality. The patient underwent comprehensive presurgical evaluation. High resolution MRI of the brain revealed a midline elliptical lesion, 7 mm. in width, 1.8 cm. in height and A-P dimension, occupying the inferior part of the third ventricle. The mass was slightly hypersignal on T2WI and FLAIR (Fig. 1a-d). Electro EncephaloGram (EEG) showed rare bilateral T3T4 interictal discharges. Ictal EEG showed rhythmic 7 Hz waves over Fp2, F4, and C4; discharges then spread to the right temporal region; and then bilaterally. Early ictal SPECT using Tc-99m Ethyl Cysteinate Dimer (ECD) injected 24 seconds after the clinical onset of a gelastic seizure showed hyperperfusion of hypothalamus and medial frontopolar regions. Interictal SPECT demonstrated hypoperfusion of the hypothalamic area (Fig. 2). She has been on several antiepileptic drugs without satisfactory improvement. Prior to operation, she experienced more than 10 laughing spells per month followed by Complex Partial Seizure (CPS) or GTC.

Fig. 1a-d Coronal and sagittal MRI reveal hypothalamic hamartoma in the third ventricle with broad attachment to tuber cinereum
At operation, the patient was placed in the supine position with head in rigid fixation in zero degree rotation and her neck was slightly flexed. Right frontal craniotomy was made with bone flap extending across the midline; posterior border extended just posterior to the coronal suture. Dura was opened based on the midline. Under microscope magnification, interhemispheric fissure was gradually split; the dissection was deepened until the corpus callosum was reached. Entry to the frontal horn of the right lateral ventricle was made through a 2.5 centimeter opening on the anterior corpus callosum. Once the right frontal horn was entered, the foramen of Monroe was identified. The size of the foramen was normal and did not allow adequate visualization of the third ventricle (Fig. 3a). Therefore, choroid plexus emerging from the foramen of Monro was gently elevated and retracted toward the midline. The tethered thalamostriate vein at the posterior limit of the foramen of Monro and the velum interpositum were divided. With further retraction of choroid plexus toward the midline, third ventricle and its contents were clearly visualized (Fig. 3b). There was an oval shiny white mass attached to the floor of the third ventricle, and to a lesser extent, to walls of the ventricle. The mass was approximately 2 cm in anterior-posterior dimension and 1 cm in transverse dimension. The massa intermedia was identified a few millimeters posterior to the mass. The mass was removed flush with the inner walls of the third ventricle.

**Results**

Postoperative period was uneventful. There was no neurological deficit. Pathological findings confirmed the hypothalamic hamartoma. The patient had no seizures during the 12 months of post operative period until rare provoked gelastic seizures occurred (Engel Class IIa)\(^1\). Post operatively; there have been improvements in memory, emotional control, independence and her quality of life.
Discussion

HH may present with Gelastic Seizure (GS), Precocious Puberty (PP), or in combination\(^{9,12}\). GS usually start in the early years of life (< 1 yr) and may be followed by drop attacks, complex partial seizures, and generalized motor seizures\(^{3,5,6,18}\). Other features include mental retardation or behavioral abnormality. Palmini, et al reported all 13 patients in their series also had delayed developmental milestones either from birth or after the onset of seizures\(^{18}\). The present case had epilepsy at a young age of onset, but without any other associated abnormality or intellectual impairment, which is interestingly uncommon.

There has been some evidence supporting the epileptogenicity of the HH\(^{13,19}\). The early injected Tc-99m ECD ictal SPECT in the present study in one GS without CPS or GTC demonstrated hyperperfusion at the midline hypothalamic region compared to the ictal injection, suggesting the HH itself as being the ictal onset zone for GS. Hyperperfusion within the hamartoma and hypothalamic regions has also been shown by others using ictal SPECT\(^{19-21}\). Moreover, stereotactic depth EEG recordings have consistently demonstrated that the ictal discharges of GS were originated and confined within the hamartoma, and stimulation of the HH could produce gelastic episodes\(^{19,22,23}\). These findings suggest that GS associated with HH is generated in the hypothalamus. The other types of seizures such as CPS, atonic or GTC, were found to be associated with cortical involvement, either frontal or temporal lobes\(^{22-23}\).

The frontomesial area is involved in triggering motor component of laughter in many studies. Restricted pericentral premotor cortex has been found to underlie ictal laughing in patients without HH\(^{26-28}\). Ictal SPECT in the presented patient also showed hyperperfusion at the frontopolar region, which is likely to represent the symptomatogenic zone for ictal laughter. The increased perfusion to the occipital lobe is considered physiologic uptake usually found in Tc-99m ECD studies as described elsewhere\(^{29,30}\).

Since Northfield and Russet reported the first successful surgical treatment of HH in 1967\(^{2}\), surgical series have been published with varying outcomes\(^{1,5,13-16,18,20,31-33}\). The surgical approaches for GS vary among neurosurgeons. These approaches can be divided into suprahypothalamic and infrahypothalamic. Suprahypothalamic techniques include transcallosal resection\(^{33}\) and endoscopic disconnection\(^{16,20,31}\). Infrahypothalamic techniques include frontotemporal\(^{4,18,32,33}\), subtemporal\(^{41}\), subfrontal\(^{21}\) and translaminar terminalis\(^{5}\) approaches.

Early surgical procedures were limited to infrahypothalamic routes\(^{1,5,14,32,33}\). With the advent of MRI, the configuration of HH and the relation of HH to the hypothalamus can be visualized with unprecedented details\(^{40}\). This has led to suprahypothalamic approaches by some authors\(^{13,16,31}\).

For the HH with most of the lesion located in the third ventricle, corresponding to Delalande type II, the suprahypothalamic approach appears to be most suitable\(^{13,16}\). Rosenfeld elegantly pioneered transcallosal interfoniceal approach for intraventricular HH with good outcomes\(^{13}\). However, transient memory loss due to bilateral fornical injury is a potential complication with this approach varying from 20-57% depending on the nature of the lesions\(^{34,36}\). Harvey, et al reported short-term memory impairment in 14 out of 29 patients following transcallosal interfoniceal approach, with four being persistent\(^{37}\).

Delalande, et al reported on 17 HH patients. Nine patients underwent endoscopic disconnection and two had an intraventricular lesion (Delandale type II). Seizure outcomes according to Engel classification\(^{17}\) were I and III respectively. For the patients with combined intraventricular and infrahypothalamic HH (Delalande type III), Engel class I were obtained in four out of nine cases using combined pterional and endoscopic approaches. This might be attributable to the extent of resection versus the boundary of epileptogenic zone beyond the HH.

In the authors’ opinion, the TCSA appeared to be feasible and effective for strictly intraventricular HH (Delalande type II HH). The authors decided to use this technique for resection of the lesion in the presented patient as it provided superior visualization of the intraventricular HH compared to other infrahypothalamic approaches. It also required much less manipulation of fornices compared to Transcallosal Inter Forniceal Approach (TIFA), thus minimizing the possibility of postoperative memory deficit. Although, TSCA enters the third ventricle slightly more lateral compared to TIFA (Fig. 4a), with the brain relaxation from the cerebrospinal fluid drainage and changing the angle of the microscope, the HH, third ventricular wall, and HH interface can be visualized in small and medium size HHs. However, in the large HH, the HH interface on the opposite site could be obscured by the column of the fornix (Fig. 4b), which makes TIFA more suitable (Fig. 4c).

Although resection of the thalamostriate veins has been a concern by many authors\(^{38-40}\), unilateral resection is well tolerated in most cases\(^{34,41,42}\).
Successful surgery without complication is demonstrated in the presented case.

Surgical outcome of the patient with HH associated with GS varies among series\(^{1,2,5,13-16,18,31-33}\). Potential complications of resection of HH include thalamic infarct, third nerve palsy, hyperphagia, hypopituitarism, diabetic incipidus and hemiparesis\(^{6,18,31}\). The extent of resection correlates directly to the seizure control. Most of the patients who have become seizure free have had total or near total resection of the HH\(^{13,14,16,18,32}\). The patients who remain seizing after resection are probably due to incomplete removal of the epilepto-genic zone. Alternative hypotheses are secondary epileptogenesis or more diffuse brain lesion indicated by the existence of HH\(^{6}\).

In the presented patient, good seizure control can be attributable to several factors. Firstly, the presurgical evaluations, including EEG and ictal SPECT,
indicated the HH to be the ictal onset zone. Secondly, the circumscribed configuration, the midline symmetrical, and the almost separable intraventricular location of the HH were amenable to complete resection. Thirdly, the convergence of the GS and the SPECT finding help in confirming the role of HH in ictal genesis. Lastly, the absence of associated abnormality and normal intellectual function in the presented patient may also be responsible for the good surgical outcome.

**Conclusion**

The ictal SPECT findings in the presented patient confirmed the likeliness of HH as the ictal origin and the mesial frontopolar region as the symptomatic-genic zone for GS. Although, the complications associated with surgical resection of hypothalamic hamartoma may be disabling\(^{(6,18,31)}\), surgical treatment still plays an important role in HH associated with medically intractable seizures. The surgical option seems justified in the presented patient who had no other associated abnormality and solely, refractory epilepsy impaired her quality of life. The transcallosal, subchoroidal approach has been demonstrated to be an effective surgical technique to the HH located in most of the third ventricle. The authors’ key to successful resection is the profile of the patient and the surgical strategy chosen\(^{(18)}\).

**References**


รายงานการผ่าตัดก้อนไฮโปธาลามิคฮามาโตมาในผู้ป่วยโรคคลื่นแบบเจลลาสติกโดยการผ่านคอร์ปัสแคโลสัมและลอดใต้คอรอยด์

ภูมิพันธุ์ บุณยะวัชร, ชัยชน โลวเจริญกุล, สุภัทรพร เทพมงคล, สุกัลยา เลิศล้ำ, ชนพ ช้างโยธิน, สุรชัย เคารพธรรม

ผู้ป่วยหญิง อายุ 19 ปี ป่วยเป็นโรคคลื่นแบบเจลลาสติกซึ่งดื้อต่อการรักษาทางยา จากการตรวจสมองด้วยเครื่องตรวจสนามแม่เหล็กพบมีก้อนขนาด 0.7 x 1.8 x 1.8 เซนติเมตรที่บริเวณกลางของโพรงน้ำสมองที่ 3 โดยลักษณะเข้ากับก้อนไฮโปธาลามิคฮามาโตมา จากการตรวจคลื่นสมองขณะชักพบคลื่นความถี่ 7 เฮิร์ทที่บริเวณ Fp2, F4 และ C4 กระจายไปถึงบริเวณ Fp1, F3 และกระจ่างไปทั่วสมอง จากการตรวจด้วยสเปกสแกนพบการเพิ่มการเรืองสารที่บริเวณไฮโปธาลามัสและบริเวณกลีบสมองส่วนหน้า ผู้ป่วยได้เข้ารับการผ่าตัดฮามาโตมาโดยผ่านคอร์ปัสแคโลสัมและลอดใต้คอรอยด์ หลังผ่าตัดผู้ป่วยไม่มีอาการชักเลยเป็นเวลา 12 เดือน หลังจากนั้นผู้ป่วยไม่มีอาการชักแน่นอนหลังผ่าตัดผู้ป่วยมีความจำ การควบคุมอารมณ์และการช่วยเหลือตัวเองดีขึ้น รายงานนี้เป็นรายงานแรกในการผ่าตัดฮามาโตมาโดยใช้การผ่าตัดที่เข้าระหว่างโฟนิกส์ซึ่งมีการรายงานมาก่อนหน้านี้