Case report

Temporal lobe epilepsy with hypothalamic hamartoma: a rare case

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Refractory gelastic seizure is one of the most common clinical manifestations in patients with hypothalamic hamartoma (HH) and HH is usually regarded as the epileptogenic focus. A young female patient with a small HH and refractory seizures is reported here. However, both the seizure semiology and results of electroencephalogram monitoring indicated the right temporal region was the epileptogenic focus. Thus a standard right anterior temporal lobectomy was performed while the hamartoma preserved. There was a marked improvement in both seizure frequency and quality of life during a 13-month follow-up. The outcome supported the concept that independent epileptogenic focus outside of the hypothalamus might occur in patients with HH.

Hypothalamic hamartoma (HH) is a congenital malformation of the hypothalamus that may be asymptomatic or manifest with gelastic seizures (GS) or other kinds of seizures, mental retardation, behavioral abnormality and central precocious puberty (CPP). In HH patients with typical symptoms of GS and CPP, there was evidence that resection or disconnection of HH could completely eliminate these symptoms. To date, the most widely accepted concept is that the tumor itself is the source of seizures in HH patients. Kramer et al reported a HH case whose video-electroencephalogram (V-EEG) monitoring revealed a well-defined epileptic focus in the left frontal region. Based on the current understanding of the nature of hamartoma-related seizures, the HH was resected while the left frontal lobe spared. A reduction of 70% in seizure frequency was observed.

Since most of the HHs are stationary, surgical treatment is not usually indicated in the asymptomatic patients. In HH patients with seizures, there has rare report of independent epileptogenic focus outside of the hypothalamus. Here we report a female HH patient with refractory seizures. Both the semiology and electrophysiological examinations indicated that the right temporal region was the epileptic focus. Thus a standard anterior temporal lobectomy (ATL) was performed. The patient kept seizure free during the follow-up of 13 months. So, this is another rare case of independent epileptic focus outside of hypothalamus in HH patients.

CASE REPORT

A 25-year-old right-handed, college graduated female had suffered from multiple seizures since the age of 18 years. Most of the episodes were complex partial seizures, which typically began with auras such as an epigastric rising sensation, dizziness and fear. The unresponsive phase often began with staring and automatisms, particularly chewing, lip smacking and hands groping without gelastic component for about 2–3 minutes. She usually had 3–5 seizures per day, sometimes followed by secondary generalized seizures, which often occurred once to twice per month. Postictal cough and amnesia were common. The epilepsy was refractory to antiepileptic drugs including topamax, lamotrigine, and valproate sodium. Her past history was unremarkable. She started her period at the age of 14 years. Her period was regular. On physical examination, her height and weight were within normal range. She had no signs of precocious puberty. Neurological examination was normal. Neuropsychological test showed that she was a little anxious, otherwise healthy. The endocrinological test was normal. Magnetic resonance (MR) scanning revealed a HH in the interpeduncular fossa, adjacent to the floor of the third ventricle. The diameter of the HH is 5 mm. The third ventricle was not distorted. The signal of both temporal lobes was normal (Figure 1A–1C).

The patient thus underwent presurgical evaluation at the Epileptic Center of Beijing Tiantan Hospital. Two episodes were recorded during 24 hours’ V-EEG monitoring: both of the epileptogenic discharges started in the right temporal region (dominantly in F8, T4 channels) (Figure 2A). To confirm the result of scalp-
Figure 1. The imaging examinations of the patient. A–C: Preoperative MR scan revealed a HH located at the interpeduncular fossa (as the black arrow pointed). D, E: The identification of electrodes on CT and X-ray scan. The tip of the 4-contact depth electrode was adjacent to the HH. Six-contact strip electrodes were put on the surface of the right temporal and frontal cortex separately. F: Postoperative MRI scan showed the right anterior temporal lobe and hippocampus were removed.

Figure 2. EEG recordings at different stages. A: The preoperative scalp-EEG showed focal spikes and poly-spike waves in the right anterior temporal region (F8 and T4 channels). The rhythmic waves were relatively normal in frontal and parietal lobes. B, C: The V-EEG results of intracranial electrodes examination. Interictal spikes over the right anterior temporal lobe were revealed (B). The earlier ictal patterns consisting of rhythmic spike activities were also detected in the anterior temporal lobe (C). F1-F6: frontal lobe cortex; T1-T6: temporal lobe cortex; H1-H4: hypothalamic region. D–F: The intraoperative EEG and ECoG results before the ATL. Rhythmic spike or sharp activities over the right anterior temporal cortex were recorded (D). Activities of spike waves were also detected by the depth EEG over the right hippocampus (E). The depth electrode over the HH showed β waves dominantly with low amplitude, no spike components were detected (F). G: Scalp-EEG 3 months after the operation showed normal cerebral electrical activities, no spike wave was detected in 48 hours.

EEG, depth and subdural electrodes were utilized for further evaluation. A 4-contact depth electrode was implanted stereotactically. Because of the size of HH, the electrode was not inserted into it. The tip of electrode was located just above the floor of the third ventricle. Two 6-contact strip electrodes were implanted on the surface of the right temporal and frontal cortex simultaneously (Figure 1D and 1E). Four habitual seizures were recorded in the 72 hours’ V-EEG monitoring. Consistent with the result of scalp-EEG, the intracranial EEG monitoring also showed that all epileptic discharges started dominantly in the right temporal region, not the adjacent region of HH.
Since the semiology of the patient showed most features of medial temporal lobe epilepsy (TLE), and the electrophysiological results indicated the epileptic focus in the right temporal region, so right ATL was performed. The intraoperative electrocorticogram (ECoG) showed multiple spikes on the temporal cortex (Figure 2D). Depth EEG showed marked spikes on the surface of hippocampus (Figure 2E) and no spikes were detected in HH region (Figure 2F). Thus the anterior 4.5 cm of temporal lobe and 3 cm of hippocampus were resected (Figure 1F). The pathology revealed moderate neuronal loss and gliosis in the hippocampus (Figure 3A) and temporal neocortex (Figure 3B).

Figure 3. The pathology revealed moderate neuronal loss and gliosis in the hippocampus tissue (A) and the temporal neocortex (B) (HE, original magnification ×100).

The patient was discharged 8 days after surgery with oral administration of valproate sodium (500 mg, twice a day) continuously. No spikes were detected during a V-EEG monitoring at 3 months after surgery (Figure 2G). The patient remained seizure free during the follow-up of 13 months.

DISCUSSION

Cascino et al reported that HH originated seizures were often mislocalized to the temporal or the frontal lobe due to the fast spreading of spike waves. In this case, temporal or frontal lobectomy did not control the seizures of the patient effectively. Thus, most authors believed even though presurgical evaluation pointed to a distinct cortical region as the source of seizures, the HH resection might alleviate the seizures in HH patients. Based on the previous concept, we initially postulated that HH was the origin of seizures. However, through detailed presurgical evaluation, we finally regarded the right temporal lobe as the origin of her seizures. The reasons are as follows: (1) The semiology: Semiologic features are highly reliable in lateralizing and localizing the seizure foci. Unlike most symptomatic HH patients, this case did not show the typical symptoms of HH including GS, mental retardation, behavioral abnormality and CPP. The only symptom was complex partial seizure with occasional secondary generalization. Her auras and automatisms were typical in medial TLE. Henkel et al reported that the probability of an abdominal aura associated with TLE was 73.6%. The evolution of an abdominal aura into an automotor seizure, however, increased the probability to 98.3%. Thus the semiology was the first clue to localize the temporal lobe as the epileptic focus. (2) The topology and size of HH: HH can be classified into “parahypothalamic type” in which the hamartoma is only attached to or suspended from the floor of the third ventricle by a peduncle; and “intrahypothalamic type” in which the hamartoma invaded or was enveloped by the hypothalamus and the tumor distorted the third ventricle. The HH in this case belonged to parahypothalamic type, since the size of HH was rather small and the third ventricle was not distorted. Arita et al reported that in parahypothalamic type of HH patients, no seizures or mental retardation were observed. Debeneix et al concluded that the clinical presentation of HH depended on its anatomy: small and pedunculated HH were associated with CPP, while large and sessile HH were associated with seizures. The origination of seizures was probably caused by the distortion of the third ventricle. Mahachoklertwattana et al also reported that 16/38 cases with mass diameter >10 mm had seizures, whereas 0/18 cases with mass diameter <10 mm had seizures. According to the size and topology of the tumor, we thought it was not responsible for the seizures in this patient. (3) Electrophysiological findings: Seizures originated in the HH and its adjacent structures have the potential to spread remotely into the temporal or frontal lobe. The characteristics of this discharge is widespread and without specific direction. Harvey et al reported the interictal discharge was bilateral synchronous and generalized when complex partial or generalized seizures evolved from HH. And in some cases, more obvious discharge could be recorded in the ipsilateral hemisphere if the HH was laterodeviated. In this case, the HH was a little deviated to the left. Thus if the tumor was the source of seizures, bilateral synchronous or left dominant discharge might be detected with scalp-EEG and discharge from HH and its adjacent regions should be earlier with depth-EEG. However all EEG results revealed a clear and definite epileptic discharge focus in the right temporal region. Based on the electrophysiological findings, we believed that the HH was asymptomatic and the right temporal lobe was the source of seizures.

We reassured the correct diagnosis and treatment after surgery in 3 aspects: (1) The pathology: The findings of moderate neuronal loss and gliosis in the hippocampus and temporal neocortex were in accordance with the
typical pathology of TLE. (2) The intraoperative spikes decreased obviously in the temporal region after ATL. Postoperative scalp-EEG monitoring showed no spikes anymore. (3) The remission of seizures after ATL.

Palmini et al\[12\] had reported a patient with HH and complex partial seizures who was cured of his complex partial seizures after temporal lobotomy. To our knowledge, this was another report that the tumor was not the origin of seizures and the patient acquired satisfying prognosis after ATL. So, we support that the independent epileptogenic focus outside of the hypothalamus might occur in patients with HH.

REFERENCES


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