Surgically Treated Status Epilepticus due to Large Cortical Tuber and Long-Term Follow-Up Results

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ABSTRACT

We present a case of invasive monitoring of a patient while he was being surgically treated in the status state. Our patient was a 27-year-old male who was hospitalized for frequent seizures, which began after a head trauma at the age of 3 years. Video electroencephalography was performed, and 25 clinical seizures were observed in 24 hours. Cranial magnetic resonance imaging (MRI) revealed a right frontal lesion which was hyperintense in T2-weighted and hypointense in T1-weighted images, and a subependymal nodule. For invasive monitoring, subdural electrodes were placed on the cortex surface via a right frontal craniotomy. The patient was re-operated, and the epileptic zone resection was performed. There was no sign of neurological deficit. Histopathological examination revealed cortical tuber, and the patient was scanned for tuberous sclerosis. There was no sign of tuberous sclerosis in other organs. The diagnosis of our patient was tuberous sclerosis, cortical tuber, subependymal nodule, epilepsy, and intermediate mental retardation. Radiological diagnosis should also be considered. Cortical tuber can be confused with focal dysplasia. Finally, staged resection may be performed as a surgical treatment in some cases.

KEYWORDS: Cortical tuber, Electroencephalography, Frontal lobe epilepsy, Invasive recording, Tuberous sclerosis

INTRODUCTION

Tuberous sclerosis (TS) is an autosomal disease characterized by hamartomas in the brain, skin, heart, eyes, and other organs. The four major intracranial manifestations of TS include cortical tuber, white matter abnormality, subependymal nodule, and subependymal giant cell astrocytoma. Cortical tubers are found in 90% of patients with TS. Cortical dysplasia in TS is mostly associated with intractable epilepsy and learning difficulties (1,7). The major neurological manifestations of TS include seizures, developmental delays, and mental retardation. Epilepsy occurs in approximately 90% of affected patients. Cortical tubers are multiple or solitary, and have a significant role in epilepsy.

We present an epilepsy case caused by a large cortical tuber that was monitored for resistant seizures within status and subsequently treated with surgery in the status state.
Brain magnetic resonance imaging (MRI) showed a central low signal and peripheral iso-high signal intensity on the T1-weighted images, high signal and radially oriented high signal intensity bands in the white matter on the T2-weighted images, and heterogeneous intensity lesion on FLAIR-MRI. One subependymal nodule was distinguished in the right lateral ventricle (Figures 1A–D).

He was operated on for invasive monitoring; a 64-grid and an 8-strip subdural electrode were placed on the frontal cortex and interhemispheric surface (Figures 2A, B).

The patient was monitored for 3 days postoperatively. Video EEG recorded frequent epileptic activity in the G26, G27, and G34 electrodes. On the other hand, less frequent epileptic activity was recorded in the G33, G35, and G50 electrodes. The recorded epileptic activity was in electrophysiological status. During the recording, three clinical seizures were observed (Figures 2C, D). The strip electrode recordings were normal.

After electrodes were removed; an epileptogenic zone resection was performed by making the necessary markings under the subdural electrodes. Resection was performed within the epileptic zone borders (Figures 3A–C).

The patient was extubated 2 days postoperatively. There was no sign of neurological deficit or seizure. His histopathological examination revealed a cortical tuber (Figures 4A–C), and he was scanned for TS. There was no family history. No skin lesions or hamartoma in the eye were detected. Renal ultrasonography and electrocardiography results were normal.

Our patient was diagnosed with TS, cortical tuber, subependymal nodule, epilepsy, and intermediate mental retardation. No genetic tests were performed. Currently, at postoperative 5 years, our patient’s seizure result scale is Engel grade 2A.

## DISCUSSION

The clinical presentations of TS are variable, and diagnostic criteria have been updated on 2012. White matter radial migration lines, subependymal nodules, subependymal giant cell astrocytomas, and cortical tubers are the major clinical diagnostic criteria (8,10).

Cortical tubers have low signal intensity or are isointense at the center on T1-weighted images, and have high signal intensity on T2-weighted images (10). Generally, 90% of the tubers are multiple, and approximately 50% of cases show calcification on computed tomography (5).

A solitary cortical lesion with no other signs of TS leads to a diagnostic dilemma. Di Paolo and Zimmerman (3) stated that possibility of neoplasms, such as ganglioglioma, oligodendroglioma, and low-grade astrocytoma, cannot be excluded.
in such cases. According to Braffman et al., 22% of cortical tubers had gyral expansion, and 57.6% were located in the frontal lobe (2). Cortical tubers are classified as type 1 with smooth surfaces and type 2 with central depressions. In our patient, the lesion was located on the frontal lobe with central depression.

Cortical tubers may be atypical and resemble focal cortical dysplasia (FCD). FCD type IIb and cortical tubers in TS are histopathologically similar; both present as epileptogenic lesions that mostly cause refractory epilepsy. Tubers could be enhanced by gadolinium in 3%-4% of cases (6). Tubers are multiple in approximately 95% of cases, whereas the remaining are solitary (3,5,10). The major clinical manifestations of TS include seizures, mental retardation, and developmental delays. All epilepsy types can occur in TS patients, but seizures may become intractable over time. Cortical tuber lesions have an important role in epilepsy and cognitive outcomes. Goodman et al. suggested that cortical tubers are reliable biomarkers for estimating the clinical severity of seizures and mental retardation (4). Our patient had...
resistant frontal seizures and intermediate mental retardation, with an IQ test score of 55.

Kassiri et al. reported that the number of cortical tubers has an important role in mental retardation among TS patients (7). Pascual-Castroviejo reported that large tubers were more epileptogenic than the small ones (9). Our patient had a large cortical tuber, and invasive EEG recording was performed during the status epilepticus, following which the epileptic focus was surgically resected. The epileptic seizure scale was Engel grade 2A, and the patient had no intractable seizures during a 5-year follow-up period.

CONCLUSION

Surgical treatment should be considered in patients with solitary cortical tubers and epileptic focus. In some patients, invasive recording could be performed again, and epileptic focus can be resected stage-by-stage.

REFERENCES


Figure 4: Histopathological examination of A) vesicular nuclei and giant cells (H&E, x20), B) dysplastic areas where there is no cortical stratification (H&E, x10), and C) giant cells (GFAP, x20).