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Intraoperative electrocorticography and successful focus resection in a case of Sturge–Weber syndrome

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This paper reports a surgically treated case of Sturge–Weber syndrome (SWS) in which the epileptic foci and haemangiomatosis were successfully resected under monitoring of intraoperative electrocorticography. The patient was a 19-month-old female infant who was referred to our hospital because of frequent hemi-tonic–clonic convulsions that were resistant to anticonvulsant therapy. Serial MRI showed progressive atrophy in the left fronto-parieto-temporal lobe, and gyral enhancement by gadolinium corresponded to venous haemangiomatosis of SWS. Three-dimensional reconstruction of the MR images was performed using the Viewing Wand System. Conventional EEG taken before the operation showed slow activity in the left frontal lobe. Intraoperative ECoG revealed spike focus at the posterior temporal cortex to the margin of the haemangiomatosis. Lesionectomy with lobar corticectomy of the total frontal and parietal lobe and part of the temporal lobe was performed. The epileptogenic focus detected by ECoG in the posterior temporal lobe was also resected. In post-excisional ECoG, epileptogenic activities had disappeared. The patient had hemiparesis and hemihypesthesia just after the surgery, but gradually recovered from the paresis and almost has normal motor function except for right-hand clumsiness up to 1 year after surgery. The present study demonstrated that lobar corticectomy of the haemangiomatosis-affected cortex with resection of the neighbouring epileptogenic focus is a good surgical alternative even if a haemangiomatosis of the SWS affected multilobar corti of the hemisphere.

Key words: Sturge–Weber syndrome; ECoG; spike and wave; corticectomy; neuronavigation.

INTRODUCTION

Sturge–Weber syndrome (SWS) is a congenital neurocutaneous syndrome characterized by: (1) a vascular naevus, usually unilateral, and typically a port-wine stain in the distribution of the trigeminal nerve and at times involving the eye and producing glaucoma; (2) typical intracranial calcification in the ipsilateral cortex underlying angiomatous malformation of the meninges; (3) seizures, generalized or focal, on the contralateral side; (4) focal neurological deficits such as spastic hemiparesis with or without atrophy, or hemianopsia thought to be secondary to atrophy of the involved cortex; and (5) frequent mental retardation. Thus, patients with SWS develop severe convulsions, almost all of which are resistant to anticonvulsant therapy. If seizures begin in early infancy, there is a high risk of progressive mental retardation with a downhill course, accompanied by severe hemiplegia and intractable seizures[1–5]. Venous haemangiomatosis of SWS leads to a reduction in the blood flow through the involved hemisphere with consequent cerebral atrophy and seizures which affect the contralateral uninvolved hemisphere, impairing its function and leading to severe mental retardation. Although the function of the remaining normal hemisphere may be preserved by prompt surgical treatment, the results of surgery are not always successful.

Here we report the successful treatment of SWS using intraoperative ECoG examination. To the best of our knowledge, there have been no previous reports on intraoperative and post-excisional ECoG for treating SWS.

CASE REPORT

A 19-month-old female infant was suffering from anticonvulsant-resistant right-side hemiconvulsions. There were no notable problems during the mother's pregnancy or at birth. At the age of 3 months, she had a hemiconvulsion and was referred to another hospi-

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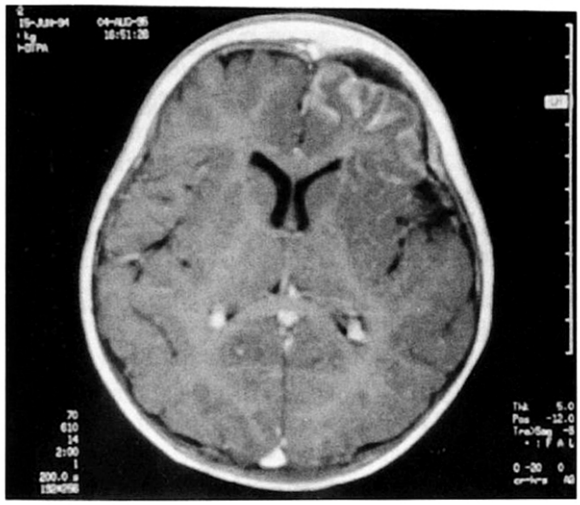


Fig. 1: Gd-DTPA enhanced MRI showing atrophy in the left frontal lobe and enhancement of the gyral surface of the left fronto-parieto-temporal lobe.

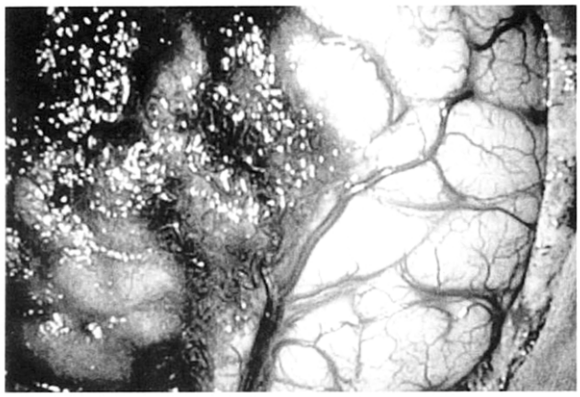


Fig. 2: Subarachnoidal haemangiomatosis was observed over the left front-parietal lobe.

tal. No abnormalities were noted in the patient's physical appearance. She was neurologically intact. EEG revealed generalized slow waves of the left frontal lobe, and a computer tomography (CT) scan showed atrophy in the left frontal lobe and a high-density signal in the white matter. Gadolinium-enhanced magnetic resonance imaging (MRI) revealed enhancement of the gyral surface of the left fronto-parieto-temporal lobe (Fig. 1). Three-dimensional reconstruction of the MRI images using a Viewing Wand System also demonstrated that the atrophy in the left hemisphere had progressively worsened and seizures became intractable despite increased anticonvulsants. A diagnosis of SWS was made, and surgical treatment was scheduled.

Surgical treatment consisted of lesionectomy by lobar corticectomy as well as excision of the cortical epileptogenic focus. A left fronto-parieto-temporal craniotomy was performed under neuroleptanalgesia using Fentanyl. Remarkable leptomeningeal haemangiomatosis was observed in the left fronto-parieto-temporal lobe with CSF collection due to cortical atrophy in the fronto-parieto-temporal region. The haemangiomatosis covered the whole frontal lobe and parietal lobe, and part of the sylvian fissure surface of the temporal lobe (Fig. 2). The use of nitrous oxide gas was discontinued 15 minutes before ECoG recording in order to confirm rapid θ or α waves using scalp electrodes placed on the occipital region. ECoG was recorded to find the epileptogenic focus. A grid electrode with 20 contacts was arranged on the surface of the haemangiomatosis. The reference electrode was placed on the left ear. A positive epileptogenic focus was located at the left posterior temporal lobe just posterior to the margin of the haemangiomatosis over the sylvian fissure (Fig. 3(a)). The ECoG over the haemangiomatosis of the frontal lobe demonstrated low-voltage delta activity. Left fronto-parieto-temporal corticectomy including the epileptic focus was performed. The left lateral ventricle was kept intact. The neuronavigation system of the Viewing Wand System is very useful to keep the lateral ventricle intact. After the resection, ECoG of the remaining gyri around the excision demonstrated no spike activity (Fig. 3(b)).

Histologically, the haemangiomatosis was observed in the subpial region. The cortical layer under the haemangiomatosis was atrophic with mild gliosis and slight calcification (Fig. 4(a)). The epileptogenic cortex, which showed periodic spike activities, demonstrated mild gliotic changes (Fig. 4(b)).

After the operation, the patient developed right hemiparesis and hemihypesthesia but became seizure-free with no anticonvulsant therapy. She began to stand up and walk within 2 months of the surgery, and she gradually became able to hold up her right arm. She was able to run and sing songs 12 months after surgery. At the present time, 25 months after surgery, she has almost normal development in both motor function and intellectual development except for a little clumsiness in the right hand. Follow-up MRI demonstrated mild ventricular dilatation on the side of the surgery but subdural fluid collection was not observed.

DISCUSSION

The main features of SWS are seizures that are resistant to anticonvulsant therapy, neurological deficits, and mental retardation. It is reported that hemiplegia becomes severe, and seizures become intractable if the patient has the first seizure attack in early infancy. It is also reported that the venous haemangiomatosis of SWS reduces blood flow in the involved hemisphere, leading to cerebral atrophy and gradual impairment of the function of the contralateral normal hemisphere[1–5]. Prompt surgical treatment can preserve the function

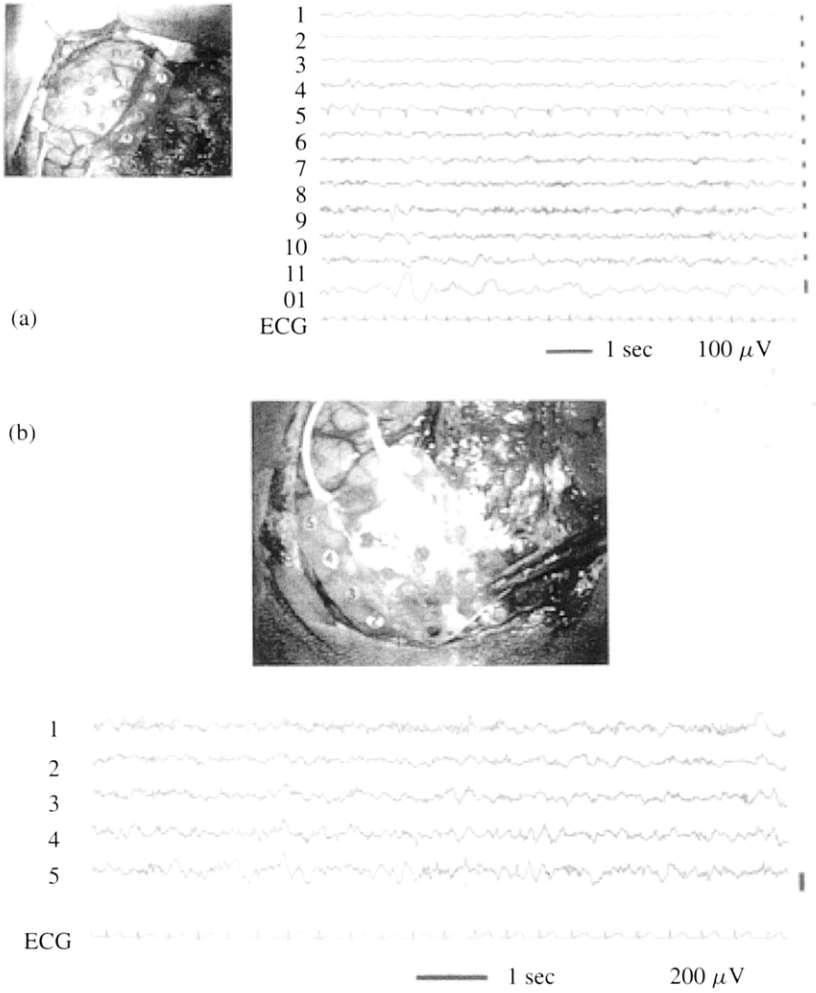


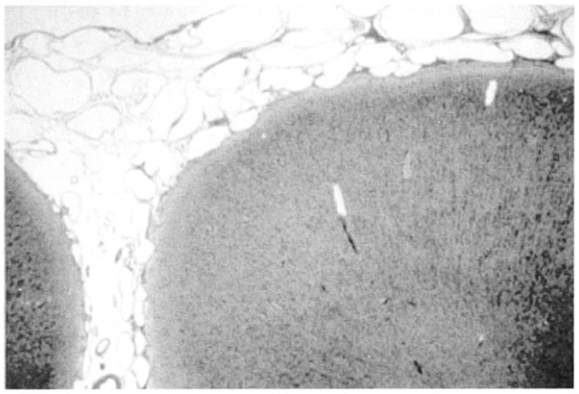
Fig. 3: (a) Intraoperative photograph showing the ECoG electrodes in position over the haemangiomatosis. Numbers indicate the position of the haemangiomatosis and electrodes, respectively. The ECoG is detecting epileptiform discharges in the posterio-temporal lobe. Note the prominent periodic spike discharge in channels 5 to 9. (b) Intraoperative ECoG after removal of the haemangioma and cortex showing no epileptogenic activity.

of the remaining normal hemisphere. Surgical treatment, including both hemispherectomy and lobectomy of haemangiomatosis at an early age has been reported to be effective for controlling convulsions which are resistant to medication[6].

According to the grading of Ogunmekan[5], the surgical results of both operations are sometimes poor. It is thought that not all of the epileptic foci are removed in the case of lobectomy, and that the patient's age is too great in the case of hemispherectomy. A hemispherectomy is better than a lobectomy for the outcome of seizures, although a hemispherectomy may result in many complications, such as cerebral hemosiderosis or hydrocephalus. Based on these observations, we performed lesionectomy, lobar corticectomy and focus resection without opening the lateral ventricle. The post-operative course was uneventful, and the patient did not develop hydrocephalus or subdural effusion.

Intraoperative ECoG has been used to detect epileptogenic foci in other neurological diseases such as epilepsies, brain tumours and AVMs[7–13]. Several reports have indicated the necessity of resecting cortical epileptogenic foci and the importance of intraoperative ECoG to detect epileptogenic foci. Awad[7] reported that maximal resection of epileptogenic foci identified by ECoG and post-operative EEGs offers the best chance of controlling intractable epilepsies. It has also been reported that intraoperative ECoG is necessary for identifying functionally important areas such as the motor area and the motorsensory area[11]. These results indicate that the detection of epileptogenic foci may be regarded as representing a high probability of developing post-operative convulsive seizures, and resection of the involved gyrus must be confined to an extent that does not cause complications or severe neurological deficits. Thus, lesionectomy with focus resection is the best choice of surgical treatment for the present case. Also, considering seizure control and neurolog-

(a)



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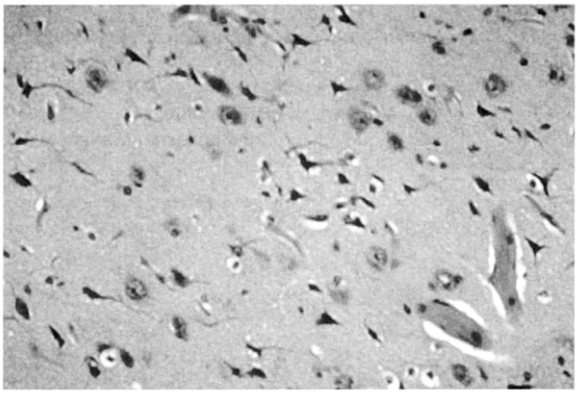

Fig. 4: Photomicrograph of the surgical specimen of (a) the venous haemangiomatosis of the SWS with gliotic cortex (H&E, ×40) and (b) the cortex of epileptogenic focus with mild gliotic changes (H&E, ×400).

ical deficits after the operation, it is better to perform surgical treatment at an early age. However, we must follow the state of patients carefully, because we cannot anticipate whether neurological deficits will improve, whether seizures will decrease, or whether mental retardation will occur. To the best of our knowledge, intraoperative ECoG of SWS has not been well studied. This is the first report of focal spiking detection by intraoperative ECoG in the surgical treatment of SWS. Although there have been several reports on surgical treatment of SWS[1–5], the post-operative neurological status has not been well described.

In the present case, epilogenic foci were detected in the apparently normal cortex, posterior to the haemangiomatosis, by ECoG as multiple spikes and periodic spike activities. After corticectomy of the haemangiomatosis and focus resection, the spike activity disappeared. After surgery, the patient developed transient right hemiparesis but gradually recovered within 7 months. The patient demonstrated no epileptic activity in the post-operative conventional EEG 1 year after the surgery and became seizure-free with no anticonvulsant therapy. The results of the present study indicate the importance of early surgery for a medically intractable epileptic child with SWS and also show the usefulness of intraoperative ECoG to confirm a focal epileptogenic focus. Moreover, lobar corticectomy without opening the lateral ventricle is an excellent alternative to the surgical treatment of SWS.

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