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Surgery for intractable epilepsy in a 14-year-old girl

一名14歲女童頑固性癲癇的外科手術

We report on a 14-year-old girl who presented with a 2-year history of simple and complex partial seizures with secondary generalisation. Monotherapy using carbamazepine and combination therapy using carbamazepine and gabapentin had been tried within the 2 years before presentation. Seizure control, however, was poor. Magnetic resonance imaging showed structural abnormalities over the right occipital and temporal lobes. Continuous scalp video electroencephalography was performed over 4 days, during which six clinical seizures were associated with electroencephalography changes at the right occipital and temporal lobes. Invasive intracranial video electroencephalography identified a focus at the right occipital lobe, a focus at the right temporal lobe that spread rapidly to the right parietal lobe, and an irritative zone over the posterior part of the right frontal lobe. Functional mapping delineated the motor and sensory cortices. Right temporal lobectomy, right occipitoparietal cortical excision, and multiple subpial transections of the posterior part of the right frontal lobe were performed. For 16 months after the surgery, the patient has been seizure-free while receiving drug treatment, and the only complication reported has been a segmental loss of the left visual field.

我們報告一名出現兩年二期簡單和複雜部分癲癇史的14歲女童。在本報告之前兩年內，使用 carbamazepine 單一療法和 carbamazepine 與 gabapentin 複合療法。然而癲癇控制很差。磁力共振圖顯現右枕部和顳葉上游結構異敘。頭皮錄像腦電描記法連續進行了 4 天，在這期間 6 次臨床癲癇與右枕部和顳葉上的腦電圖變化相關連。侵入性顱內錄像腦電描記法識別出在右枕葉上的一病灶，右顳葉上已快速擴散到右壁葉的一個病灶，和右前葉後部的一個刺激區。手術切除了右顳葉/右枕壁皮質和右前葉後部的多重軟膜。在手術後16個月裡，患者在服藥治療時無癲癇發作。所報告的唯一併發症是左視區有部分失明。

Key words:
Brain diseases/surgery;
Child;
Electroencephalography;
Epilepsy, temporal lobe/surgery;
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關鍵詞：
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Introduction

Epilepsy is a common neurological problem in children. Although in most cases, drug treatment is adequate for seizure control, 5% to 10% of cases remain medically intractable. Surgical therapy may offer a cure for intractable localisation-related epilepsy.[1]

The lack of a uniform definition of medically intractable epilepsy would, in general, imply that all therapeutic attempts with single or combined drug therapy for a sufficient period of time would fail to control the seizures. It is not known, however, how long medical treatment should last and how many drugs should be tried.[1-4] In some medically intractable

types of epilepsy, the likelihood of an improvement in seizure control following surgery is high. Examples include epilepsy caused by a structural lesion or mesial temporal sclerosis.[5] Other factors in deciding whether to perform surgery in children are the adverse psychosocial consequences brought about by the disease, the quality of life, and the side effects of the drugs used. Furthermore, frequent seizures in infants and young children can have a disastrous effect on their development. We report on a patient with intractable epilepsy, who subsequently received epilepsy surgery.

Case report

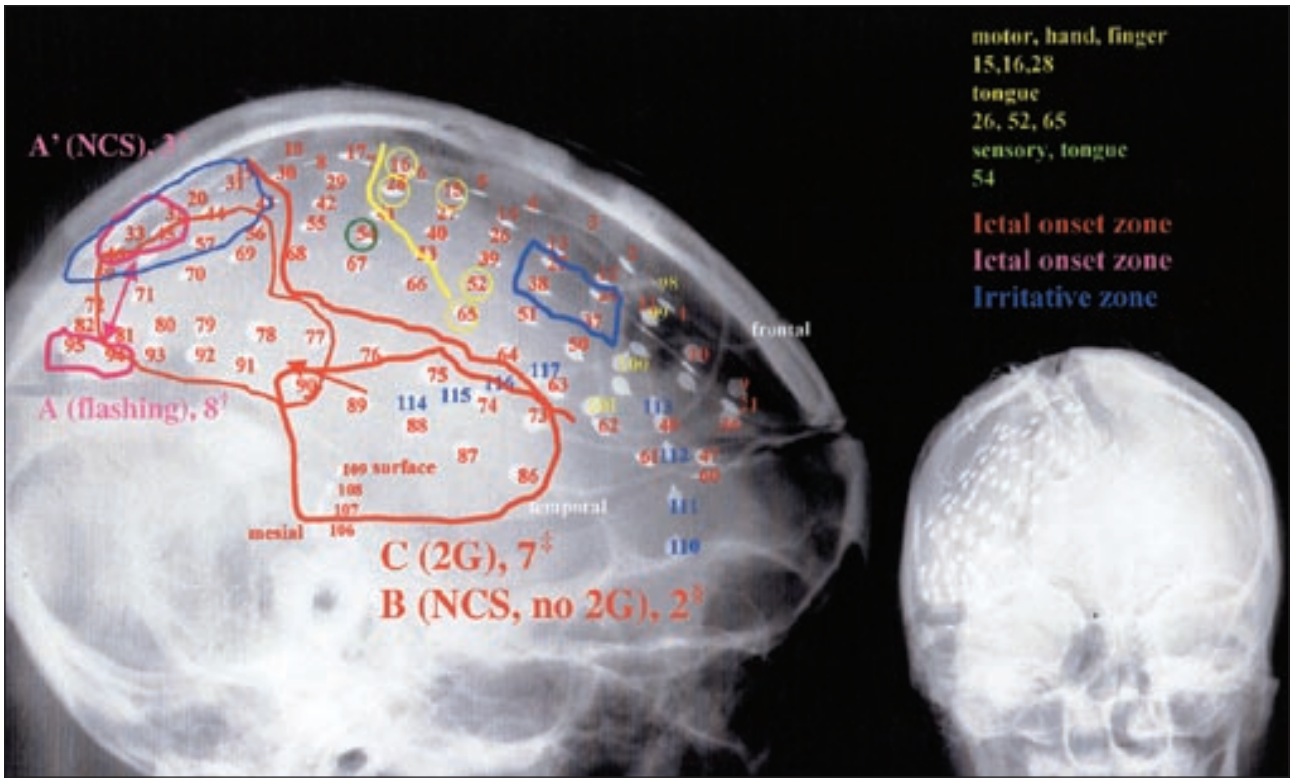
A 14-year-old right-handed girl presented to the Hospital for Sick Children, Toronto, Canada on 5 November 1997. Her birth and development had been normal, but she had started to have seizures in May 1997. Her seizures were of multiple types: the first type was characterised by seeing flashes of light, the second was the sensation of a strange taste, and the third was characterised by staring. These features would precede left-side or whole-body clonic activity. From the descriptions given, the seizures were partial in onset and with or without secondary generalisation. The experience of seeing flashes of light suggested a seizure arising from the occipital lobe, whereas the strange taste sensation was typical for seizures arising from the mesial temporal lobe (uncinate fit). Scalp electroencephalography (EEG) had previously shown an epileptic focus in the right posterior hemisphere involving the posterior temporal, parietal, and occipital regions. Initial drug treatment using carbamazepine 600 mg twice daily had been tried but without success. Gabapentin 600 mg three times per day had been added later. The patient experienced approximately a dozen seizures a day.

Magnetic resonance imaging (MRI) showed structural abnormalities over the right occipital and temporal lobes that were indicative of right mesial temporal lobe sclerosis. There was also right occipital atrophy. Continuous scalp video EEG was performed over 4 days, during which six clinical seizures were associated with EEG changes at the right occipital and temporal lobes. No abnormal discharge was noticed over the left hemisphere. A functional study of cerebral metabolism using interictal positron emission tomography (PET) defined a large region of hypometabolism that covered the right occipital parietal, temporal, and inferior frontal regions. This finding suggested the presence of an interictal epileptic focus in this region. A functional study of cerebral blood flow using interictal single-photon emission computed tomography (SPECT) demonstrated hypoperfusion at the right temporal lobe, thus suggesting an interictal epileptic focus in this region.

Neuropsychological assessment recorded normal language and memory function. There was weakness, however, in the patient's organisation of perceptual information. Functional MRI showed a left hemispheric dominance for language. All the above pieces of information suggested a large epileptogenic zone in the right hemisphere, which was secondary to right mesial temporal sclerosis and right occipital atrophy. Surgery was considered for improving seizure control and cognitive impairment. If further repeated seizures were allowed, brain damage would have been likely to follow.

On 23 August 1999, a 95–surface-electrode array subdural grid, and several depth electrodes were implanted into the right hemisphere and were monitored for 4 days. During the monitoring period, 20 clinical or subclinical electrographic seizures were captured. Ictal zones were found at the right occipital lobe and at the right temporal lobe with a rapid spread to the right parietal lobe. In addition, an irritative zone over the posterior part of the right frontal lobe was identified. The epileptic foci are indicated in the Figure, which is an X-ray of the skull with the implanted electrodes. The motor cortex for the left hand and fingers was defined and found to be close to the irritative zone at the posterior part of the right frontal lobe. The child was asked to label objects, count numbers, and name letters and months while electrical cortical stimulation was applied to the electrodes over the right temporal and inferior frontal regions. No speech arrests or errors were noted, thereby indicating that the dominant language centre was not in the right hemisphere. The patient was thus expected to have no significant language problems if the right temporal lobe or right inferior frontal region were resected.

On 27 August 1999, the subdural grid was removed and right temporal lobectomy and right occipitoparietal cortical excision were performed. For the focus at the posterior part of the right frontal lobe, multiple subpial transections were performed. Early postoperative complications included subgaleal fluid collection, which was managed by the application of a pressure bandage over the scalp and lumbar drainage of cerebrospinal fluid. Another complication was transient diplopia, which was probably due to the manipulation of the third and fourth cranial nerves during the removal of the mesial temporal region. No seizures were



* 3 seizures originated from right occipital lobe with no clinical sign
† 8 seizures originated from right occipital lobe with the clinical sign of seeing flashes of light
‡ 7 seizures originated from right temporal lobe with secondary generalisation
§ 2 seizures originated from right temporal lobe with no sign nor secondary generalisation

Fig. Skull X-ray of an epileptic patient showing electrodes of the subdural grid and epileptic foci
Marked areas represent epileptic foci over the right occipital lobe (pink); right temporal lobe (red) with rapid spread to the right parietal lobe; and an irritative zone on the posterior part of the right frontal lobe (blue). The yellow regions correspond to the mapped area for movement of the right hand and fingers, and tongue

recorded after the operation. The patient was discharged home 2 weeks postoperatively. For 16 months after the surgery, the patient has been seizure-free while receiving drug treatment, and the only complication reported has been a segmental loss of the left visual field, but this loss in visual field has had minimal impact on her daily life.

Pathological examination of the posterior occipital lobe revealed cortical microdysgenesis, whereas pathological examination of the mesial temporal lobe revealed mesial temporal sclerosis.

Discussion

In this case of intractable epilepsy, two epileptic foci were found: one over the right temporal lobe and one over the right occipital lobe. Hence, this would not be an ideal case for epilepsy surgery. However, structural lesions—namely right mesial temporal sclerosis and right occcipital atrophy—were found on the MRI scans. Because the presence of structural lesions is a good prognostic factor for the success of surgery, these MRI findings suggested a reasonably high likelihood of cure from surgery such as multilobar resection. Thus, the patient in this case was evaluated for epilepsy surgery.

The success of epilepsy surgery lies in the identification of a well-defined epileptogenic zone and the complete removal of that epileptogenic zone.[6] The location of this zone is suggested by the symptomatology of seizures, although this strategy is not totally reliable. For example, this patient's experience of seeing flashes of light is quite typical of seizures arising from the occipital lobe. The initial investigations to locate the epileptogenic zone included EEG and MRI. Depending on the availability of resources, other investigations such as PET and SPECT, which were also used in this case, are helpful to identify the epileptogenic zone. The aim is to achieve concordance from all the diagnostic studies. In this case, there was some discrepancy about the extent of the epileptogenic zone on the right hemisphere; thus, invasive monitoring using subdural grid and depth electrodes was arranged. Simultaneous recording of ictal events using this method may be considered the gold standard of recording epileptic activity, because the electrical field is recorded directly from the cortical surface. There is

no masking effect from the skull and scalp, which would be present in scalp electrode recording.

In this case, a subdural grid that covered the convexity of the right frontal, parietal, temporal, and occipital lobe was inserted. In addition, four depth electrodes towards the right mesial temporal lobe were inserted. Depth electrodes are necessary to document the origin of the seizures in the deep-seated areas such as the mesial temporal lobe. During the monitoring, 20 seizures were captured; this figure is sufficient for defining the epileptogenic zone. Furthermore, an irritative zone over the posterior part of the right frontal lobe was noticed. This region revealed significant potential epileptogenicity. It is usually characterised by a pattern of spikes or sharps consistently over a focus. The irritative zone, as well as the epileptogenic zone, must be included in the surgical procedure if surgery is to be successful.[7]

Functional mapping in this case prevented the resection of important cortex. In this patient, mapping for motor function, speech, and speech-related memory were performed. The visual cortex was not mapped, however, because the epileptogenic zone was extensive and involved the right occipitoparietal region and right temporal lobe—regions that must both be resected to achieve a good outcome. Thus, the visual cortex on the left side was certain to be sacrificed, thereby making mapping of the visual area unnecessary. Segmental loss of the left visual field was the only postoperative complication reported.

In this case, right temporal lobectomy and right occipitoparietal cortical excision were performed. Lobetomy is the removal of the whole lobe and cortical excision is the removal of grey matter. Multiple subpial transection—a relatively new technique—was carried out for the focus at the posterior part of the right frontal lobe, because of its closeness to the motor area for the hand and fingers. The technique interrupts the intracortical, horizontally oriented fibres, while preserving the main vertically or radially aligned elements, because epileptiform activity is generated through side-to-side connecting fibres, whereas normal cortical physiological functions are mediated through vertically arranged fibres. Hence, this technique is adopted when an epileptic focus lies in an important cortex, such as the motor cortex or eloquent cortex. The outcome of multiple subpial transection from a large series is still pending. In a report by Sawhney et al,[8] cortical resection and multiple subpial transection were performed in 12 patients, with 11 of these showing a worthwhile decrease in seizure frequency.

In this case, the epilepsy had dual pathology—namely mesial temporal sclerosis and microdysgensis of the occipital lobe. This is not an uncommon situation. Cendes et al[9] showed that coexistent hippocampal atrophy (indicating dual pathology) was present in 25 (15%) of 167 patients with temporal or extratemporal partial epilepsy. Raymond et al[10] demonstrated an association of hippocampal sclerosis with cortical dysgenesis in 15% of 100 patients with hippocampal sclerosis.

Conclusion

This case illustrates when and why surgery is indicated for cases of intractable epilepsy. Furthermore, presurgical investigation is very important for identifying the epileptogenic zone and in defining the resection margin. Epilepsy surgery requires a team of devoted specialists. The result can be an improvement in seizure control, attainment of normal development and schooling, and a better quality of life.

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