Clinical commentary

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Bilateral occipital dysplasia, seizure identification, and ablation: a novel surgical technique

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ABSTRACT – MRI-guided thermal ablation is a relatively new technique utilising heat to ablate both tumours and epileptogenic lesions. Its use against epilepsy offers some patients a new and relatively safe way of reducing or aborting seizures. Most cases of MRI-guided thermal ablation have been performed in patients with isolated lesions. Placement of depth electrodes prior to laser ablation has been rarely performed. We present a case with bilateral independent lesions traversing eloquent cortex, which, after sampling for seizures and successful ablation, retained normal function. The patient is, to date, seizure-free.

Key words: focal epilepsy, thermal ablation, occipital dysplasia, periventicular dysplasia, depth electrode, pharmacoresistant

Epilepsy surgery is often considered after failure of two to three antiepileptic drugs (AED) (Berg and Kelly, 2006). Factors negating surgery include the potential for impairing primary function (language, motor, sensory, vision, cognitive outcome, and memory) and multiple or bilateral deeply-located sites of seizure onset in the same individual (Helmstaedter and Kockelmann, 2006). Palliative procedures, special diets, and the addition of further medications are therefore the only options available to some individuals with epilepsy (Clarke *et al.*, 2007). Extra-operative techniques such as VEEG, MEG, PET, fMRI, and SPECT, as well as direct cortical stimulation, have helped to guide surgical procedures in an attempt to minimise surgical morbidity. This still does not address the necessity of traversing normal cortex to remove epileptogenic foci successfully. The opportunity for using alternative means of aborting or significantly reducing seizures is therefore a welcomed option. Less invasive techniques, such as gamma knife have been attempted with varying success in cases with isolated lesions (Bartolomei *et al.*, 2008). Gamma knife is not without radiation and associated risks, there is often a significant delay to seizure freedom, and it has not been shown to be more effective than traditional surgery (Régis *et al.*, 2004). Stereotactic EEG-guided

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radiofrequency ablation (RFAB) has also been applied to epileptiform lesions with varying success, though with some limitations of specificity due to a lack of real-time image-guided monitoring (Catenoix *et al.*, 2008). A relatively new technique uses laser-induced heat, monitored via MRI in real time, to ablate both tumours and epileptogenic lesions. Its use against epilepsy offers some patients a new and relatively safe way of reducing or aborting seizures. Most cases thus far have involved isolated lesions, and sampling for seizures with depth electrodes prior to ablation is rare. We present a case with bilateral independent lesions traversing eloquent cortex, which, after sampling for seizures and successful ablation, retained normal function. The patient is, to date, seizure-free.

Case study

A 22-year-old, left-handed university student presented with focal-onset epilepsy caused by bilateral linear cortical malformations affecting both occipital and right temporal lobes, abutting and adjoining peri-ventricular nodules. She had normal birth and developmental history. Her neurological examination was normal, including no visual deficits in visual confrontation. She began having seizures at 11 years of age. Eleven antiepileptic medications failed to render her seizure-free. For fear of affecting her vision, surgical resection was not previously offered. A vagus nerve stimulator (VNS) was implanted, which aborted clusters, but 2-4 daily seizures persisted significantly, affecting her ability to drive, causing her to miss classes, and impaired her overall daily activity. During her seizures, an aura of subtle changes in vision was described as "fuzziness". This evolved into seeing numbers (often the number 9), occasionally certain words, and ultimately visual loss. Visual changes often initially involved her left hemi-field or both fields of vision were simultaneously involved. Early in the course of her illness, she described occasional associated auditory hallucinations or hearing specific phrases, but this was no longer experienced. Alteration of consciousness was rarely documented by the patient, but her parents described an occasional confessional state with paucity of speech. During what was described as a typical seizure, the patient's manifestations evolved with staring, oro-buccal automatisms, and inability to follow commands.

MRI demonstrated subependymal heterotopic grey matter involving the atria, occipital, and temporal horns. The right hemisphere exhibited occipital and temporal linear dysplasia approximating, but not involving, the right mesial temporal structures, and left per-ventricular heterotopic grey matter, extending linearly into the occipital lobe (*figure 1A*). Scalp video electroencephalogram (VEEG) recordings revealed interictal posterior discharges (greater on the right side than the left). The right temporal/occipital region was maximally involved in most ictal events. Subtraction SPECT demonstrated hyper-perfusion involving the right posterior temporal, occipital, and inferior parietal regions (*figure 1B*). Functional MRI revealed bilateral BOLD signal hugging the regions of cortical dysplasia, after testing monocular and bilateral visual evoked potentials (*figure 1C*). DTI/tractography revealed normal anterior-posterior, lateral and caudal-rostral tracts, except for those which separated and surrounded the dysplasic tissue in both occipital lobes (*figure 1D*). Neuropsychological testing suggested frontal-temporal dysfunction bilaterally with greater involvement of her non-dominant hemisphere. WADA testing suggested 92 and 100% recall of verbal and visual stimuli to left-sided injection, and 42 and 50% verbal and visual stimuli to right-sided injection, respectively. Language dominance was, therefore, deemed to be left-sided. Therefore, focal resection of the right temporal and occipital regions would unlikely affect language; however, visual loss would be inevitable as well as a possible reduction in memory. It was decided to place bilateral depth electrodes lateral to, and partially traversing, bilateral regions of dysplasia (*figure 2A*), although the neurophysiology and SPECT suggested right-sided maximal activity. This decision was made in an attempt to negate a lack of rapid spread from the left sided dysplasia to the right.

Due to the patient's VNS, she required both MRI and CT for stereotactic planning. The patient underwent MRI and was then brought to the CT scanner where she was placed under general anaesthesia and had placement of the Leksell stereotactic frame. A CT scan was obtained and the patient was brought to the operating suite. MRI and CT scans were merged and stereotactic planning for bilateral occipital 8×5-mm PMT depth electrodes was performed. Depth electrodes were placed adjacent to the areas of cortical dysplasia using standard stereotactic techniques with the Leksell stereotactic frame. Following post-implant MRI, the patient was transported to the Epilepsy Monitoring Unit (EMU).

The lateral placement was performed in order to allow more mesial and precise placement of the ablation catheter. This meant abutting normal white matter tracts. Cortical stimulation, the gold standard for defining eloquent cortex, might potentially have produced false specificity in this case because of the depth placement trajectory, although it may still have been confirmatory if no visual changes were experienced. Surgical planning to avoid functional areas, however, relied heavily on pre-surgical planning. During 3.5 days

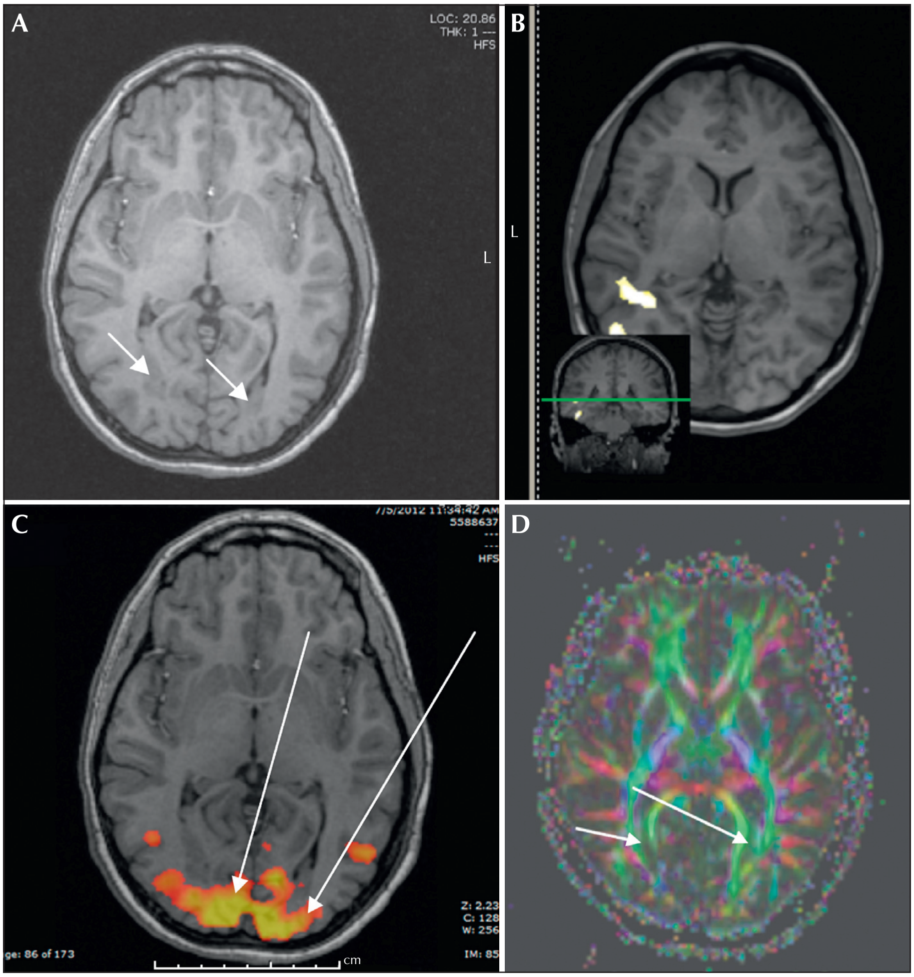

Figure 1. (A) Bilateral cortical dysplasia extending into both occipital lobes (greater in the right side than left) from both peri-ventricular regions (arrows). (B) Subtraction SPECT demonstrated hyper-perfusion involving the right posterior temporal, occipital, and inferior parietal regions. (C) Functional MRI with bilateral bold signals hugging the regions of cortical dysplasia (arrows). (D) DTI/tractography revealing tracts which separate and surround the dysplasic tissue in both occipital lobes (arrows).

of VEEG, the patient had six subclinical events (four with right-sided onset and two with left sided onset) and five electroclinical seizures (two right sided onset, two left sided onset with rapid spread to the right, and one with diffuse onset evolving to right-sided maximal activity). Though ictal onset occurred from both areas independently, right-sided activity was involved during the evolution of all seizures (*figures 2B* and *2C*), therefore reflecting the right-sided maximal activity observed on the scalp EEG and the positive right-sided SPECT. This similar ictal evolution also accounted for the stereotyped clinical features (a description of visual change in brief events to altered consciousness with oro-buccal automatisms and occasional versive head and eye deviation to the left in longer events, greater than 60 seconds).

Depth electrodes were removed on the morning of the ablation procedure and the patient underwent a new MRI scan, and again was brought to the CT scanner, placed under general anaesthesia, and underwent application of the Leksell stereotactic frame. A CT scan was obtained and the patient was brought to the operating suite. The MRI and CT scans were merged and stereotactic planning was performed for placement of bilateral occipital laser fibres that were placed using standard techniques with the Leksell stereotactic frame. Due to the proven bilateral involvement, we elected to place the laser fibres for the ablation *via* a new trajectory in order to better encompass the shape of the lesions. The applicator assembly comprised an inner 1.6-mm diameter laser optical fibre with a diffuser tip (10 mm) and an outer polycarbonate saline cooling catheter (Visualase® Inc., Houston, TX). The catheter contained a stainless steel stiffening stylet and was inserted through an anchor bolt. The right-side fibre was placed in an elongated area of heterotopia, lateral to the trigone and posterior temporal horn of the ventricles. The left-side fibre was

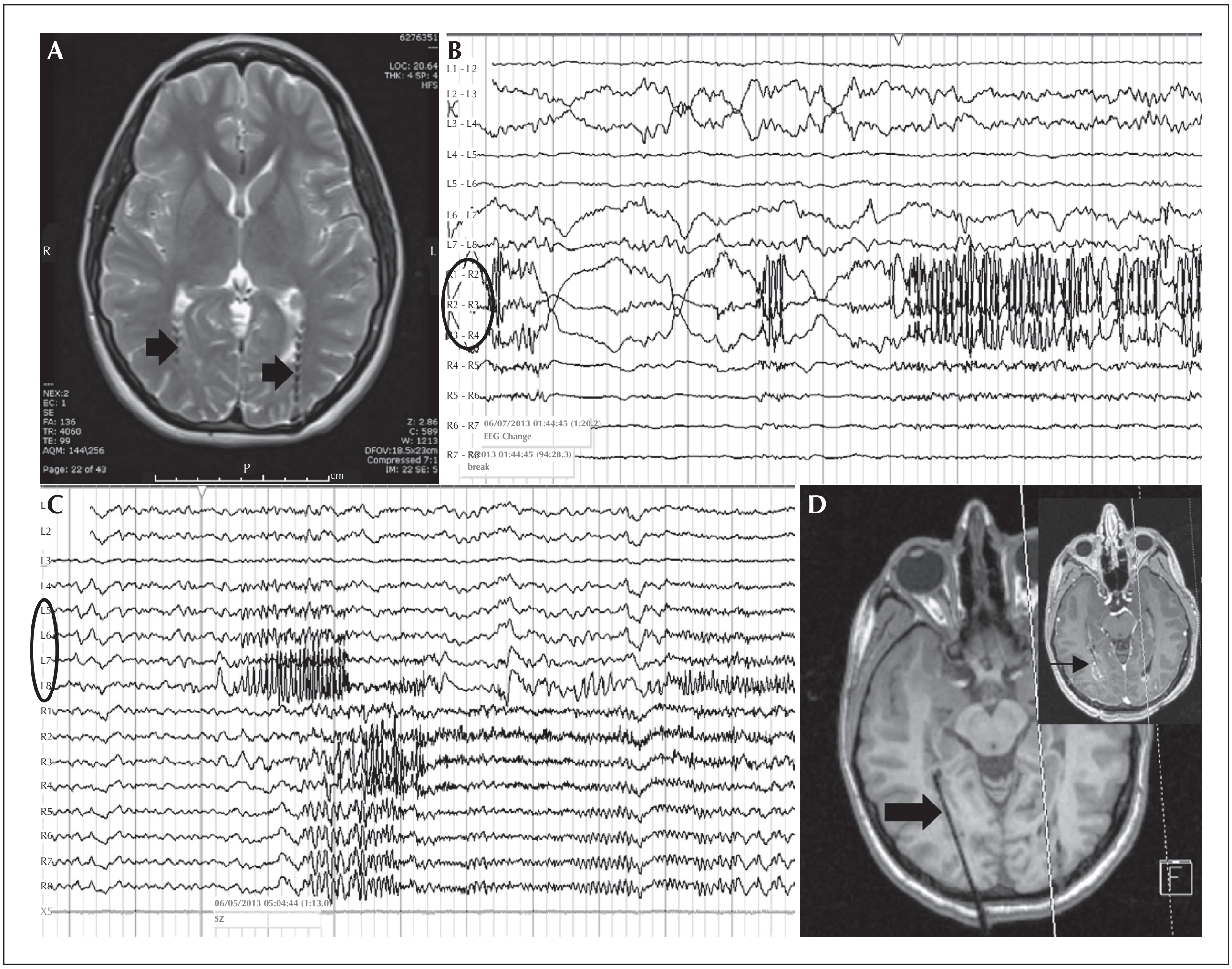

Figure 2. (A) Bilateral depth electrodes lateral to, and partially traversing regions of, dysplasia. (B). Seizure onset and evolution involving the right (R) depth electrode (circle). (C) Seizure onset involving the left (L) depth electrode (circle), then rapidly spreading to the right. (D) Ablation catheter (thick arrow) and area ablated (thin arrow).

placed in an area of heterotopia capping the trigone and extending through an area of heterotopia in the roof of the trigone of the ventricles (*figure 2D*). Leksell frame was removed and the patient was transported to the MRI suite. During the laser ablation technique used, heat sensors during the MRI and the compatibility of the saline-cooled laser applicator allow for real-time MR-guided heat monitoring and ablation (Curry *et al.*, 2012; Jethwa *et al.*, 2012). Temperature safety limits set by the user automatically terminate laser delivery if the temperature exceeds 45 degrees, therefore avoiding and protecting the integrity of surrounding tissue and conservatively restricting thermal damage to a 2-mm radius around the lesion. The laser fibre and cooling lines were then routed from the patient in the scanner to a Visualase workstation (Visualase® Inc., Houston, TX) in the control room for real-time MRI and modelled estimates of the necrosis zone. Bilateral sequential procedures allowed for thermal ablation of both linear heterotopias with sparing of the mesial temporal structures (*figure 2D*). The patient was discharged the day after the procedure with what she described as "occasional visual blurring" for approximately one week, however, she required no assistance with activities of daily living and has experienced no further symptoms. Ophthalmological perimetry testing performed at three months revealed no deficits and she has remained seizure-free to date (eight months post-ablation).

Discussion

A surgical option would seldom be offered in cases of bilateral dysplasia approximating eloquent cortex.

Others have advocated subdural electrode evaluation and focal resection after focal EEG findings and supportive functional imaging. However, based on fMRI and tractography data in this patient, it would be nearly impossible to avoid visual impairment. Memory dominance and language were left-sided. However, the best possible outcome for this college student would be complete avoidance of any deficit of memory, which makes avoiding removal of mesial temporal structures critical (Scoville and Milner, 1957). Kuzniecky *et al.* (1997) described 10 cases of cortical malformations in the occipital lobes, presenting predominantly with ictal phenomenon characterised as visual changes. Pre and post-operative deficits, or lack thereof, suggested visual re-organisation. Whereas outcome was uniformly favourable in the series of Kuzniecky *et al.*, a larger cohort revealed patients with cortical dysplasia to have a worse seizure outcome than those with tumours (Kuzniecky *et al.*, 1997; Aykut-Bingol *et al.*, 1998). Stereotactic EEG-guided radiofrequency ablation (RFAB) has been applied successfully to epileptiform lesions. Seizure freedom described in the literature has been limited to the destruction of smaller lesions and its use often previously described for palliation (Parrent and Blume, 1999; Guénot *et al.*, 2004; Catenoix *et al.*, 2008). A study utilising it specifically for amygdalohippocampectomy, however, demonstrated 78% seizure freedom over two years (Liscak *et al.*, 2010). The laser ablation technique is MRI-compatible whereas RFAB is not due to radiofrequency interference. The lack of real-time MRI-guided imaging and a protective cooling device limits RFAB specificity of the ablation zone and reduces the ability to preserve surrounding tissue. The conductive spread of RFAB heat also potentially results in a wider transition zone when compared with the heat generated from the laser.

In this case, there was shifting or displacement of visual fields away from areas of dysplasia, as evident on her fMRI and tractography, and preoperative perimetry testing revealed no changes. Cortical stimulation remains the gold standard, and though it might have been falsely localising in this case, it may have provided added security if no visual changes were experienced. The controlled nature of the real-time ablation, safeguards against thermal injury of surrounding tissues, detailed pre-surgical workup (fMRI, DTI), and subclinical events with no deficits described until spread and prolongation of the seizure, together made all significant visual deficits less likely with no objectively-defined deficits post-surgery. There was, however, an extensive discussion prior to consent with regards to placement of the depth electrodes, catheters, and ablation, as well as the risks of visual deficits or visual loss. The patient requested to precede secondary to the severity of seizure burden. A specific bolt (PMT inc.) created after, and as a direct result of this case, now allows for placement of the depth electrode and laser applicator along the same trajectory. This allows for better specificity and sensitivity if and when cortical stimulation is required.

Epilepsy surgery is still required by most epilepsy patients who are candidates (Wyllie *et al.*, 2007). Traditional epilepsy surgery allows for larger sampling for the epileptogenic zone, removal or disconnection of more tissue if required, and more detailed cortical stimulation. This is, however, a continually evolving process and functional mapping, stereotactically-placed depth electrodes, and other techniques may allow for a shift in procedural planning and mitigate the need for extensive grid placement, large exposures, and resections. Individuals deemed suitable for this procedure may benefit from low morbidity and shorter hospital visits (Curry *et al.*, 2012). Thermal ablation could also be life-altering for some individuals in whom epilepsy surgery may not be an option or risk of harm negates involvement of functional cortex. □

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