Case Report

Surgical outcome in neocortical resections of type IIId focal cortical dysplasia with accompanying medial temporal pathology

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ABSTRACT

Focal cortical dysplasia (FCD) type IIId is a newly proposed type associated with early-life brain insults. Such patients are often considered unsuitable for resective epilepsy surgery, given the usually wide extent of the lesion and the poor correlation of MRI to the epileptogenic pathology. Two patients with intractable epilepsy, early-life ischemic/traumatic injury and MRI findings of extensive unilateral cystic-gliotic and ipsilateral medial temporal sclerotic-malformative lesions were subjected to presurgical evaluation revealing well-localized neocortical ictal onsets. They underwent tailored neocortical resections sparing medial temporal areas and achieved Engel class I postsurgical outcomes. Histopathology was consistent with type IIId focal cortical dysplasia. Successful outcomes with tailored resections may be achieved in cases with this subtype of focal cortical dysplasia, in the presence of converging and well-localized semiological, EEG and functional imaging data, even on a background of complex and extensive MRI abnormalities. Medial temporal pathology, although often present in this setting, is not necessarily the site of ictal onsets, and its resection may not be always mandatory for a favorable outcome.

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1. Introduction

The new proposal for classification of focal cortical dysplasia (FCD) recognizes a particular type, IIId FCD, associated with an epileptogenic lesion acquired in early life (i.e., traumatic injury, ischemic injury or encephalitis) [1]. This is classified among “type III” FCD group associated with certain other epileptogenic lesions (in particular, hippocampal sclerosis, tumors and vascular malformations). Histopathologically, type IIId FCD consists of altered cortical architecture and cytoarchitectural composition, occurring adjacent to the principal early-life lesion. There is not yet much information regarding the prevalence of these cortical dysplastic changes among patients with these kinds of early-life lesions, and imaging-pathologic correlations are only at the beginning; nevertheless, it could be inferred that such an association is probably high.

Epileptic seizures secondary to early-life brain injuries often follow a protracted and drug-resistant course. Given the destructive nature and involvement of multiple brain regions by initial injury, many of these patients are not considered suitable for epilepsy surgery. Others with well-lateralized hemispheric lesions undergo extensive procedures (hemispherectomy/hemispherotomy) which, although associated with successful postsurgical seizure control, may induce new or aggravate previously existing neurological deficits [2,3]. Results of focal resective surgery are overall poor. This work aims to denote that a) selected cases with converging semiological, electrophysiological and structural/functional imaging evidence of localized ictal onsets may show favorable postsurgical results to focal-tailored resections and b) accompanying mesio-temporal pathology ipsilateral to the gross lesion may not contribute to seizure generation.

2. Materials and methods

2.1. Case 1

Patient 1 was a 10-yr-old boy originally diagnosed with left hemispheric atrophy and an ipsilateral parietotemporopariietal porencephalic cyst. He developed right-sided upper extremity hemiparesis, mild left superior quadrantanopia, and drug-resistant seizures (for history, presurgical evaluations and treatment details, see the Supplementary data). Recent MRI verified the presence of left hemispheric atrophy and large left-sided parietotemporopariietal porencephalic cyst and revealed an ipsilaterally malformed hippocampus medial temporal region with increased FLAIR signal (Fig. 1A). An ipsilateral multilobar, parieto-occipital resection, sparing the mesial temporal structures, rendered the patient seizure free (Fig. 1B).
2.2. Case 2

Patient 2 was a 25-yr-old man with lifelong history of intractable seizures secondary to serious posterior left head trauma sustained at the age of 4 months. Magnetic resonance imaging (MRI) presented posterior left hemispheric atrophy, a prominent area of tissue loss and gliotic lesion over the lateral temporoparietal region and ipsilateral hippocampal atrophy plus temporal horn enlargement (Fig. 2A). Surgical treatment involved extensive resection of the lateral temporo-parieto-occipital area, including the principal gliotic region and the lateral-inferior occipital region, sparing the medial temporal area (Fig. 2B), and achieved seizure freedom.

3. Discussion

Given the recent introduction of the new classification scheme for FCD, it is not surprising that there is not yet much information regarding the incidence of IIId FCD among patients with epilepsy with early-life-acquired lesions and corresponding surgical outcomes. It is very likely that this type of pathology eluded detection in previous years, the neuropathologic focus being the "principal" lesion. Some deductions, however, concerning surgical outcome could perhaps be attempted. Krsek et al. [4] reported on a series of 25 children having suffered pre- or peri-natal (mostly ischemic) injury, all with a broad spectrum of accompanying cortical dysplastic changes, mostly type I FCD (which apparently conforms to type IIIId current classification). Slightly more than half of these patients were offered lobar and multilobar resections with class I Engel outcomes achieved in ≈ 50% of them. There was a high incidence of extensive and not well-localized interictal and ictal EEG abnormalities in this series, suggesting the presence of large and diffuse epileptogenic zones. Tassi et al. [5] reported on a large surgical series of type I FCD. A subgroup of eight cases had had an early-life-acquired anoxic–ischemic or inflammatory lesion. All eight patients underwent multilobar resections with class I Engel outcomes achieved in only 25% of them. No data are available concerning seizure semiology and surface EEG localization. In contrast to the abovementioned studies, Iida et al. [6] reported Engel class I outcomes in 6/8 patients with congenital porencephalic cysts undergoing focal cortical resections guided by acute intraoperative ECoG. Most of these patients had semiological evidence plus well-localized and lesion-concordant surface EEG findings in the presurgical work-up, implicating discrete cortical areas for ictal genesis.

An interesting feature in both our cases was the MRI presence of additional medial temporal pathology on the side of operation. This could be compatible with medial temporal sclerosis in patient 2, while patient 1 presented more complex features of hippocampal–parahippocampal region white–gray matter blurring, increased T2/FLAIR signal and malformed/abnormally rotated hippocampus. These findings should raise the possibility of medial temporal ictal onsets, given literature data suggesting such a relationship in patients with congenital extratemporal porencephalic cysts and medial temporal sclerosis [7–9]. However, neither semiology nor surface EEG localization supported such a hypothesis. A subtemporal strip placed in patient 2

Fig. 1. Presurgical (upper row, A) and postsurgical (lower row, B) FLAIR MR images of patient 1.

Fig. 2. Presurgical (upper row, A) and postsurgical (lower row, B) T1 MR images of patient 2.
demonstrated only late involvement of the mediobasal temporal region during seizures. In addition, both patients enjoy excellent seizure control, 30 months and 18 months postoperatively, without having resected medial temporal structures. Among 29 patients with pre- or peri-natal hypoxia and low-grade FCD [4], 6 were found to have medial temporal sclerosis on MRI. All of these patients underwent extensive resections (mostly hemispherectomies) including medial temporal structures, so it is difficult to deduce the epileptogenic potential the lesioned medial temporal region might possess. Among 24 patients with congenital extratemporal porencephaly, Burneo et al. [8] reported 9 with additional medial temporal sclerosis; an incidence very close to that of Krsek’s study. Five of these patients had ipsilateral temporal lobe ictal onsets and achieved class I Engel outcomes following temporal lobectomy. However, 3 others had extratemporal ictal onsets, based on video-EEG findings. Unfortunately, there is no information regarding whether these patients underwent operation. Ho et al. [7] reported evidence of medial temporal atrophy/sclerosis, based on MRI hippocampal and amygdalar volumetry in 21 out of 22 cases with congenital porencephaly. Only 4 patients underwent operation, all temporal lobectomies, with favorable results. Again, no information is available regarding patients with extratemporal ictal onsets. Iida et al. [6], in contrast, reported ictal onsets localized in the vicinity of congenital extratemporal porencephalic cysts with excellent results (Engel class I) following tailored resections. However, there are no data regarding the presence of medial temporal pathology. Concerning neuropathologic classification, the recent proposal [1] does not offer a specific subcategory among type III FCD for cases with the constellation of FCD, early-life injury and medial temporal sclerosis, or any other medial temporal malformative lesion, if present distant to the principal early-life lesion and neighboring dysplasia.

4. Conclusion

Our findings, along with literature data, suggest that the organization of the epileptogenic substrate in type IIId FCD probably varies along a spectrum including on the one side cases with extensive ictal onset zones and non-satisfactory outcomes following focal resections and on the other side cases with well-localized ictal onsets and Engel class I outcomes following tailored resections, even though MRI may suggest more extensive and complex pathology. Within such complex imaging lesions, the epileptogenic potential is not necessarily uniformly distributed, and abnormal areas (such as the hippocampal-medial temporal region) may actually not contribute to ictogenesis, while in other cases be an essential component of it. The real challenge in presurgical evaluation is, therefore, to carefully determine which part, if any, of the whole abnormality is critical for seizure onsets. Our experience suggests that stereotyped focal-onset seizure semiology along with concordant localized interictal/ictal EEG and functional data is essential for identifying such candidates.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.ebcr.2012.11.002.

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