Seizure xxx (2010) xxx–xxx

Contents lists available at ScienceDirect

Seizure

journal homepage: www.elsevier.com/locate/yseiz

Epilepsy surgery of posterior quadrant dysplasia in the first year of life: Experience of a single Centre with long term follow-up

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ARTICLE INFO

Article history:
Received 8 April 2010
Received in revised form 14 September 2010
Accepted 17 September 2010

Keywords:
Posterior quadrant dysplasia
Hemi-hemimegalencephaly infancy
Surgery
Drug-resistant epilepsy
Cognitive delay

ABSTRACT

Posterior quadrant dysplasia (PQD) is a rare variant of cortical dysplasia involving the posterior regions of a single hemisphere. It is always associated with early onset, refractory epilepsy often characterized by a "catastrophic" evolution. The experience on its surgical management during the first year of life is limited to sporadic, isolated cases.

Between 2002 and 2005, four children less than one-year-old and affected by drug-resistant epilepsy associated with PQD were admitted to our Institution and underwent surgical treatment.

One patient remained seizure-free during all the follow-up (Engel I). The remaining three children showed a recurrence of the seizures, requiring subsequent surgical procedures in two cases. In one case (Engel II), the seizure control has been obtained thanks to pharmacological treatment. The other two patients respectively had only a partial (Engel III) and a less relevant reduction of the number of seizures (Engel IV).

Both the epileptic and the neuropsychological outcome of our series were significantly influenced by persistent contralateral interictal anomalies rather than by the timing of the surgical procedure.

Unpredictable results should be expected in this kind of patients if there is the detection of contralateral independent epileptiform activities on the EEG at diagnosis. Parents and relatives should be aware of the results' variability, even though a reduction of seizures may be expected, enabling an easier handling of the child's condition.

1. Introduction

A significant proportion of candidates to epilepsy surgery are affected by malformations of cortical development (MCD).[1] Focal cortical dysplasias (FCD) currently represent up to one fifth of MCD, as result of the improved diagnosis by means of high resolution MRI in the last decade. A successful long term follow-up after surgical resection is often reported in FCD, the seizure-free rates being about 75%.[2,3] On the other hand, the more diffuse forms of cortical dysplasia, which possibly extend over multiple lobes, are burdened by a worse outcome.[4,5]

Posterior quadrant dysplasia (PQD) involves the posterior areas of a single hemisphere, namely the occipital lobe and the posterior part of parietal and temporal lobes. Its management is particularly challenging. Actually, PQD is a rare variant of cortical dysplasia, and only a few cases have been reported in the literature so far. The experience on its surgical management during the first year of life is limited to sporadic, isolated cases. The proximity or overlapping of eloquent areas, the poor visualization of the edges of the abnormal tissue, and the difficult localization of the epileptogenic foci give reason of the failures in the surgical treatment.[4–7]

Here, we present a series consisting of four children who required surgery during the first year of life because of drug-resistant epilepsy sustained by PQD. The goal is to report on the long-term follow-up of these patients to provide further information on such an uncommon and challenging pathological condition.

2. Patients and methods

Between 2002 and 2005, four children less than one-year-old (2 boys and 2 girls, mean age 6 months, range: from 5 to 7 months) and affected by intractable epilepsy associated with PQD were admitted to our Institution to be considered for a possible surgical management. All the patients experienced a period of non-effective drug treatment (mean duration 6 months, range: from

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doi:10.1016/j.seizure.2010.09.015

5 to 7 months) that included at least four drugs in monotherapy or in combination. Before and after surgery, they were assessed according to the following procedures:

- detailed clinical history and physical examination;
- ictal/interictal prolonged scalp video-EEG recordings;
- high resolution MRI;
- neuropsychological tests; and
- behaviour observation.

After surgery, the patients were followed with serial assessment every 6 months during the first year and, afterwards, yearly.

2.1. Seizure classification

Epilepsy was classified according to the International League Against Epilepsy (ILAE) classification.[8]

Seizure outcome was assessed using the Engel's scale.[9]

2.2. Video-EEG

Videopolygraphic study was performed using 11 EEG electrodes according to the 10/20 International System. Deltoid surface electromyogram (EMG) also was recorded. Every patient underwent numerous recordings when awake and repeated, sometimes prolonged, sleep recordings; each patient had at least one nocturnal polysomnography video-EEG recording lasting >24 h.

2.3. Neuroimaging

All the patients were examined using a 1.5 Tesla MR system (Horizon Echospeed/Excite, General Electric, Milwaukee, USA). Angio-MRI or angio-CT scans were also performed.

2.4. Neurological evaluation

Motor function was estimated pre and post-operatively through a careful neurological examination. Visual function was assessed using a test battery that includes: ocular movements (spontaneous and in response to a target), the ability to fix and follow a target, and the visual field.

2.5. Neuropsychological examination

The developmental assessment was obtained using the Griffiths Mental Development Scales. The Griffiths' scales[10] were performed to assess the developmental quotient according to the single scales (locomotor, personal social, hearing and language scale, hand–eye coordination, performance, practical reasons) and the general developmental quotient (GQ). The Vineland Adaptive Behaviour Scales[11] were administered to evaluate the adaptive behaviour. Mental development disorders were defined according to the Diagnostic and Statistical Manual of Mental disorders (DSM V-TR).

3. Results

3.1. Pre-surgical data

The details of each patient are summarized in Table 1.

3.2. Clinical history

Three patients had uncomplicated pregnancy and normal delivery; one patient (# 1) had respiratory distress at birth. Family history of epilepsy was noticed in two cases (# 1 and 4).

3.3. Epileptic findings

The mean age at epilepsy onset was 1 month (range: 4 days to 3 months). Three patients presented the first seizures within the first week of life. Seizures were partial in all cases, meanly lasting 1–4 min, and followed by clusters of asymmetrical spasms. The frequency was as high as 20–100 episodes per day, despite multiple antiepileptic drug administration (including ACTH).

The interictal EEG mainly showed asymmetrical background abnormal activity (slow waves) and repetitive paroxysmal activity of high amplitude spikes and poly-spikes on the affected hemisphere, with predominance on the posterior and temporal regions. In three cases (# 1, 3 and 4) independent contralateral abnormalities were detected; in particular, contralateral subclinic epileptic discharges were recorded in one case (# 3). In all patients a clear asymmetrical hypsarrythmic pattern appeared 2–3 months after epilepsy onset.

In case of partial seizures, ictal EEG showed discharges of theta-rhythmic activity localized on the posterior regions of the affected side, with contralateral and anterior spreading. In case of spasms, it documented diffuse high-voltage slow waves followed by low voltage rapid activities, exclusively or predominantly on the affected hemisphere.

Corresponding clinical signs during ictal discharges were not always detectable and sometimes asymmetrical spasms were evident only at the end of the discharges.

3.4. Neurological and neuropsychological findings

Axial hypotonia, asymmetric movements with slight hemiparesis contralateral to the brain lesion, severe impairment of the visual function and strabismus were detected in all cases. The head was constantly rotated contralaterally to the affected side, as result of possible visual hemi-inattention. Two patients showed nystagmus (# 1 and 4).

A severe developmental delay was found in one case (# 1), while the remaining three children had a low average GQ. A disharmonic developmental profile with worse results in motor, performance, and hand-eye coordination scales was observed. All patients showed stereotyped movements, lack of eye contact, poor responsiveness and irritability.

3.5. Neuroimaging

Neuroradiological findings of PQD were found in all cases. PQD involved the left hemisphere in all but one patient (# 2). The affected cortex showed a hyperintense MRI signal on T1 and hypointense on T2. A dysmorphic aspect of the homo-lateral ventricle was constantly present. Two main different morphological patterns of irregular cortical thickening were noticed: pachygyria in two children (# 2 and 4) and polimicrogyria in the remaining two (# 1 and 3) (Figs. 1–3).

3.6. Surgical procedures

Age at surgery ranged from 5 to 7 months (mean: 6 months). The total excision of the PQD was obtained in one case (# 2) and a subtotal removal in another one (# 4). Because of the poor differentiation between normal and abnormal tissue, a partial excision of the dysplasic tissue was realized in the remaining two children (# 1, 3) together with functional hemispherectomy. However, these two patients required a further operation due to the persistence of clinical and subclinical ictal activities on the affected side, associated with contralateral paroxysmal discharges (# 3). The excision of the residual PQD was performed in both cases, 1 year (# 1) and 4 years (# 3) after the first operation.

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Table 1
Summary of the pre-operative and post-operative findings.

Pre-surgery

Case #/sex	Age at seizure onset	Seizure findings and AED	EEG	Neurological findings	Griffiths (DQ; mental age: MA)	Visual function	VABS (adaptive level)
1/M	4 days	Partial seizures, followed by clusters of asymmetrical spasms (frequency: 100/die). VPA, PB, CBZ, CLB, CNZ, VGB, TPM, ACTH	Interictal: (*at onset*) asymmetrical BA and subcontinuous left CPTO sW and SW; (*at 3 months*) asymmetrical hypsarrythmic pattern (L > R); independent and rare right isolated S. Ictal: (*focal seizures*) left PTO theta-rhythmic activity; (*spasms*) left high-voltage sW followed by low voltage fast activities	Axial hypotonia, slight R hemiparesis and visual hemi-inattention, nistagmus	DQ: 28 (at 7 months)	Fixation, visual attention and OKN: abnormal. Acuity cards: not testable	Communication, daily living skills and socialization: low
2/F	3 months	Partial seizures, followed by clusters of asymmetrical spasms frequency: 20/die). VGB, TPM, CBZ, CLB, ACTH	Interictal: (*at onset*) asymmetrical BA and subcontinuous right CPTO SW and sW, spreading on the right side; (*at 6 months*) asymmetrical hypsarrythmic pattern (R > L). Ictal: (*focal seizures*) right PTO theta-rhythmic activity; (*spasms*) right high-voltage sW followed by low voltage fast activities	Axial hypotonia, slight L hemiparesis and visual hemi-inattention	DQ: 90 (at 7 months)	Fixation, visual attention, OKN and acuity: abnormal	Communication, daily living skills and socialization: adequate
3/M	7 days	Partial seizures, followed by clusters of asymmetrical spasms (frequency: 80/die). VPA, VGB, TPM, CNZ, BMZ, ACTH	Interictal: (*at onset*) asymmetrical BA and asymmetrical hypsarrythmic pattern (L > R); (*at 2 months*) independent right isolated S and SW. Ictal: (*focal seizures*) left PTO theta-rhythmic activity; (*spasms*) left high-voltage sW followed by low voltage fast activities. Subclinical theta/rhythmic discharges on the right posterior regions	Axial hypotonia, slight R hemiparesis and visual hemi-inattention	DQ: 83 (at 5 months)	Fixation, visual attention and OKN: abnormal. Acuity cards: not testable	Communication, daily living skills and socialization: low
4/F	3 days	Partial seizures, followed by clusters of asymmetrical spasms (frequency: 50/die). VPA, VGB, CNZ, BMZ, ACTH	Interictal: (*at onset*) asymmetrical BA and subcontinuous left CPTO sW and SW; (*at 2 months*) asymmetrical hypsarrythmic pattern (L > R); independent and rare right isolated S. Ictal: (*focal seizures*) left PTO theta-rhythmic activity; (*spasms*) left high-voltage sW followed by low voltage fast activities	Axial hypotonia, slight R hemiparesis and visual hemi-inattention, nistagmus	DQ: 87 (at 5 months)	Fixation and visual attention: abnormal. Acuity and OKN: not testable	Communication, daily living skills and socialization: low

Post-surgery

Case #	Age at surgery and follow-up duration	Epileptic findings (Engel)	EEG	Motor abilities	Griffiths	Visual function	VABS (adaptive level)
1	7 months and 17 months FU: 6 years	III AED at the follow-up end: TPM, CLB	Interictal: asymmetrical BA; no hipsarrythmic patterns. After *3 months* left FCT S and sW; after *9 months* interictal right focal spikes. Ictal (*9 months*): left fast activity predominat on FCT	Improved, still not able to walk	DQ: 29 (MA 17 months)	Fixation, visual attention and OKN: improved. Acuity: unchanged	Communication, daily living skills and socialization: improved
2	7 months FU: 5 years	I AED at the follow-up end: TPM	Interictal: **a**symmetrical BA; no hipsarrythmic patterns. After *8 months* right FCT S and sW	Improved, able to walk without support at 2 years	DQ: 97	Fixation, visual attention and OKN: improved. Acuity: borderline	Communication, daily living skills and socialization: improved adequate (improved)
3	5 months and 4 years 5 months FU: 7 years	IV AED at the follow-up end: DPA, CLB	Interictal: asymmetrical BA; no hipsarrythmic patterns. After *2 months* left FCT S and sW; (*at 9 months*) right interictal focal spikes. Ictal (*at 16 months*): left fast activity predominat on FCT, spreading on the right side. Subclinical theta/rhythmic discharges on the right posterior regions	Improved, still not able to walk	DQ: 39 (MA: 11 months)	Fixation: improved. Visual attention, acuity and OKN: unchanged	Communication, daily living skills and socialization: improved
4	5 months FU: 8 years	II AED at the follow-up end: DPA, TPM, CLB	Interictal asymmetrical BA; no hipsarrythmic patterns. After *12 months* left FCT SW; *21 months* independent right PTO S and SW	Improved, able to walk without support at 4 years	DQ: 53	Visual attention: unchanged. Fixation and acuity: improved	Communication, daily living skills and socialization: improved

BA: background activity; sW: slow waves; SW: spike-waves; S: spikes; PTO: parieto-temporal-occipital; CPTO: centro-parieto-temporal-occipital; FCT: fronto-centro-temporal; M.A: menta. Age. P: pathological. AED: antiepileptic drugs; VPA: valproate; PB: phenobarbital; CBZ: carbamazepine; CLB: clobazam; CNZ: clonazepam; BMZ: bromazepam; VGB: vigabatrin; TPM: topiramate; OKN: optocinetic nystagmus; and VABS: Vineland Adaptive Behaviour Scales.

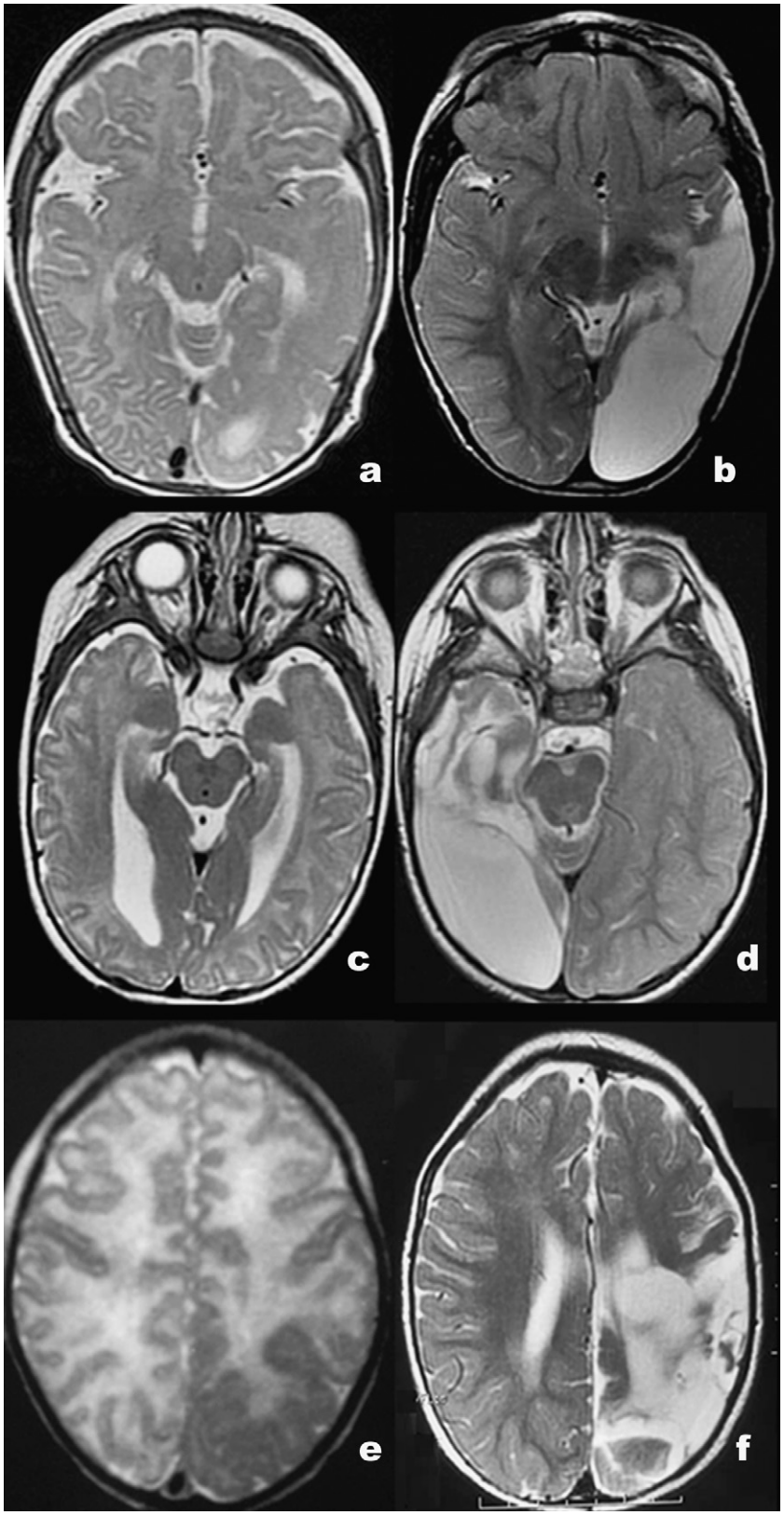

Fig. 1. Preoperative (a, c and e) and postoperative (b, d and f) axial T2w MR images of cases 1 (a and b), 2 (c and d) and 3 (e and f) showing volume reduction of the affected occipital and temporo-parietal lobes, associated with diffuse irregular cortical thickening with prevalent polimicrogyric pattern in cases 1 and 3, and pachigyric pattern in case 2; the altered cortex is characterized by mildly hypointensity if compared to unmyelinated subcortical white matter. The post-operative images show the temporo-parieto-occipital CSF-filled surgical cavities.

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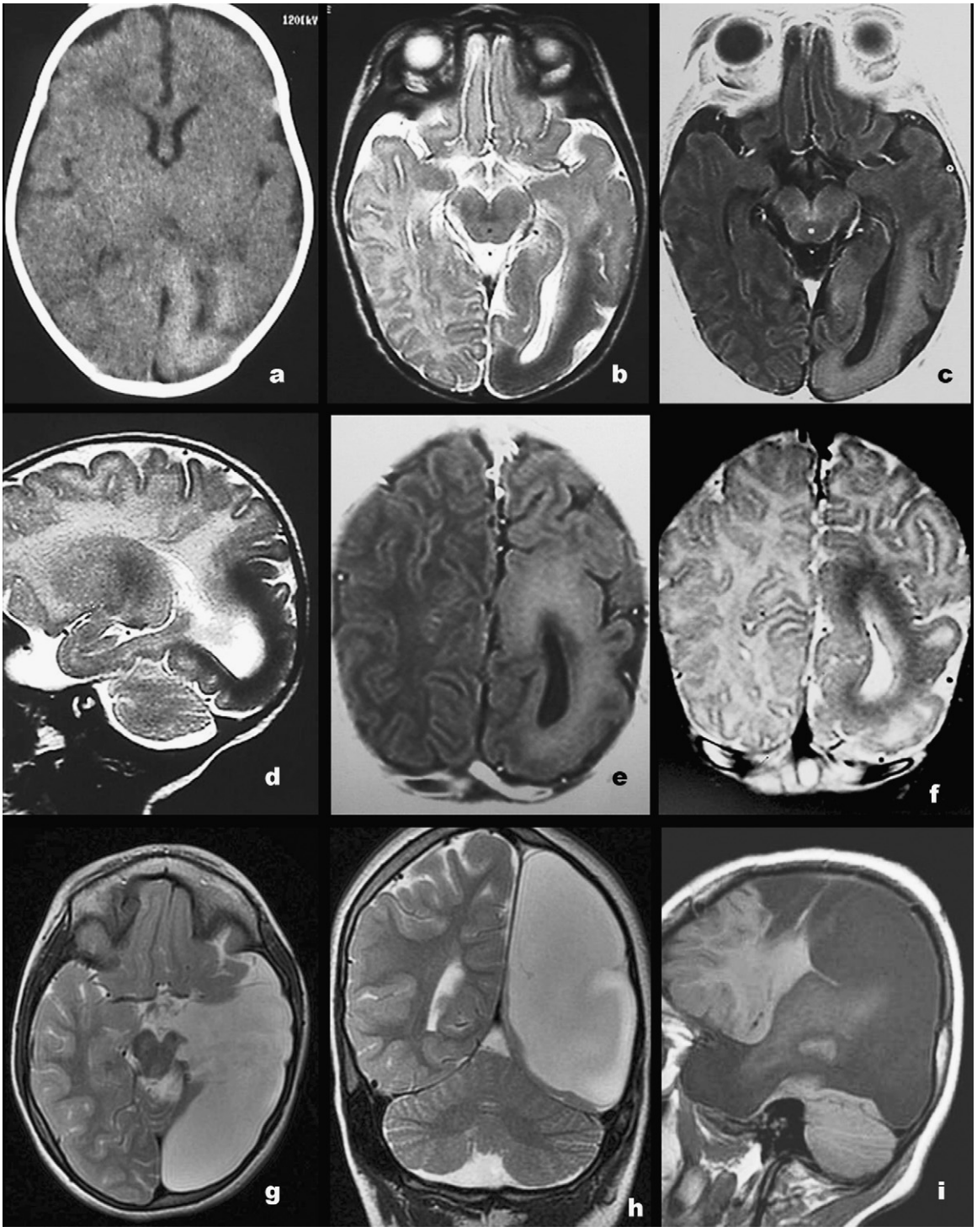

Fig. 2. Case 4: (a) axial CT scan image, (b and c) axial T2w and T1w images, (d) sagittal T2w image and (e and f) coronal T1w and T2w images. CT and MR images show volume reduction of left temporal and occipital lobes, associated with marked cortical thickening, with prevalent pachigyric pattern; the occipital cortex is smooth, with few cerebral gyri and sulci, characterized by mildly hyperdensity on CT scan image, hyperintensity on T1w images and hypointensity on T2w images, if compared to unmyelinated subcortical white matter. The left lateral ventricle is abnormally dilated. (g) Axial T2w image, (h) coronal T2w image and (i) sagittal T1w image. MR images show temporo-parieto-occipital CSF-filled surgical cavity with complete removal of the lesion.

The postoperative course was uneventful since no major complications were observed after all the procedures. Patient # 1 required the endoscopic marsupialisation of a left periventricular cyst that developed 2 years after the second operation and caused the recurrence of left fronto-central seizures.

3.7. Pathology

The pathologic analysis of surgical specimens revealed structural abnormalities consistent with severe cortical dysplasia. Typical disorganization of the laminar architecture was observed,

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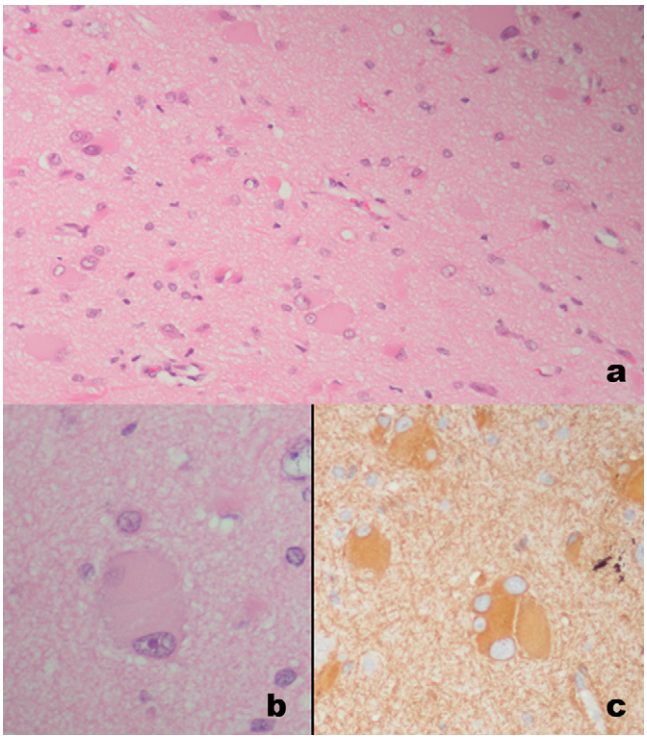

Fig. 3. Representative histopathological images: (a) cortical dysplasia with abnormal cortical architecture (H&E stain), (b) balloon cells with glassy eosinophilic cytoplasm lacking cellular processes are visible on higher magnification (H&E stain) and (c) abundant pericellular immunostaining for synaptophysin is observed over the ectopic neurons in the subcortical white matter.

with sparse balloon cells inside the cerebral cortex and the subjacent white matter.

3.8. Post-surgical follow-up

The follow-up ranged from 5 to 8 years (average: 6.5 years).

3.9. Epileptic outcome

One patient (# 2) remained seizure-free during all the follow-up (Engel I). The remaining three children showed a recurrence of the seizures respectively by 3, 6 and 30 months from the first surgical procedure. They mainly presented with partial motor seizures, gelastic and autonomic seizures. Patients # 1 and 3 had showed independent paroxysmal activities (spikes) contralateral to the lesion on preoperative EEG assessment (patient # 1 also presented subclinic epileptic discharges on the unaffected side).

Early postoperative EEG recordings showed an asymmetrical background activity without paroxysmal abnormalities or hipsarrythmic patterns. In all cases, interictal paroxysmal activities (spikes and spike-wave complex) were detected on the residual regions of the affected hemisphere three months after the first operation. During the follow up, clinical and subclinical ictal activities were detected on the affected side in two cases (# 1 and 4), and contralaterally in one (# 3). Contralateral interictal focal abnormalities (spikes) were observed in three cases (#1, 3 and 4).

Patient #4 currently maintains a seizure control (Engel II) thanks to the pharmacological treatment (VPA, TMP and CLB). In patients #1 and 3 only a significant (Engel III) and a less relevant reduction of seizure severity (Engel IV) has been achieved, respectively.

3.10. Neurological and neuropsychological outcome

Hemi-paresis and strabismus still persist in all cases. Axial hypotonia disappeared in one case (# 2), who started walking alone at two years of age; it was significantly reduced in the other three patients even though only one of them was able to begin walking at the age of 4 (case # 4). Nystagmus improved in all patients. A significant improvement of visual attention was observed in all patients.

Two patients show a severe mental retardation (cases # 1 and 3) and one a moderate grade of mental delay (case # 4). Only one child has a borderline cognitive development (case # 2). Adaptive behaviour is better in the two children with minor emotional disorders and without a motor impairment (cases # 2 and 4).

The quality of life as experienced by the parents has improved in all cases.

4. Discussion

PQD, also addressed as hemi-hemimegalencephaly,[4] represents a challenging problem. This malformation actually involves the occipital, parietal and temporal lobes of one hemisphere, and is always associated with early onset, refractory epilepsy often characterized by a "catastrophic" evolution. Consequently, the patient's psychomotor development is progressively impaired and the quality of life definitely poor. This condition advises the epilepsy specialists to consider the surgical treatment as early as possible to avoid such a clinical deterioration. Only a few data from the literature are currently available with regards of children affected by PQD and operated on early, that is within the first year of age. Indeed, infants are sporadically described, usually as part of series including older children and adults, so that only anecdotic cases are reported.[4,5,12]

In this subset of children, as demonstrated by our experience, the EEG at onset is often characterized by a focal pattern that tends to evolve into a hypsarrhythmic pattern within 2 or 3 months. Due to the refractory epilepsy and the progressive cognitive worsening, early surgical treatment is indicated, as well as in other cases of hypsarrhythmia associated with focal cortical injuries, as suggested by other authors.[13–15]

In our series, all patients presented with infantile spasms associated with partial seizures. Despite being a generalized epilepsy frequently associated with multiple epileptogenic zones, the occurrence of infantile spasms should not be considered as a contraindication for surgery, in particular when focal features are observed, namely focal lesion, focal neurological signs, history of prior or ongoing focal seizure and focal interictal EEG findings.[13–16] Moreover, patients with infantile epileptic encephalopathy may have a better post-surgery outcome, especially in terms of developmental gains. Indeed, they often undergo surgical procedures at an earlier age, with a consequent shorter duration of epilepsy.[14,15] On the other hand, the surgical outcome seems to be mostly influenced by the duration of the clinical history. A significant difference of outcome has been actually observed in patients with infantile spasm depending on whether they had been controlled by drugs or they were medically refractory from the beginning.[15]

The patients affected by infantile spasms often present increased contralateral interictal spikes, contralateral background slowing and ipsilateral and contralateral paroxysmal fast activity (PFA), as we observed in all our patients. Precocious contralateral epileptic abnormalities are usually considered as unfavourable prognostic factor. In contrast, Wyllie et al.[17] recently reported a study on a series of patients affected by refractory symptomatic epilepsy presenting generalized or bilateral findings on pre-operative EEG; post-surgical outcome (72% rate of seizure-free patients) was similar to that of a

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comparison group of patients with similar MRI findings and only ipsilateral electrical abnormalities. They supposed that the contralateral epileptiform discharges may come from a potentially reversible secondary epileptogenesis resulting from an interaction between the early lesion and the developing brain. These findings were not confirmed in our experience: 3 out of the 4 patients with pre-operative independent controlateral interictal abnormalities showed recurrent seizures respectively 3, 6 and 30 months after the first surgical procedure. Accordingly, unpredictable results should be expected in patients affected by posterior quadrantic dysplasia with the detection of contralateral independent activities. Parents and relatives should be aware of the results' variability, even though a reduction of seizures may be expected, enabling an easier handling of the child's condition.

As for the epileptic outcome, also the neuropsychological outcome of our series was significantly influenced by persistent contralateral interictal anomalies rather than by the timing of surgical procedure (see Table 1). Anyway, an overall improvement of the quality of life was observed in all cases with a better daily management of these patients.

As far as commonly reported, the more extensive the excision of the PQD, better the outcome.[18–20] In our small sample, the worse results were obtained for those patients (# 1 and 3) who underwent partial lesionectomy as first surgical step and who showed an abundance of pre-operative independent contralateral interictal EEG abnormalities.

One of the possible mechanisms concurring to the variability of the results is the difficulty in differentiating the borders of the abnormal tissue from the normal cortex which makes it difficult to obtain a complete removal of the lesion. Moreover, even with the aid of intra-operative electrophysiological monitoring, the surgical attitude should be to tailor the excision considering the high risk of damaging functional areas, and to limit to selected cases the complete removal of the lesion as first surgical step. On these grounds, some authors[5,12] advocated the association of functional posterior disconnective procedures with lesion excision. In the multicentric study of D'Agostino et al.,[4] surgical treatment was tailored on the single patient, 4 patients undergoing primary posterior disconnection beyond the removal of the dysplastic lesion at the last follow-up (range, 1.5–7 years; mean, 4 years). Such an attitude resulted in complete seizure control in one patient, Engel Class II in another one and Class III in the remaining two patients. According to these authors, even a class III outcome with rare seizures has to be considered worthwhile, allowing the resumption of neurocognitive developmental progress and an improvement of behaviour. Better results were obtained by Daniel et al. who operated on two infants who showed an optimal epileptic outcome (Engel I). No patients in both studies presented pre-operative independent controlateral interictal abnormalities.

Major early postoperative complications have been reported in 5% of the cases following hemispherotomy, including significant intraoperative blood loss, electrolyte imbalance, and hypothermia; moreover, late complications, namely hydrocephalus and postoperative haemorrhages, have been observed.[21] No major early or late complications were observed in our series, even in the two patients who underwent hemispheric disconnective procedures at first surgery.

5. Conclusions

Early surgical treatment represents the mainstay in the management of children with cortical dysplasia and refractory epilepsy, also when multiple lobes are involved as in quadrantic forms. Though on a relatively small number of patients, our series confirms that, even in children operated on in the first months of life, the most important negative prognostic factor for the seizures outcome is represented by the persistence of controlateral slowing and paroxysmal fast abnormal EEG activity after surgery. Persistent abnormal controlateral EEG findings, as expected, also negatively influence the neurocognitive outcome, independently from the age at surgery. On the contrary, the final extent of surgical excision of the dysplastic tissue did not seem to relate with the final prognosis.

Conflict of interest statement

The authors report no conflict of interest.

Acknowledgement

The authors thank the association "AREF onlus", www.arefonlus.it, for its support.

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Please cite this article in press as: Novegno F, et al. Epilepsy surgery of posterior quadrant dysplasia in the first year of life: Experience of a single Centre with long term follow-up. Seizure: Eur J Epilepsy (2010), doi:10.1016/j.seizure.2010.09.015