



Combined SEEG and source localisation study of temporal lobe schizencephaly and polymicrogyria

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ARTICLE INFO

Article history:

Accepted 26 June 2009

Available online 24 July 2009

Keywords:

Schizencephaly

Polymicrogyria

Temporal lobe epilepsy

Source localisation

Stereo-EEG

ABSTRACT

Objectives: Type 1 schizencephaly (SZ) is a cerebral malformation characterised by a cleft lined and surrounded by a polymicrogyric cortex, extending from the pial region to the peri-ventricular heterotopia. Our purpose was to combine and compare dipole source imaging technique and Stereo-EEG (SEEG) technique in determining the irritative and epileptogenic zones in a case of type 1 schizencephaly.

Methods: High-resolution (64-channel) video-EEG with electrical source imaging and SEEG recordings were performed during a pre-surgical evaluation for medically intractable epilepsy.

Results: Anatomico-electro-clinical correlations based on SEEG and source localisation identified two irritative and epileptogenic zones partially overlapping the polymicrogyric cortex surrounding the SZ: an anterior medio-lateral network primarily involving dysplastic limbic structures and a lateral network involving the anterior and middle part of the cleft and polymicrogyric cortex. The most posterior part (at the temporo-parieto-occipital junction) displayed a normal background activity.

Conclusions: Both epileptogenic and electrophysiologically normal cortices coexisted within the same widespread malformation: only the anterior part belonged to the anterior medio-lateral epileptogenic network defined by the SEEG.

Significance: In cases of widespread cortical malformation such as SZ, source localization techniques can help to define the irritative zone and relevant targets for SEEG.

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1. Introduction

Schizencephalies (SZs) are cerebral malformations characterised by a cleft, lined and surrounded by a dysplastic and/or polymicrogyric cortex, extending from the pial region to the lateral ventricle and/or peri-ventricular heterotopia (Yakovlev and Wadsworth, 1946). SZs are divided into two types: (i) type 1 or “closed lip” SZ, when the marginal layers of the cortical banks join each other; and (ii) type 2 or “open lip”, when the banks are separated from each other. In a new genetic and developmental classification of cortical malformations, SZs have been put together with polymicrogyria (PMG) in the category of malformations related to abnormal cortical organisation (Barkovich et al., 2005). Prevalence of focal infra-sylvian SZs in temporal lobe epilepsy has been estimated to be around 0.4% (Lehericy et al., 1995).

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In type 1 SZ, epilepsy is the principal mode of presentation and might be medically intractable. Published case reports of type 1 SZ do not demonstrate a clear link between the lobar localisation of SZ and the presumed anatomical origin of the associated partial seizures. Some data suggest that the anatomical origins of seizures are related to SZ, particularly in frontal and parietal SZ (Granata et al., 1996). There are only a few case reports of type 1 SZ explored with video-EEG (Leblanc et al., 1991; Landy et al., 1992; Silbergeld and Miller, 1994; Maehara et al., 1997). According to these studies, there is no clear relationship between epileptogenic zone and SZ: in some cases, seizures may arise from the malformed area (e.g. primary sensory-motor cortex seizures associated with rolandic SZ); in other patients, seizures could originate exclusively from distant sites (e.g. temporal lobe seizures associated with rolandic SZ) (Silbergeld and Miller, 1994). A recent stereo-EEG (SEEG) study of PMG (Chassoux et al., 2008) included two cases of focal SZ and showed that the polymicrogyric cortex surrounding the cleft displayed intrinsic epileptogenicity. In these cases, the limited spatial extent of the malformation overcame the issue of sampling bias inherent to this technique. However, in cases of larger cortical

malformations, the source localisation technique may provide complementary information about the relationship between the malformation and irritative zone (Burneo et al., 2004a,b; Gavaret et al., 2004).

Our aim was to compare contribution of dipole source imaging, which is a non-invasive technique, and SEEG in determining the irritative and epileptogenic zones in a case of complex and large temporal malformation involving type 1 SZ, PMG and dysplastic limbic structures.

2. Methods

2.1. Patient

Our patient was a right-handed, 27-year-old woman with no history of neurological or psychiatric disorders. She had undergone normal education. Epilepsy had started at the age of 18 years, with diurnal complex seizures and rare secondary generalisations. Usual seizures started with an initial sensation of ascending warmth associated with a dreamy state. There was an early loss of contact. Seizures lasted more than 1 min and occurred several times a week, with more than two secondary generalisations per year.

Pre-surgical evaluation included medical history, neuropsychological testing, 10/20 scalp video-EEG monitoring, high-resolution video-EEG recordings (64-channel, 10/10 international system), cranial MRI, cranial FDG-PET study and SEEG recordings.

2.2. High resolution video-EEG recordings and source analysis

Recordings were made with 64 new EEG-MRI sensors (Koessler et al., 2007, 2008) taped with collodion onto the head of the subject according to the 10/10 international system (Oostenveld and Praamstra, 2001). EEG data were referenced to Fpz. The signal was recorded at a 1 kHz sampling rate (SD64 Headbox, Micromed, Italy). Spatial localisation of the electrodes was carried out by automatic localisation and labelling of the EEG sensor (ALLES), as described by Koessler et al. (2008).

Source analysis of inter-ictal and ictal events was performed using an equivalent current dipole model (Scherg and Berg, 1991), MUSIC (Mosher et al., 1992) and distributed sources imaging (sLORETTA) (Pascual-Marqui et al., 2002) implemented with ASA software (ANT[®]; Enschede, Netherlands). The MUSIC approach is a scanning method which uses the same source model as a dipole fit, but the number of sources does not need to be known in advance. After principal component analysis, the MUSIC scan metric gives a distributed view of the contribution of each generator distributed in the cerebral volume. sLORETTA is based on reconstruction of cerebral electric activity in each point of a 3D grid of solution points, the number of points being much larger than the number of measurement points on the surface. Each solution point is considered as a possible location of a current source. The particular constraint in this case is that the method selects the solution with a smooth spatial distribution.

2.3. Stereotactic placement of intracerebral electrodes and SEEG recording

The goal of SEEG was to delineate the epileptogenic zone (Chauvel et al., 1996). The electrode implantation sites were chosen according to non-invasive data collected during the earlier phase of the investigation. Eight electrodes were placed in the right hemisphere targeting the amygdala, hippocampus, para-hippocampal gyrus and anterior part of the SZ, middle temporal gyrus, anterior lip of the parieto-occipital part of the SZ, entorhinal and inferior temporal gyri, and posterior lip of the parieto-occipital part of the SZ (see

Fig. 1 and Table 1 for details). One electrode was placed in the left hippocampus and middle temporal gyrus.

Stereotactic placement of the intracerebral electrodes (Dixi Medical, Besançon, France), consisting of 5–15 contiguous contacts (Talairach et al., 1974), was performed as follows: after induction of general anaesthesia, the Leksell G-frame (Elekta S.A, Stockholm, Sweden) was positioned on the patient's head and a stereotactic MRI (3D SPGR T1 weighted-sequence, TR: 20 ms, TE: 6 ms; matrix 512 × 512, with double injection of gadolinium) was carried out. MRI was imported into a computer-assisted stereotactic module (Leksell Surgiplan[®]; Elekta S.A, Stockholm, Sweden), and electrode trajectories were calculated according to pre-operative planning, with careful avoidance of vascular structures. A post-operative stereotactic CT-scan was then carried out and fused with pre-operative MRI to determine the exact position of each electrode.

SEEG and video monitoring were performed 24 h/day for 4 days in the Neurology Department, University Hospital, Nancy. The signal was recorded at a 1 kHz sampling rate on a 128 channel amplifier (LTM 128 Headbox; Micromed, Italy). Electrical stimulations delivered direct current via a 50 Hz train of 3–5 s or 1 Hz shocks. Impulsion was biphasic and 1000 µs width; intensity was 0.2–3 mA (IRES 600; Micromed, Italy).

2.4. Pre-operative cranial MRI

Pre-operative cranial MRI (Signa 1.5 Tesla; General Electric Medical System, Milwaukee, United States) revealed a right occipito-temporal closed lip SZ with polymicrogyric clefts, extending from the lateral temporal cortex to the hippocampus and nodular periventricular heterotopia lining the right lateral ventricle. A malformation of the hippocampal formation was also observed characterised by a folding fault of the right hippocampus, whose body and head had a round shape, associated with dilation of the right temporal horn. The SZ was surrounded by a polymicrogyric cortex involving the occipito-temporal, inferior, middle and superior temporal gyri and the temporo-parieto-occipital junction (Fig. 1).

2.5. ¹⁸F-DG-PET study

PET scans were obtained during the inter-ictal state, in the resting position with eyes closed, 4 days after the last seizure. PET and CT images were recorded at least 4 days after the last seizure on a Biograph hybrid system involving a 2-detector CT for correction attenuation (Siemens, Knoxville, Tennessee, USA). An activity of 300–400 MBq of ¹⁸F-FDG was injected intravenously after an overnight fast, patients being set in a quiet room with eyes closed. Thirty to 40 min later, CT imaging was initiated and immediately followed by a 3D PET recording of 15 min.

PET images were reconstructed with a 3D-OSEM method (3 iterations and 8 subsets) and displayed with 3.0 × 3.0 × 3.0 mm³ voxels.

3. Results

3.1. 10/20. scalp video-EEG recordings

Inter-ictal EEG showed right temporal theta activity, and two types of paroxysms: spike-waves projecting on FT10 and F8 (Fig. 2A and B), and 8 Hz polyspikes followed by a high amplitude delta wave projecting on T4, T6 and FT10 on 10/20 montages (Fig. 3A). Three seizures were recorded with scalp video-EEG monitoring. Clinically, the patient warned and felt a warm ascending sensation associated with the feeling of being where the last seizure had taken place (strong sensation of familiarity associated with recollection). There was an initial facial flushing

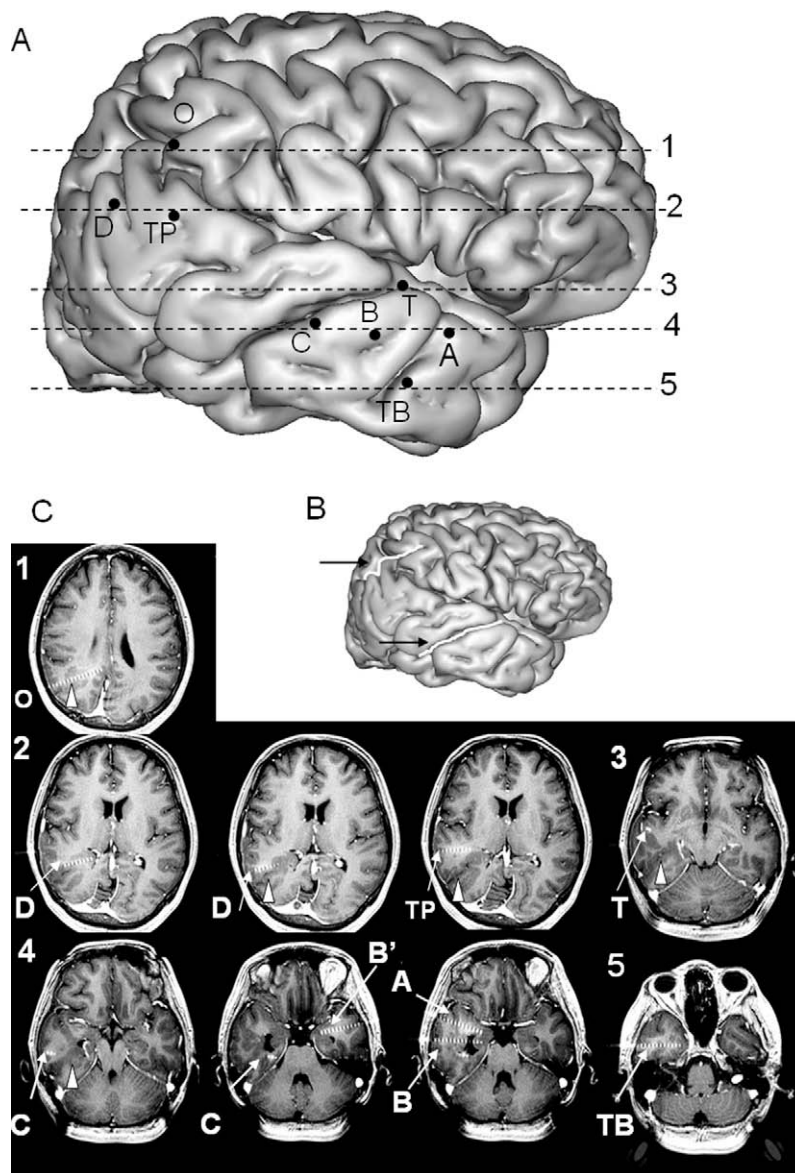


Fig. 1. (A) Depth electrodes scheme superimposed on the 3D surface reconstruction of the patient's cranial MRI (right lateral view). (B) Same lateral view highlighting (white) the schizencephaly (black arrow). (C) Axial slices showing the electrodes trajectories (white arrows) and their position relative to the schizencephaly (white arrow heads). Electrodes placement was the following: in the right hemisphere, electrode A explored the amygdala and the middle temporal gyrus; B explored the anterior hippocampus; anterior schizencephaly and middle temporal gyrus; C explored the posterior hippocampus, anterior schizencephaly and middle temporal gyrus; TB explored the ento-rhinal cortex and the inferior temporal gyrus; T explored the anterior part of the superior temporal gyrus and sulcus; TP explored the posterior schizencephaly and superior temporal gyrus; D explored the posterior schizencephaly; O explored the posterior cingulate gyrus and posterior schizencephaly. In the left hemisphere, B' explored the anterior hippocampus and middle temporal gyrus.

and mydriasis. Loss of contact was inconstant, occurred during the second part of the seizure and was associated with left upper limb elementary gestural automatism. Secondary generalisation began with a left brachio-facial tonic contraction. First ictal EEG modifications were characterised by a right hemispheric flattening followed by a focal right temporal (FT10, F8, T4, FP10) alpha (8 Hz) rhythmic discharge and ending with an irregular high amplitude delta activity.

3.2. Scalp high-resolution (64-channel) video-EEG recordings

10/10 64-channel video-EEG recorded the same two types of inter-ictal paroxysmal activity: (i) focal spike-waves characterized by a negative field projecting on FT8, FT10, FP2 and T8. without

phase reversal, consistent with the modelled radial equivalent dipole (Fig. 2); (ii) more widespread polyspike-waves characterized by a negative field projecting on F8, FT8, P8, FT10, TP8, F6, FC6, C6 and CP6 and a widespread but weaker positive field on averaged referential montage. Using MUSIC and single moving dipole algorithms, source localizations of the first type of inter-ictal event was consistently localised in the right temporal pole (Fig. 2C and D). Source localizations of the polyspike-waves were consistently localised in the lateral and basal part of the right temporal lobe. Sequential sLORETA analysis of the inter-ictal polyspike-waves consecutively localised the modelled generators in the middle and anterior part of the SZ (Fig. 3B and C, steps 1 and 2), then in the basal and lateral temporal gyri (Fig. 3B and C, steps 3–8).

Table 1

Location of depth electrodes and significant results of electrical stimulation.

Electrodes names	Internal contacts	Middle contacts	External contacts	Sites whose stimulation triggered seizures
<i>Right hemisphere</i>				
A	Amygdala	–	Ant MTG	Amygdala (0.6 mA by train, 2 mA by shock)
B	Ant hippocampus	–	Middle MTG	Ant hippocampus (0.6 mA by train)
C	Post H	Ant SZ	Post MTG	–
T	STS	–	Ant STG	–
TP	Post SZ	–	Post STG	–
D	Post SZ	Post SZ	Post SZ, TPOj	–
O	PCG	Post SZ	Post SZ, TPOj	–
TB	EC	Collateral sulcus	ITG	EC (0.6 mA by train)
<i>Left hemisphere</i>				
B'	Ant hippocampus	–	Ant MTG	–

Abbreviations: MTG, middle temporal gyrus; STS, superior temporal sulcus; STG, superior temporal gyrus; post, posterior; SZ, schizencephaly; ant, anterior; CG, cingulate gyrus; TPOj, temporo-parieto-occipital junction; EC, entorhinal cortex and ITG, inferior temporal gyrus.

4. SEEG

Study of the inter-ictal recordings identified two types of paroxysmal events (Fig. 4): (i) abundant spike-wave paroxysms, simultaneously recorded in the amygdala (A, internal leads) and hippocampus (B and C, internal leads) sometimes propagating to the anterior middle temporal gyrus (A and B, external leads); and (ii) bursts of polyspike-wave paroxysms recorded in the anterior part of the middle and inferior temporal gyri (B, C and TB, external contacts), propagating to the anterior superior gyrus (T) and rarely (immediate post-ictal phase) to the temporo-parieto-occipital junction in the posterior part of the SZ (D, external contacts).

These results delineated an irritative zone consisting of two partially overlapping networks: an anterior medio-lateral network and a neocortical lateral and basal network. These two networks were not independent since the latter was activated when the former was depressed in the post-ictal phase.

Identification of the epileptogenic zone relied on the recordings of five spontaneous seizures and six seizures triggered by electrical stimulation. Two types of seizure could be identified: (i) the first type (Fig. 5) corresponding to the usual clinical seizures involved an anterior medio-lateral temporal network. The seizure onset was characterized by a high amplitude sharp complex involving right amygdala and hippocampus partially followed by an initial overlapping fast low voltage discharge (FLVD) localized in the same structures (hippocampus: B, internal leads, C, internal leads and amygdala: A, internal leads). This discharge kindled less than 2 s later the anterior part of the superior, middle and inferior temporal gyri (A, B, C and TB, external leads; Fig. 5). The discharge progressively slowed and took the form of continuous rhythmic spikes (12 c/s) propagating secondarily to posterior schizencephaly (anterior bank: D) and inconstantly to right posterior superior temporal gyrus (TP). The entire seizures lasted on average 60 s. Clinically, the patient initially warned and reported a constant ascending warm sensation, and sometimes a complex visual scene of a previous seizure corresponding to a dreamy state. Swallowing automatisms and gestural elementary automatisms of the left or right hand (abdominal rubbing) and secondary partial loss of contact were inconstant. This first type of spontaneous seizure was reproduced by shock and train electrical stimulation of the amygdala (A, internal leads), hippocampus (B, internal leads) and entorhinal cortex (TB, internal leads). Stimulation of A and B elicited a local rapid fast low voltage discharge secondarily slowing (12 c/s) and propagating to lateral temporal cortex (external contacts of A, B, C and TB) and posterior schizencephaly (D). Stimulation of TB first elicited an increase of inter-ictal spikes in hippocampus and amygdala followed by a typical spontaneous seizure (ii) the second type of seizure (Fig. 6, lateral, basal) was observed once and was not reproduced by electrical stimulation. Electrically, the initial fast

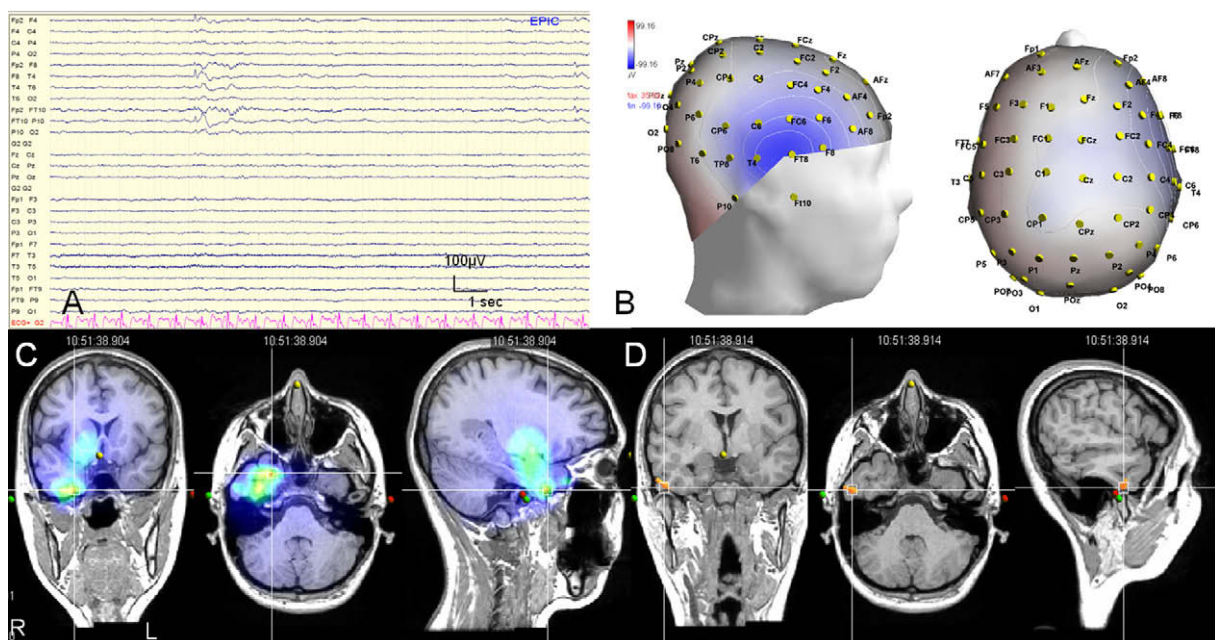


Fig. 2. (A) Bipolar 10/20 longitudinal montage showing an anterior temporal inter-ictal spike-wave; (B) 10/10 international system amplitude cartography of the inter-ictal event; and (C and D) source localisation (C: MUSIC; D: single dipole) showing the temporo-basal and temporo-polar modelled generator of the inter-ictal event.

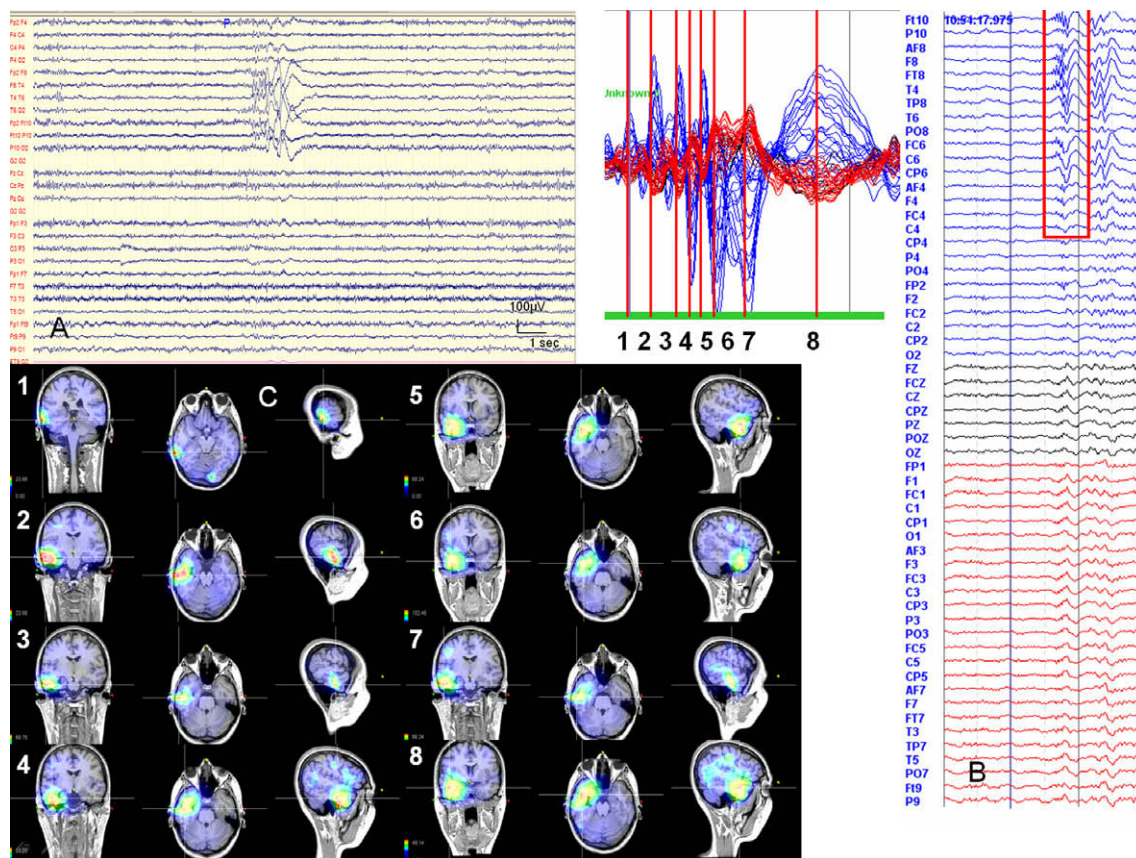


Fig. 3. (A) Bipolar 10/20 longitudinal montage and (B) 10/10 referential montage, showing an anterior and middle temporal inter-ictal polyspike-wave; (C) sLORETA distributed sources analysis consecutively localised modelled generators in the middle and lateral part of the schizencephaly (steps 1 and 2), then in the middle and anterior basal temporal area (steps 3–8).

low voltage discharge spared the medial temporal structures and involved not only the anterior part of the SZ (C, middle contacts) but also the superior, middle and inferior temporal gyri (T, A, B, C and TB, external contacts) and the anterior bank of the posterior SZ (D, Fig. 6). Secondly, it spread to the medial temporal structures (entorhinal cortex: TB, internal contacts and posterior hippocampus: C, internal contacts). Clinically, the patient did not spontaneously warn but when asked, reported a visual blurring in all visual fields. There was no clinically objective sign.

4.1. FDG-PET study

FDG-PET showed a heterogeneous pattern of the malformation: the anterior part belonged to a larger anterior temporal region with decreased metabolism (Fig. 7B) involving the temporal pole, basal and limbic structures and matching the anterior temporal sources of the inter-ictal spikes (Fig. 7A). In contrast, the metabolism of the posterior part of the SZ appeared to be increased compared with the contra-lateral temporal cortex (Fig. 7D). This functional zone lay just behind the sources of the inter-ictal polyspike-waves (Fig. 7C).

4.2. Surgery

A right anterior temporal lobectomy involving the temporal pole, amygdala, hippocampus, the anterior and middle part of the SZ and polymicrogyric cortex and sparing the posterior part of the SZ was performed in January 2008. Post-operative EEG monitoring did not show any paroxysm and the patient remains seizure-free after a follow-up of 14 months.

5. Discussion

We present the anatomo-electro-clinical correlations of a patient with medically intractable partial epilepsy associated with temporal closed lip SZ, regional PMG and malformation of the hippocampal formation.

5.1. Comparison of non-invasive source localisation technique and SEEG in delineating the irritative zone

In contrast to focal cortical dysplasia (FCD) (Chassoux et al., 2000), inter-ictal paroxysmal activity within the irritative zone was discontinuous, as described previously in PMG (Chassoux et al., 2008). SEEG identified two irritative networks: an anterior medial-lateral network and a lateral-basal temporal network corresponding to the anterior part of the SZ/PMG complex. Source localization identified two possible generators corresponding to the two types of surface inter-ictal events: one was localized in the right temporo-polar region (Fig. 2), the other one was best described as a network consisting of the sequential activation of the anterior schizencephaly, lateral and basal right temporal gyri and then the temporal pole (Fig. 3). Dipole source imaging did not identify hippocampal sources of inter-ictal spikes. This is consistent with previous studies showing that only the lateral component of medio-lateral spikes can be reliably identified and localized on high-resolution scalp-EEG (Alarcon et al., 1994; Merlet and Gotman, 1999; Gavaret et al., 2004). We therefore believe that the temporo-polar source localizations reflected the occasional basal and lateral propagation of the medial inter-ictal spikes identified on SEEG (Fig. 4). Interestingly, the lateral network iden-

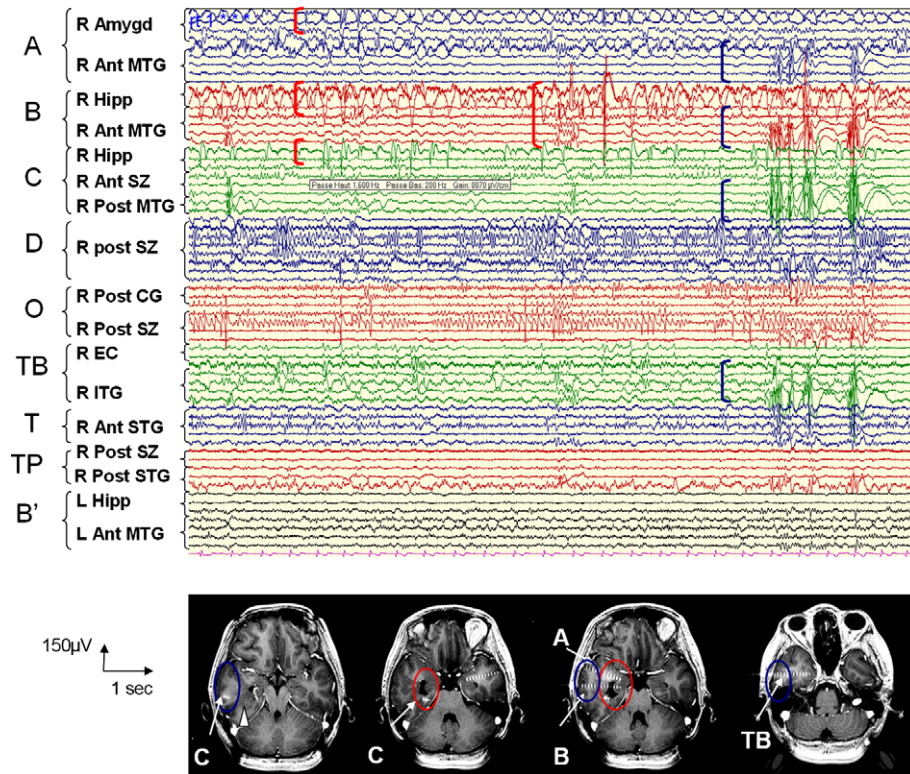


Fig. 4. SEEG recordings of inter-ictal events. Red markers correspond to frequent spike-wave activity within the right hippocampus (electrodes B and C, internal contacts) and amygdala (A, internal contacts). Blue markers correspond to independent bursts of polyspike-waves recorded in the right middle temporal gyrus (A, B and C, external contacts) and inferior temporal gyrus (TB, external contacts). The anterior and posterior schizencephaly (C, D and O), right anterior and posterior superior temporal gyrus (T and TP), right rhinal cortex (TB), and left hippocampus (B') display normal inter-ictal background activity. *Abbreviations:* R, right; Amygd, amygdala; ant, anterior; MTG, middle temporal gyrus; Hipp, hippocampus; SZ, schizencephaly; post, posterior; CG, cingulate gyrus; EC, entorhinal cortex; ITG, inferior temporal gyrus; STG, superior temporal gyrus and L, left. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

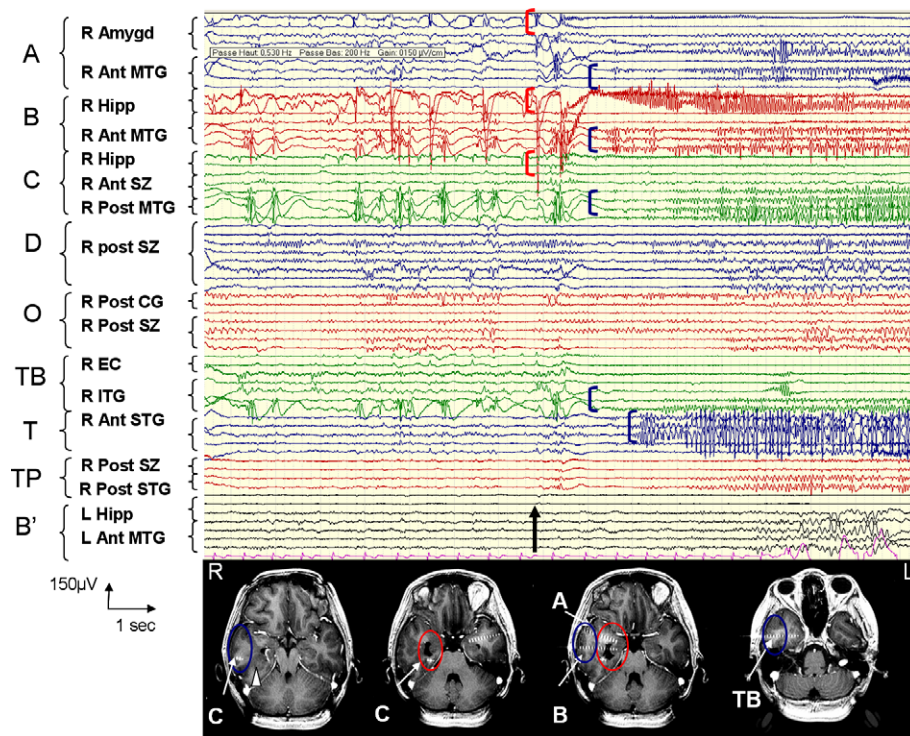


Fig. 5. SEEG recordings of a right anterior medio-lateral temporal seizure. Black arrow indicates seizure onset characterised by an initial high amplitude spike-wave and fast low voltage discharge localised in the amygdala (A, internal contacts) and hippocampus (B and C, internal contacts, red markers), and then 2 s later in the middle and inferior temporal gyri (A, B, C and TB, external contacts), followed by a slower rhythmic alpha spike discharge in the superior temporal gyrus (T). *Abbreviations:* R, right; Amygd, amygdala; ant, anterior; MTG, middle temporal gyrus; Hipp, hippocampus; SZ, schizencephaly; post, posterior; CG, cingulate gyrus; EC, entorhinal cortex; ITG, inferior temporal gyrus; STG, superior temporal gyrus and L, left. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

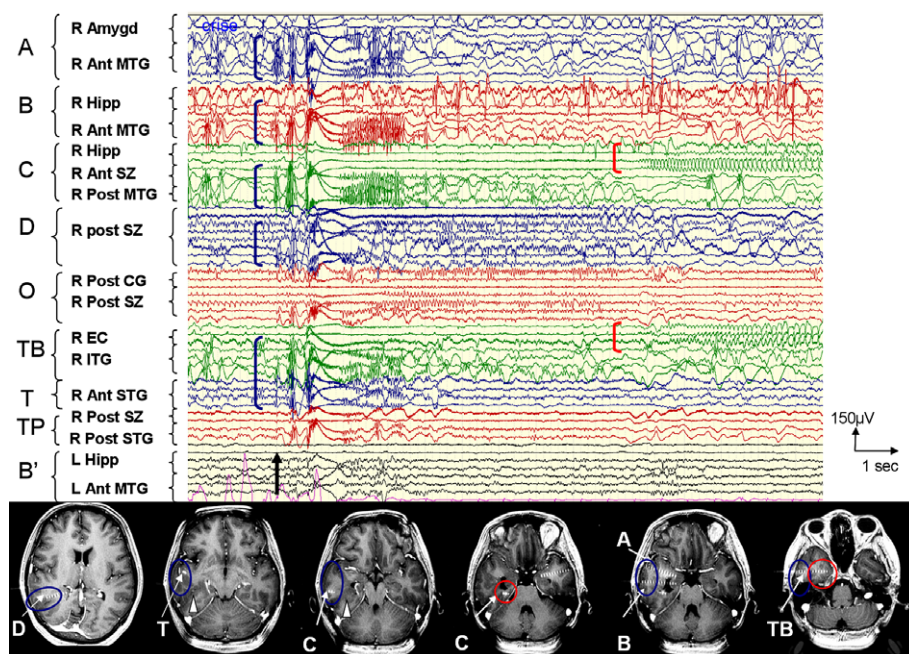


Fig. 6. SEEG recordings of a right lateral, basal and posterior temporal seizure. Black arrow indicates the seizure onset characterized by a polyspike immediately followed by a fast low voltage discharge localized (blue markers) in the middle temporal gyrus (A, B and C, external leads), inferior temporal gyrus (TB, external leads) and superior temporal gyrus (T and TP) extending to the posterior schizencephaly (D). Secondary propagation to right hippocampus and ento-rhinal cortex (C and TB, internal leads) is indicated by red markers. *Abbreviations:* R, right; Amygd, amygdala; ant, anterior; MTG, middle temporal gyrus; Hipp, hippocampus; SZ, schizencephaly; post, posterior; CG, cingulate gyrus; EC, entorhinal cortex; ITG, inferior temporal gyrus; STG, superior temporal gyrus and L, left. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

tified by dipole source imaging was consistent with the lateral irritative zone identified by SEEG involving the anterior schizencephaly and the middle and inferior temporal gyri (Fig. 4) and sparing the posterior part of the SZ. The heterogeneous pattern of inter-ictal activity of the SZ correlated well with its heterogeneous metabolic

pattern (Fig. 7): the anterior epileptic part displayed decreased metabolism while the most posterior part displayed normal if not increased metabolism compared with the normal cortex. Patterns of increased metabolism have been described in distributed areas extending beyond the epileptogenic network of cryptogenic

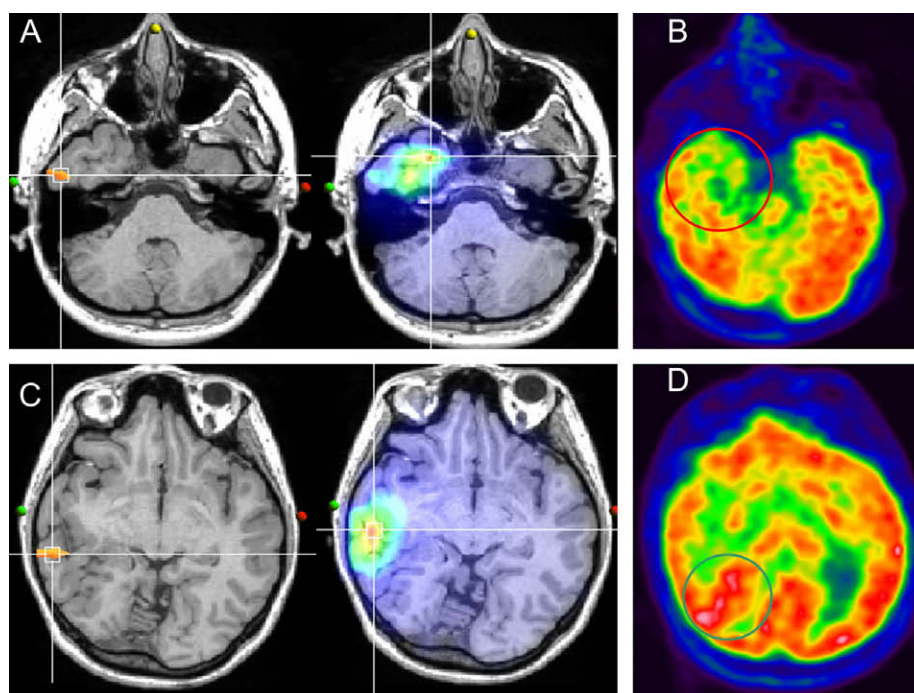


Fig. 7. Right temporal pole modelled generator of the inter-ictal spike-wave identified by single dipole and MUSIC analysis (A), with a corresponding inter-ictal PET scan metabolic decrease in the right anterior and medial temporal lobe (B). (C) Right middle temporal source localization of the inter-ictal polyspike-wave localised by single dipole and MUSIC analysis; and (D) projecting ahead of the posterior cleft which displays an apparent metabolic increase.

temporal lobe epilepsies (Franceschi et al., 1995) and were hypothesised to represent either “a pathophysiological state predisposing to epileptic discharge diffusion”, or “a network of inhibitory circuits activated to prevent the diffusion of the epileptic discharge”. In our case, the apparent hypermetabolism in the ectopic dysgenetic cortex extending along the cleft was most probably explained by the extension of normal grey matter metabolic activity inside the white matter (Van Bogaert et al., 1998). Inter-ictal metabolic and electrophysiological data support the view that a SZ/PMG complex can be functionally heterogeneous and simultaneously have pathological activity in one part (anterior part in this case) and normal inter-ictal electrical and metabolic activities in the other (Leblanc et al., 1991; Spreer et al., 2000). Source localisation was very valuable because it combined both structural and functional data in the anatomical space of the patient (Burneo et al., 2004a,b) and helped define the targets for depth electrode implantation. In such cases of very large structural lesions, this method provides accurate and regional data, which in combination with SEEG overcome the classical limitations of intra-cranial EEG related to partial sampling of cortical activity.

5.2. Identification of the epileptogenic zone by SEEG

SEEG identified two epileptogenic networks. The first one explained most of the clinical seizures. Ictal symptoms and signs recorded indicated right temporal lobe involvement, homo-lateral to the malformation. The initial viscerosensitive symptom associated with déjà-vu and reminiscence supports an initial medial temporal involvement (Williamson et al., 1993; Maillard et al., 2004; Vignal et al., 2007) confirmed by the ictal SEEG recordings. According to the classical definition of the epileptogenic zone (Talairach et al., 1974), ictal electrical patterns of triggered and first type spontaneous seizures support an epileptogenic zone involving primarily medial and also lateral anterior temporal cortex corresponding to the electro-clinical medio-lateral sub-type (Bartolomei et al., 2001; Maillard et al., 2004). The lateral part of this epileptogenic zone included the anterior part of the SZ. The second network identified by the SEEG was a lateral neocortical network, including the anterior and part of the posterior SZ (Figs. 1 and 6) and spreading secondarily to the medial temporal structures. This network could not be triggered by electrical stimulation. So only the anterior part of the SZ belongs to the anterior medio-lateral primary epileptogenic zone involving also right temporal limbic structures and amygdala.

In contrast to focal cortical dysplasia, whose electrical stimulation easily reproduces spontaneous seizures (Chassoux et al., 2000), focal electrical stimulations of the polymicrogyric clefts did not elicit seizures. It may be that in SZ, a larger volume of cortex has to be recruited to generate seizures but this hypothesis needs further validation on other similar cases. This different electrophysiological property could be related to the distinct developmental origin of SZ and of type 2 FCD, the former being related to abnormal cortical reorganisation, the latter being characterised by the presence of dysmorphic neurons and balloon cells related to abnormal proliferation (Barkovich et al., 2005).

For many authors, the success of surgery depends on the completeness of resection of the visible malformation (Edwards et al., 2000; Hamiwka et al., 2005). Others have emphasized that it holds true, provided that the malformation is congruent with the primary epileptogenic zone identified by electroclinical data and SEEG (Sisodiya, 2000; McGonigal et al., 2007). In our particular case, given its large extension from the temporal pole to the temporo-parieto-occipital junction, the malformation could not be completely removed without causing obvious cognitive alterations. Clinical findings, EEG and SEEG completed by dipole source imaging consistently delineated the irritative and epileptogenic zone in

the anterior right temporal lobe and prompted an anterior lobectomy sparing the posterior part of the SZ. The favourable current outcome (seizure-free period of 14 months) supports the view that resection of SZ/PMG does not have to encompass the whole malformation to obtain complete removal of the epileptogenic zone. This needs further validation in other similar cases.

In conclusion, this is the first study to combine source localisation and intraslesional recordings to confirm the functional and epileptic heterogeneity of SZ. In this case, the irritative zone defined by high-resolution EEG was consistent with SEEG and was useful to define targets for SEEG. In such cases of very large structural lesions, it could provide accurate and regional data, complementary from SEEG since it overcomes the classical limitations of intra-cranial EEG related to partial sampling of cortical activity. On the other hand, SEEG can record epileptic activity from limited deep generators like hippocampus, not detectable by scalp-EEG whatever the number of electrodes (Gavaret et al., 2004).

Disclosure information

The authors do not have any financial interest to disclose.

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