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Intracranial video-EEG and surgery for focal atonic seizures

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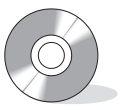
ABSTRACT – Atonic seizures are epileptic attacks characterised by a sudden loss or diminution of muscle tone. Structures corresponding to inhibitory cortical areas, such as the primary negative motor area or the supplementary negative motor area, could be responsible. We present findings observed in a patient with atonic seizures due to focal epilepsy, who underwent intracranial video-EEG monitoring and epilepsy surgery, and discuss possible underlying mechanisms. [*Published with video sequences*]

Key words: focal atonic seizures, epilepsy surgery, intracranial video-EEG, high-field MRI, neuropathology

Atonic seizures are epileptic attacks characterised by a sudden loss or diminution of muscle tone, affecting areas ranging from a circumscribed segment, such as a limb or the head, to antigravity muscles of the entire body, leading to falls and trauma (epileptic drop attacks) (Proposal for revised clinical and electroencephalographic classification of epileptic seizures, 1981; Blume *et al*., 2001; Zhao *et al*., 2010). In these cases, loss of consciousness is usually of very short duration (Satow *et al*., 2002; Zhao *et al*., 2010). Atonic seizures are often seen in patients with generalised epilepsies, but in a few cases, atonic semiology has also been reported to occur in patients with focal epilepsy (Zhao *et al*., 2010; Kovac and Diehl, 2012). Focal atonic seizures are partial seizures and the ictal manifestation consists of paresis or paralysis of one or more parts of the body (Kovac and Diehl, 2012). Mechanisms causing focal atonic seizures have not yet been fully identified, but structures corresponding to inhibitory cortical areas, such as the primary negative motor area (PNMA), located in the inferior frontal gyrus, or the supplementary negative motor area (SNMA), in front of the supplementary sensorimotor area, could be responsible, at least in part, for the phenomenon. Stimulation of these areas produces predominantly contralateral atonia of limb muscles (Lüders *et al*., 1995; Zhao *et al*., 2010; Lüders, 2008).



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We present findings observed in a patient with atonic seizures due to focal epilepsy, who underwent intracranial video-EEG (VEEG) monitoring and epilepsy surgery, and report results of the neuropathological analysis of the tissue excised.

Case Study

A 25-year-old, right-handed woman with a history of non-complicated febrile seizures suffered with epilepsy for over 13 years. The patient reported seizure onset at the age of 12, comprising epigastric discomfort, loss of consciousness, manual automatisms, sphincter relaxation, and postictal confusion. Ictal semiology changed over time. Current symptoms include unresponsiveness, head drops with slight rotation to the right, sudden atonia of axial muscles, and frequent falls. Family history was unremarkable. Pregnancy and birth were normal.

At the time of admission, repeated EEGs showed right temporal spikes. Extensive right temporo-parieto-occipital hypointensity on T2* (gradient echo) image was observed by 3.0T MRI and confirmed to be calcified based on CT and interpreted as a large calcified dysplastic lesion (*figure 1*). Coeliac disease was ruled out.

The patient was receiving the following medications: zonisamide at 250 mg/d, levetiracetam at 3,000 mg/d, oxcarbazepine at 2,100 mg/d, and clonazepam at 3 mg/d. She had previously been prescribed carbamazepine, lamotrigine, topiramate, and valproate which had failed to control seizures.

During VEEG recording, 5 episodes of loss of consciousness, sudden head drops with slight deviation to the right, and upper limb atonia were recorded. Tonic elevation of both upper limbs was also observed on one occasion. More recently, right hand automatisms were recorded. Postictal features included confusion and difficulty in naming objects (see *video sequence*). In 3 of 5 seizure episodes, ictal EEG showed generalised voltage attenuation followed by brief, rhythmic activity in the right posterior temporal region. During one seizure, ictal onset was bitemporal (*figure 2*). Interictal EEG showed a right temporal focus and independent left temporal spikes.

Due to suspicion of a dysplastic lesion and because of a lack of clear correlation between the symptomatogenic area (atonic seizure) and the temporal lobe lesion, we decided to perform invasive VEEG using subdural electrodes, with the intention of better defining the epileptogenic zone. Electrodes were placed over the lesional area (temporal lobe) and the parietal lobe (suspected to be the symptomatogenic area). Two extra strips were placed on the medial frontal lobe, which is also a possible atonic seizure area. Two 32-contact subdural grids, one over the right parieto-occipital region (above the lesion) and the other over the right frontal lobe, together with 2 subdural strips containing 8 contacts (right temporal and subtemporal), were implanted. Due to difficulties in accurately localising the ictal onset site, 2 right-sided

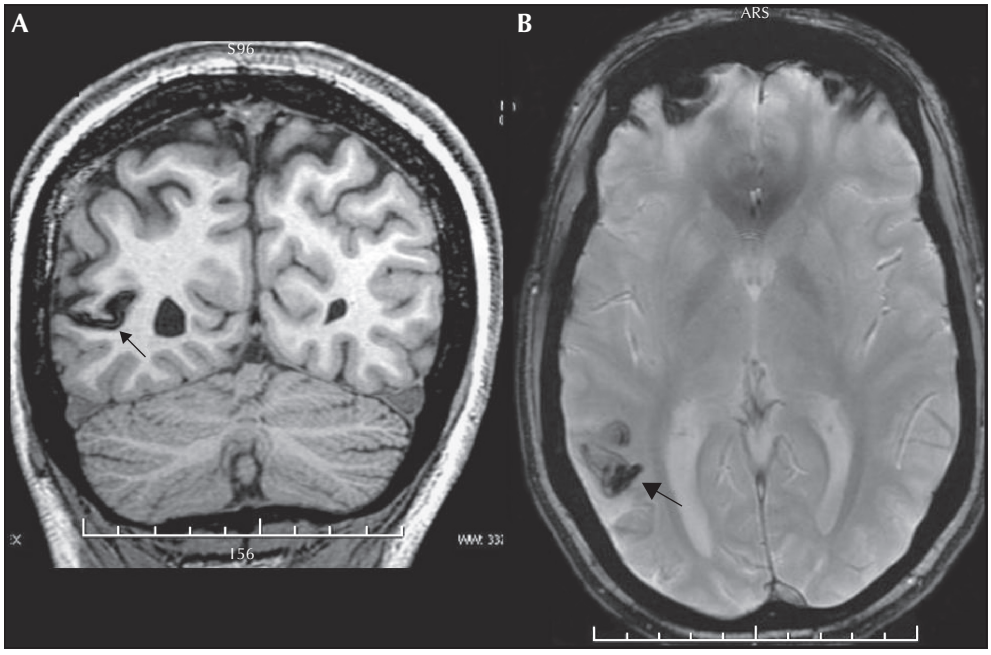

Figure 1. MRI 3.0T epilepsy protocol. A) Coronal 3D showing linear hypointensity (arrow). B) T2* right temporo-parieto-occipital hypointense image (arrow).

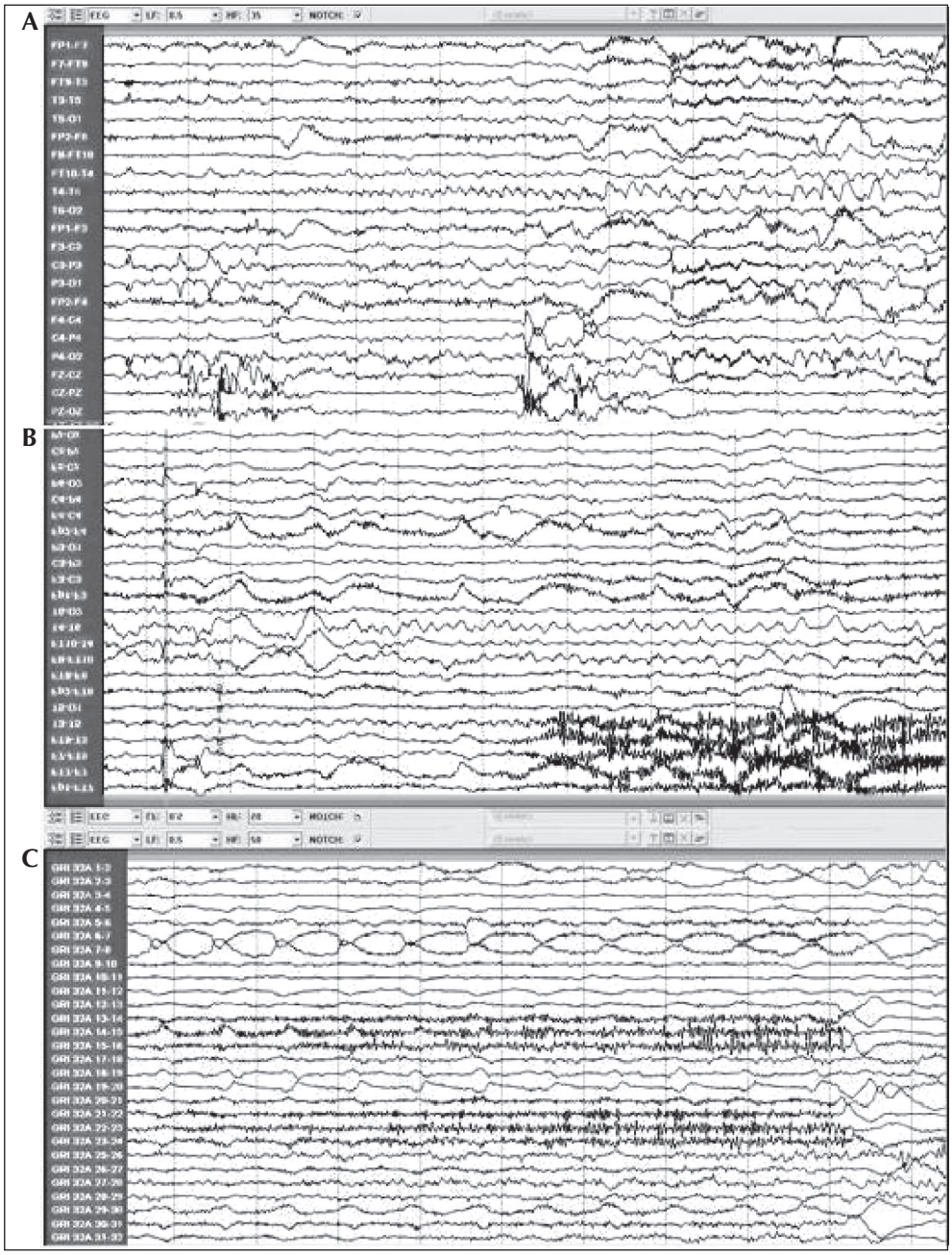

Figure 2. Double banana montage. A) Scalp EEG showing ictal activity in the right posterior temporal region (T4). B) Interictal scalp EEG showing spikes in the right temporal lobe. C) Intracranial EEG showing ictal activity in contacts 14-15 and 22-23 from the anterior centro-parietal grid (32A).

extradural implants were added (a 20-contact grid over the posterior temporal region and an 8-contact strip over the fronto-temporal region) (*figure 3*). In total, 37 seizures were recorded during intracranial VEEG, as described.

Coinciding with clinical onset, voltage attenuation was observed in all anterior centro-parietal grid contacts, followed by fast recruitment activity, which ultimately spread to the subtemporal strip. On the basis of these results, lesionectomy and tailored corticectomy were performed.

A 6-contact strip was used during intraoperative electrocorticographic (ECoG) monitoring showing temporal, parietal, and perilesional areas of high frequency epileptiform paroxysmal discharges. Before the resection was performed, spike and polyspike

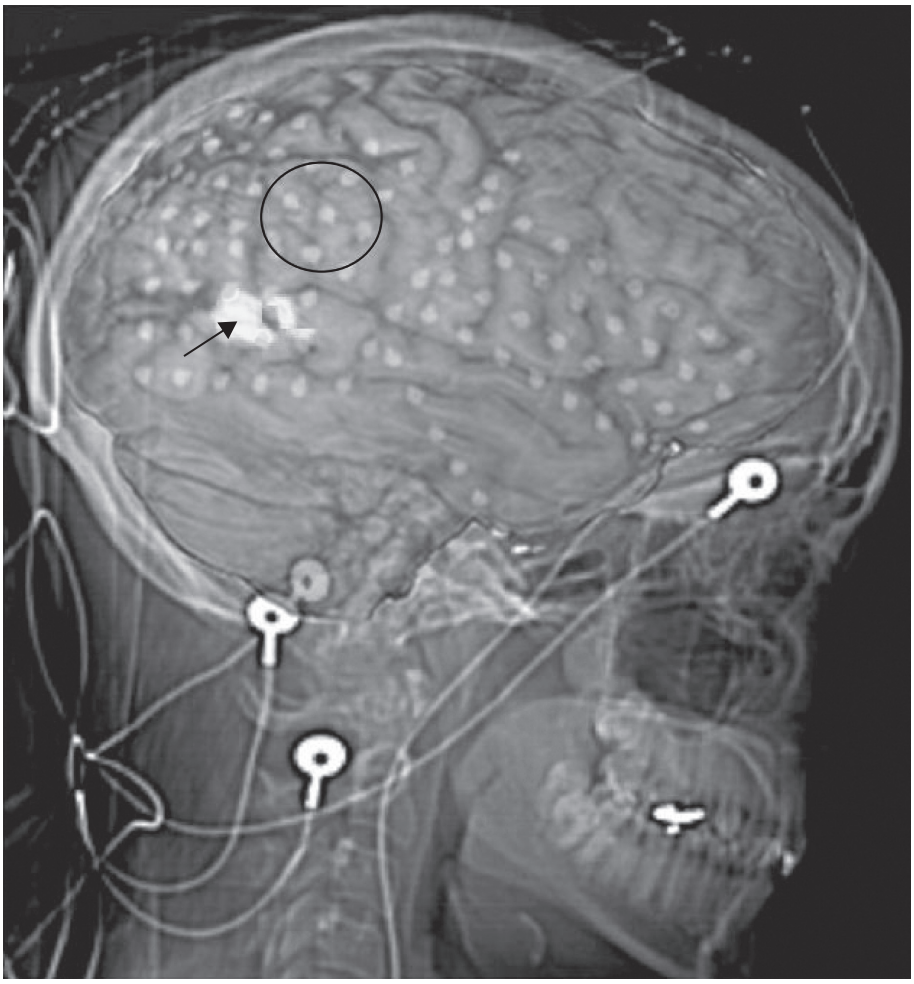

Figure 3. Reconstructed image by fusion of MRI 3.0T epilepsy protocol and skull X-ray to confirm implant placement, using MRIcro 1.40 build 1 program.
The reconstructed calcified lesion is shown in white and pointed by the black arrow. Contacts from centro-parietal grids showing ictal discharges (14-15 and 22-23) are circled.

paroxysms were recorded on parieto-temporal cortex near the lesion and spikes of high frequency were observed on parietal cortex above the lesion, coinciding with interictal discharges observed on invasive VEEG. During the intraoperative ECoG, epileptiform paroxysms were recorded in perilesional areas, especially in temporal and parietal cortex. These areas were resected. After the resection, polyspikes of high frequency outside the area of resection, on posterior temporal areas, persisted.

Histopathological analysis revealed calcified IIA dysplasia with intense reactive astrogliosis, secondary to calcium deposits (*figure 4*).

Three years after epilepsy surgery, the patient remained seizure-free and was reported to have an improved quality of life.

Discussion

Atonic seizures of partial epilepsies have previously been classified by Tassinari *et al*. as two separate types (Satow *et al*., 2002; Engel *et al*., 2008). In the first, the fall is preceded by sudden tonic posturing, whereas in the second, falls are not associated with any overt motor sign. In the case presented here, the seizures of the patient clearly corresponded to the second group, with a manifestation of ictal phenomena consisting of sudden loss of consciousness, head drops, and axial and upper limb atonia; all findings were confirmed on clinical examination during episodes.

The ictal EEG pattern showed diffuse voltage attenuation followed by rhythmic activity in the centro-parietal

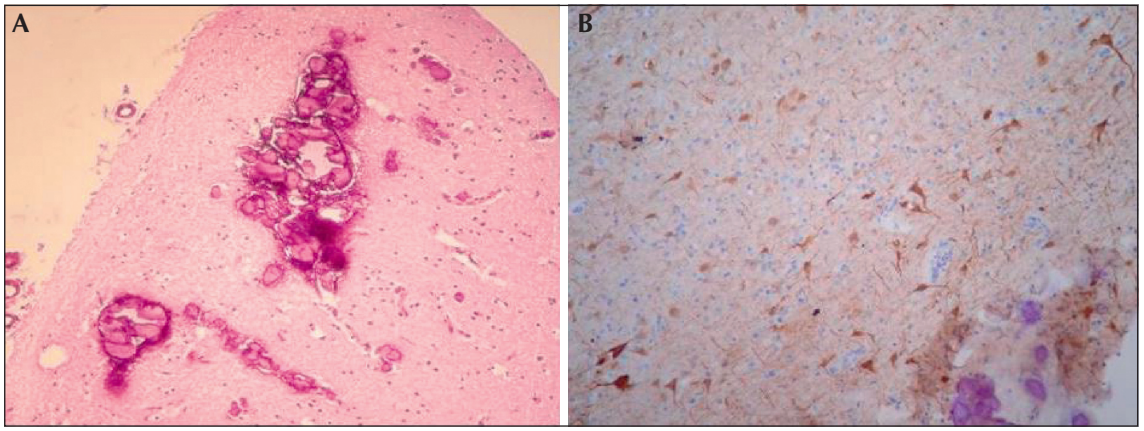

Figure 4. A) Haematoxylin and eosin staining at 20x magnification showing brain tissue with calcified cortical lesions. B) NF (neurofilament) immunohistochemical staining showing grouped, dysmorphic neurons with loss of polarity and disrupted architecture.

area, as described, similar to descriptions observed by Satow *et al*. (2002) and Zhao *et al*. (2010).

The PNMA and SNMA are suspected to be responsible for causing atonia in this kind of seizure. Electrical stimulation of these areas was shown not to initiate or maintain voluntary movements (Satow *et al*., 2002; Engel *et al*., 2008). It has been hypothesized that lesions, or focal discharges in the primary somatosensory cortex, may activate the inhibitory motor system represented by the negative motor areas (Kovac and Diehl, 2012).

Atonia is an under-recognised ictal phenomenon in focal seizures. There is a lack of consistent nomenclature which contributes to a misconception of the semiology and pathophysiology of this seizure type (Kovac and Diehl, 2012).

When focal atonic seizures are suspected, admission for VEEG monitoring and epilepsy protocol high-field MRI are recommended in order to increase diagnostic accuracy, as well as rule out possible surgical lesion-related aetiology. □

Legend for video sequence

Video sequence

The patient sits on the bed. Suddenly, she has a head drop with slight deviation to the right with upper limb atonia.

Key words for video research on www.epilepticdisorders.com

Syndrome: focal non-idiopathic (localization not specified)
Etiology: dysplasia (architectural)
Phenomenology: aura (abdominal); atonic seizure (drop attack); head deviation
Localization: multilobar including temporal

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