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Bilateral symmetric tonic posturing suggesting propagation to the supplementary motor area in a patient with precuneate cortical dysplasia

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ABSTRACT – We report a patient manifesting seizures with bilateral symmetric tonic posturing, which were markedly reduced after resection of the left precuneus. A 16-year-old man had sudden onset, complex partial seizures with bilateral symmetric tonic posturing since the age of eight years. Magnetic resonance fluid-attenuated inversion-recovery imaging revealed a hyperintense lesion in left precuneus. In almost all focal seizures recorded during an invasive EEG evaluation, ictal onset was detected from the inferomesial aspect of the lesion, but fast paroxysmal discharges from the ipsilateral supplementary motor area (SMA) were observed just before the clinical onset. After surgical excision of the EEG onset zone, including the lesion, seizure frequency was markedly (> 95%) reduced. By the 20th month after surgery, there were only brief nocturnal seizures involving slight elevation of both shoulders and slight abduction of both arms, with preservation of consciousness occurring once every few days. Invasive EEG findings and surgical outcome suggested that the epileptic activity originating from the epileptogenic zone may have propagated to the symptomatogenic zone including mainly the ipsilateral SMA. In summary, we report an interesting case of bilateral symmetric tonic posturing suggesting propagation to the SMA. MRI and invasive EEG confirmed the epileptogenic focus as a precuneate cortical dysplasia lesion. *[Published with video sequences]*

Key words: bilateral symmetric tonic posturing, cortical dysplasia, precuneus, supplementary motor area, focal seizures, symptomatogenic zone, parietal lobe seizures

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Surgery for parietal lobe epilepsy is much less common than temporal or frontal procedures, being as low as 8% of all epilepsy surgeries (Salanova *et al.* 1995). In a clinical series of 82 patients with parietal lobe epilepsy treated surgically, 28% exhibited tonic posturing and 61% had epileptogenic zones involving the superior parietal lobe (Salanova *et al.* 1995). However, this article did not indicate whether cases with precuneate lesions were involved. The medial parietal lobe traditionally receives little attention because of the rarity of lesions (Cavanna *et al.* 2006). Moreover, only a few reports have documented seizures in patients with precuneate lesion. Ikeda *et al.* (2002) reported a case of left precuneate cortical dysplasia presenting as supplementary motor area (SMA) seizures, and subdural investigation suggested the propagation of epileptic activity from the lesion to the SMA. Although SMA seizures are characterized by sudden and brief tonic posturing of one or more extremities (Morris *et al.* 1988), tonic posturing is described in the literature as asymmetric, classically as a fencing posture or M2e (Ajmone-Marsan *et al.* 1957), or not in detail.

We present a rare case of a patient with complex partial seizures manifesting with bilateral symmetric posturing, while electrophysiological findings and neuroimaging localized the epileptogenic zone to the left precuneate lesion.

Case report

The present case was a 16-year-old man with a history of medically refractory seizures since the age of eight years. His habitual seizures were characterized by symmetrical elevation of both shoulders with flexion of neck and symmetrical abduction of the upper extremities. The seizures began abruptly without warning and were often followed by bilateral symmetrical tonic posturing of the four extremities, particularly the upper extremities, with impaired consciousness *(see video sequence 1)*. Magnetic resonance (MR) fluid-attenuated inversion-recovery imaging (FLAIR) revealed a hyperintense lesion at the left precuneus *(figure 1A, B* and *C)*. During long-term EEG and closed circuit television (EEG/CCTV) monitoring with scalp electrodes, interictal epileptiform discharges were observed at Cz. In the recorded seizures, ictal discharges (ID) were prominent at Cz. Interictal cerebral blood flow was examined by single photon emission tomography (SPECT) with ^{123}I-labeled N-isopropyl-p-iodoamphetamine (^{123}I-IMP) and the central benzodiazepine receptor was analyzed with ^{123}I-iomazenile (IMZ) SPECT. The two examinations identified an area of hypoperfusion and a low IMZ binding area, respectively, in the posterior region of the left parietal lobe including the precuneus lesion. Although interictal technetium-99m-ethyl cysteinate dimer (^{99m}Tc-ECD) SPECT also showed hypoperfusion around the lesion, ictal ^{99m}Tc-ECD SPECT showed hyperperfusion *(figure 1D* and *E)*. The Wada test showed that the eloquent area of language and memory was located in the left hemisphere. To identify a more detailed epileptic focus and for the cortical mapping, subdural grid electrodes (center-to-center inter-electrode distance of 1 cm) were implanted over the lesion (6 × 3) and the frontoparietal convexity (8×4). Additionally, two strip electrodes were placed in the interhemispheric fissure (6×1) so as to cover the supplementary motor area (SMA), because tonic posturing suggests that the SMA may be involved in the patient's seizures *(figure 2A* and *B)*. The result of cortical mapping revealed that interhemispheric electrodes (LHS3 and 4, and LHI2 and 3) covered the SMA region and proximal electrodes of the interhemispheric grid (A4 and 5, B5, and LHS5 and 6) were placed at the motor area of the foot *(figure 2C)*. Interictal epileptiform discharges were frequently observed, with a wide involvement of the SMA, left prefrontal area, and left parietal convexity, as well as the inferomesial portion of the lesion.

More than 100 seizures were recorded, including simple partial seizures (SPS) characterized by slight neck flexion with elevation of the shoulders and abduction of both arms, and complex partial seizures (CPS) manifesting as neck flexion followed by bilateral symmetric tonic posturing of all four extremities, but predominantly the upper extremities. In SPS, mono- or biphasic waves followed by low amplitude and fast spike activity started at the inferomesial aspect of the lesion (A2) less than one second before clinical onset. Ictal activity then spread to the prefrontal area (F2 and G2) and SMA (LHS3 and RHS3) *(figure 2D)*. Low amplitude and fast spike activity at A2 continued for about 7-10 s after the seizure had ended *(figure 2D)*. Similarly, in CPS, the epileptic discharge appeared at A2 just before clinical onset *(figure 3A)*. An enlarged EEG recording revealed that epileptic discharge at the SMA (LHS3) started soon after EEG onset at A2 electrode *(figure 3B)*. However, the activity not only spread to LHS3 and RHS3, but also involved the frontoparietal convexity within less than 1 second, before onset of the symmetric tonic posturing *(figure 3A)*. Further examinations were performed including cortical mapping and recording of somatosensory-evoked potentials for confirmation of sensory and motor areas.

These invasive evaluations suggested that the paroxysmal epileptic discharges probably propagated to the frontal area, particularly to the mesial frontal region containing the SMA, and its evolution could reflect the habitual seizures. We speculated that the epileptogenic zone was located mainly in the left precuneus. We performed resection of the left precuneus, including the lesion, taking the functional mapping into account *(figure 1F, G* and *H)*. Histopathological study of the surgical specimen revealed cortical dysplasia and balloon cells, and therefore Taylor-type focal cortical dysplasia (TFCD) was diagnosed. After surgery, a transient motor deficit appeared in the right foot for one month. Although a slight sensory disturbance

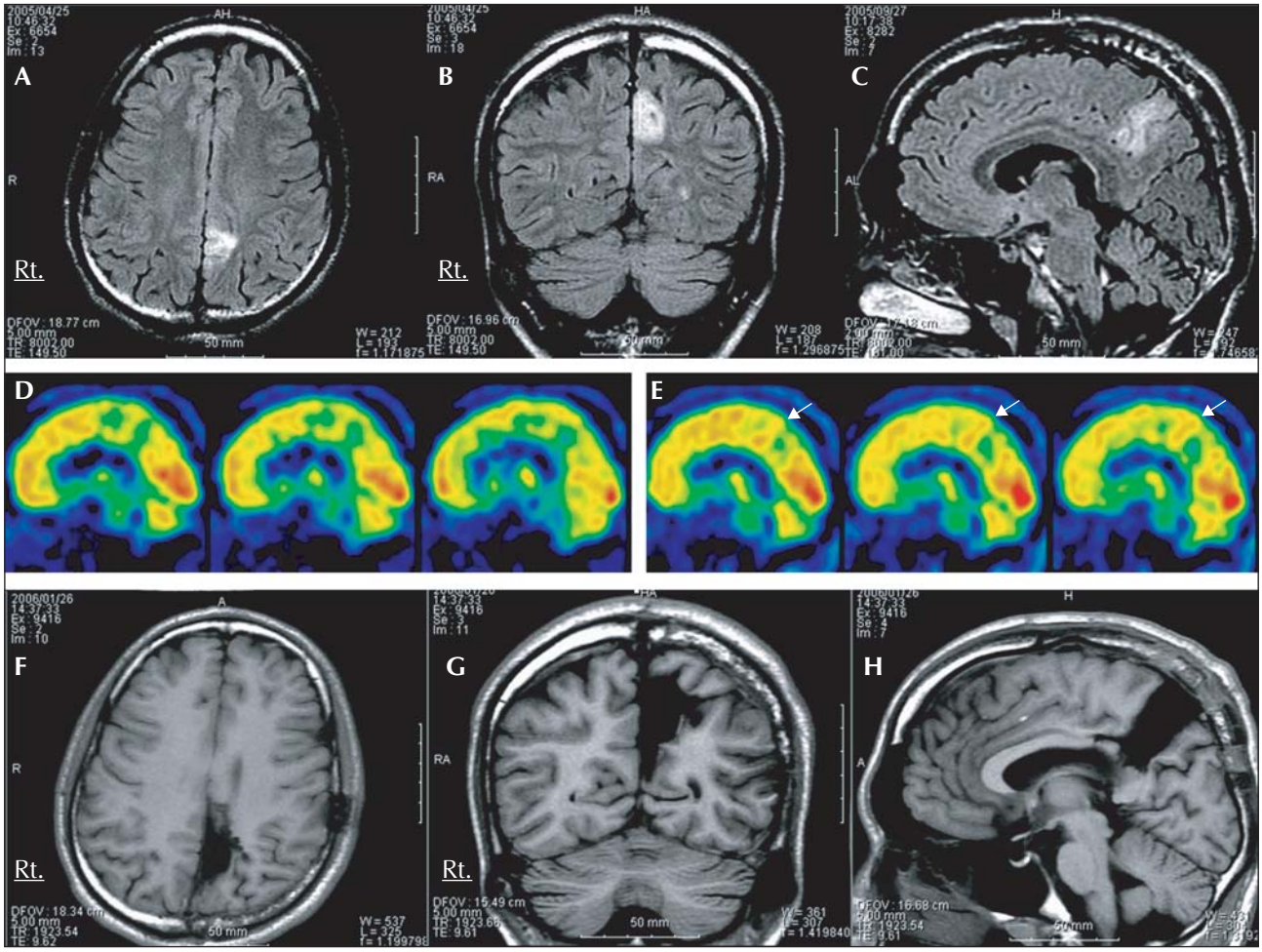

Figure 1. A), **B**), and **C**) Preoperative magnetic resonance fluid-attenuated inversion-recovery images revealing a lesion with high intensity signal in the left precuneus; axial, coronal, and sagittal, respectively. **D**) Consecutive sagittal images of interictal technetium-99m-ethyl cysteinate dimer (^{99m}Tc-ECD) SPECT showing an area of hypoperfusion around the precuneate lesion. **E**) Consecutive sagittal images of ictal ^{99m}Tc-ECD SPECT demonstrating increase of cerebral blood flow around the precuneate lesion, especially the anterior portion, during seizure (arrows). **F**), **G**), and **H**) Postoperative magnetic resonance T1-weighted images demonstrating the left precuneus gyrectomy including the cortical dysplastic lesion: axial, coronal, and sagittal, respectively.

(8/10) of the right distal foot, predominantly in the sole, persisted, higher-order cognitive function was normal. The patient was followed while on medication. Over the follow-up period of 20 months following surgery, only nocturnal SPS were reported, and these occurred once every few days.

Discussion

The postero-medial parietal cortex, including the precuneus, is traditionally under-studied because it is buried in the interhemispheric fissure and encased by the sagittal sinus and bridging veins (Cavanna *et al.* 2006). However, modern neuroimaging technology has made it possible to explore the morphology and function of this area and a central role of the precuneus has been speculated, including self-centered mental imagery strategies and successful episodic memory retrieval (Cavanna *et al.* 2006). In the present case, the patient's habitual seizures were reduced markedly (> 95%) after excision of the precuneus. Histopathological examination led to a diagnosis of TFCD. The characteristics of TFCD have been reported as follows: 1) balloon cells associated with cortical laminar disorganization; 2) high seizure frequency compared to other types of cortical dysplasia; 3) hyperintense lesion on MRI T2-weighted image is commonly localized; 4) the epileptogenic zone is mainly extratemporal (Tassi *et al.* 2002). Because nocturnal seizures remained after surgery, the epileptogenic zone might not have been restricted entirely to the precuneus. However, the remarkable decrease in seizure frequency suggests that the major epileptogenic zone was present in the resected precuneus. Because there

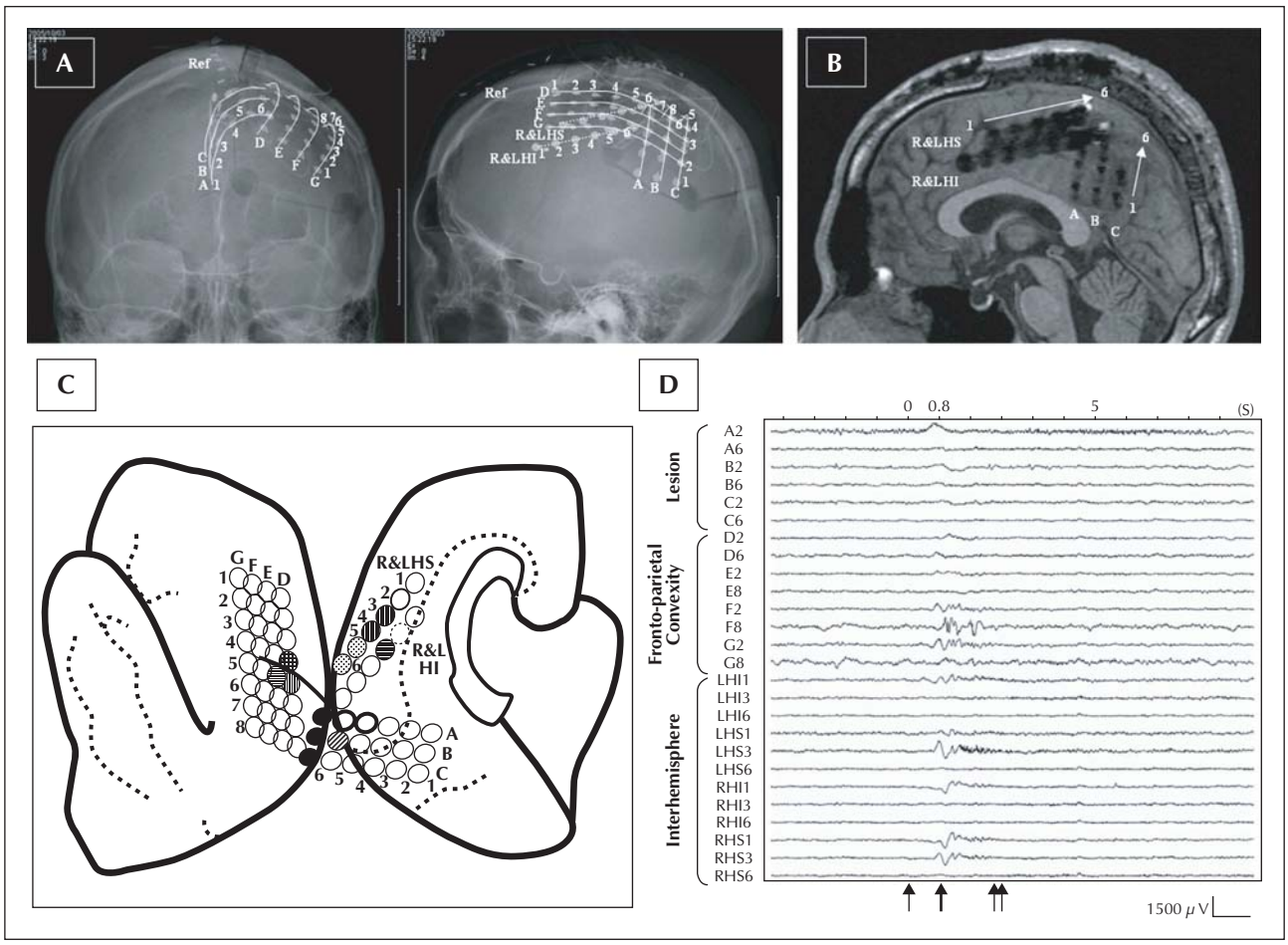

Figure 2. A) Skull X-rays demonstrating montage of subdural electrodes implanted in the prefrontal convexity, parietal convexity, and interhemispheric fissure (Ref: reference, R: right, L: left, HS: strip implanted in the superior hemispheric fissure, HI: strip implanted in the inferior hemispheric fissure, 1 to 8: number assigned to each electrode, A to G: letter assigned to each row of grid electrode).
B) Sagittal view of T1-weighted image demonstrating subdural electrodes implanted in the interhemispheric fissure. A grid consisting of 18 electrodes, each of which was named A, B and C, and covered the precuneate lesion as shown in *figure 1C*.
C) Schematic depiction of the intracranial electrodes and the results of cortical mapping. Closed circle; sensory area of right foot, bold open circle; motor area of right proximal foot, broken open circle; motor area of right shoulder (abduction), circle with bold horizontal lines; motor area of right shoulder and upper extremity (abduction), circle with fine slanted lines; motor area of right ankle joint, circle with fine vertical lines; sensory area of right neck and shoulder, circle with fine horizontal lines; sensory area of right upper extremity and shoulder, circle with crossed lines; motor area of right third and fifth fingers, dotted circle; motor area of right digits of foot (flexion), circle with bold vertical lines; motor area of feet (extension).
D) Ictal EEG demonstrating with reference montage (reference electrode is one of the subcutaneous strip electrodes shown in *figure 2A*). Propagation of the intracranial epileptiform discharge during a simple partial seizure manifesting as slight elevation of shoulders with symmetric abduction of both upper extremities. EEG onset (thick arrow) is marked by the appearance of mono-phasic waves, followed by low voltage fast activity at A2 starting approximately 0.8 s before clinical onset (fine arrow). Left SMA (LHS3) and prefrontal region (F2 and G2) are first involved just before clinical onset and followed by low-voltage fast activity. Double arrows demonstrate the point at which the seizure ended.

are limitations to the extent of electrode placement, the picture of seizure discharges obtained from the intracrnaial EEG using electrodes placed on the brain surface, does not necessarily provide an accurate picture of the actual spread of the epileptic activity. However, the remarkable seizure reduction following resection of the precuneus may explain the ictal spread from the precuneus to the SMA. The residual postoperative seizures may be due to incomplete removal of the epileptogenic zone.

The clinical symptomatology of supplementary motor seizure is characterized by sudden and brief tonic posturing of one or more extremities, vocalization, and initially preserved consciousness (Morris *et al.* 1988). Despite the implantation of strips, with electrodes adhered on both sides, into the interhemispheric fissure, we were not able to confirm precisely to what extent the right SMA was related to the seizures. However, in almost all of the recorded seizures, clear EEG onset consisting of mono- or

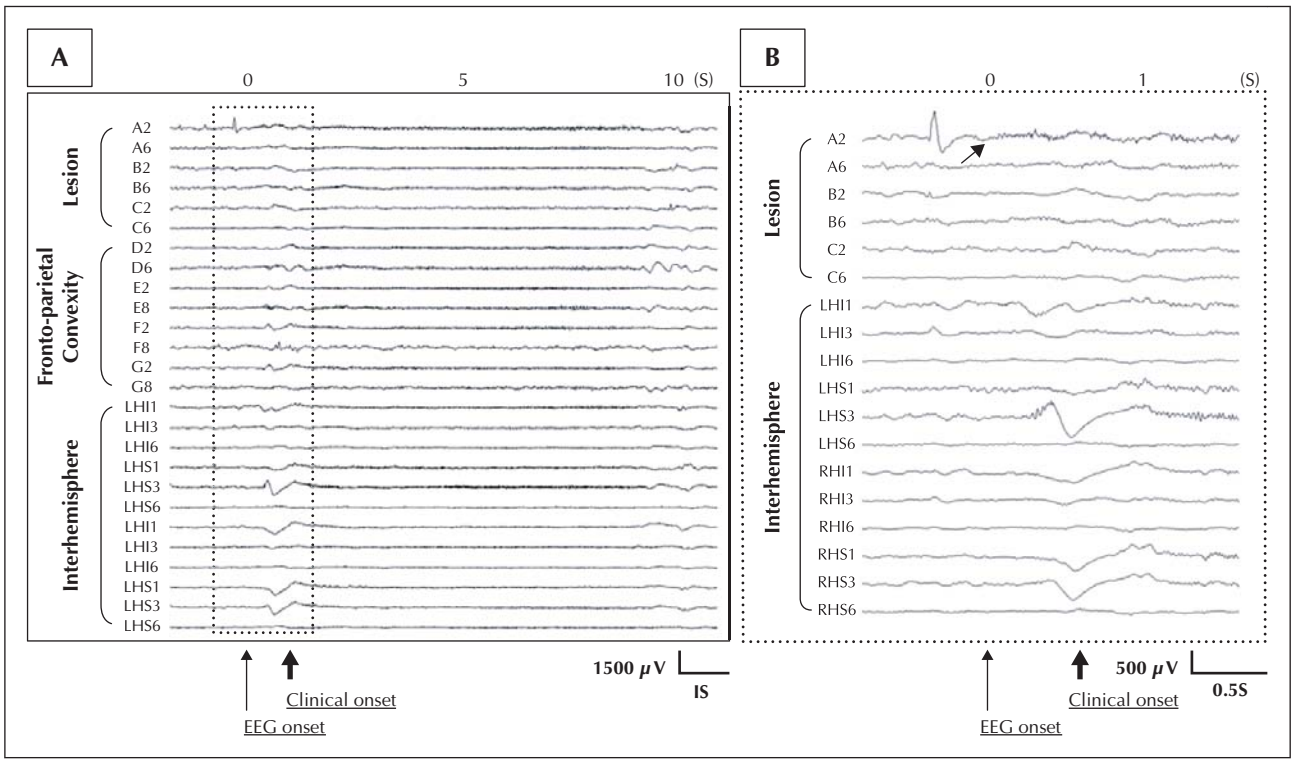

Figure 3. A) Ictal intracranial EEG, demonstrated with reference montage (reference electrode is one of the subcutaneous strip electrodes shown in *figure 2A*), during an habitual complex partial seizure. Paroxysmal discharges propagated not only to left SMA and prefrontal lobe, but also to the parietal convexity. Spontaneous EEG onset (fine long arrow), just before clinical onset (bold arrow), was defined as the appearance of low voltage fast activity at the electrodes covering the inferior lesion (A2). **B**) An enlarged EEG recording from selected electrodes for the period marked by the dotted line enclosed area in *figure 3A*. EEG onset (fine long arrow) is observed approximately 0.6 s before the EEG change at the SMA electrode (LHS3). The abbreviated letters and numbers shown in this figure are in accord with those presented in *figure 2*.

biphasic waves followed by fast activity was recorded from the electrode (A2) placed on the precuneate lesion, and was followed one second later by EEG changes at the electrodes for the left SMA and left prefrontal convexity, just before clinical onset. Considering both the clinical symptoms and electrophysiological findings, propagation of epileptic activity to the SMA is suggested. Although SMA seizures are classified as a frontal lobe epilepsy by ILAE (Commission on Classification and Terminology of the International League Against Epilepsy 1989), its motor symptoms are unique, but not quite definitive (Ohara *et al.* 2004). Our patient manifested symmetric tonic posturing: we could find no reports documenting the difference between asymmetric and symmetric seizures.

Although SMA seizures can be defined as seizures originating from or secondarily involving the SMA, propagation of epileptic discharges from the precuneus to the SMA has been rarely documented (Ikeda *et al.* 2002). Extensive connections between the precuneus and the SMA have been demonstrated. Furthermore, the corticocortical projections from the precuneus to the lateral parietal areas and prefrontal cortex have been suggested to play a pivotal role in the hand-eye coordination (Ferraina *et al.* 1997). Using electrical stimulation studies, Lim *et al.* (1994) demonstrated that SMA-type motor positive responses were elicited not only from the SMA, but also from the paracentral lobule, cingulate gyrus and precuneus. In the present case, the first paroxysmal discharges originating from the lesion (A2 electrode) and involving the SMA (LHS electrodes) and prefrontal area (F2 and G2 electrodes), were stereotypical findings. These observations are compatible not only with the above-mentioned neural connections but also with previous PET findings suggesting that the precuneus belongs to a mesial prefrontal-midparietal neural network (Cavanna *et al.* 2006; Malouin *et al.* 2003).

Conclusion

A rare case of precuneate cortical dysplasia manifesting as bilateral symmetric tonic posturing is reported. The symptomatology, invasive EEG findings and surgical outcome suggest that the epileptogenic zone was located mainly in the precuneate lesion and that epileptic activity propagated from the lesion to the SMA. □

Legend for video sequence

The patient was playing with a portable game. The seizure began abruptly, without warning, and was followed by bilateral symmetrical tonic posturing of the upper extremities and impaired consciousness. About 10 seconds later, he appeared to be smiling, which was followed by stiffening for several seconds. Finally, he answered "OWATTA" meaning "the seizure has ended" and he was alert.

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