## Video case report

Epileptic Disord 2006; 8 (3): 232-7

# Pilomotor seizures associated with sequential changes in magnetic resonance imaging

Pascal Masnou<sup>1,2</sup>, Jean-Paul Gagnepain<sup>2</sup>, Amal Fouad<sup>1</sup>, Denis Ducreux<sup>3</sup>, David Adams<sup>1</sup>

<sup>1</sup> Department of Neurology,

<sup>2</sup> Department of Clinical Neurophysiology and Epileptology,

<sup>3</sup> Department of Neuroradiology, University Hospital Kremlin-Bicêtre, Paris, France

Received September 19, 2005; Accepted June 12, 2006

**ABSTRACT** – Piloerection is rarely described in seizures. This symptom has been most frequently observed in patients with temporal lobe epilepsy and is rarely the principal clinical feature of seizures. No specific etiology of epilepsy associated with pilomotor seizures has been reported. We present the first case of a patient who experienced sudden and transitory epilepsy with pilomotor seizures occurring several times a day for months, and associated with sequential changes of the left hippocampus demonstrated by magnetic resonance imaging.

[Published with video sequences]

**Key words:** pilomotor seizures, transient MRI signal abnormality, hippocampus atrophy, autonomic seizures, TLE

Pilomotor seizures are classified as a subtype of autonomic seizures, socalled cutaneous autonomic seizures (Baumgartner *et al.* 2001). Piloerection is rarely reported as an ictal manifestation. It may occur unilaterally or bilaterally. Lateralizing and localising value of ictal piloerection is still the subject of debate (Loddenkemper *et al.* 2004). However, this symptom has been most frequently observed in patients with temporal lobe epilepsy. No specific etiology of epilepsy associated with pilomotor seizures has been found.

We describe an additional case with pilomotor seizures as the main feature of temporal lobe epilepsy, studied with video-EEG recording, and associated with sequential changes in magnetic resonance imaging (MRI).

## **Case report**

A 35-year-old, right-handed woman, with no relevant clinical history, was referred for sudden epilepsy with a seizure frequency reaching 30 per day. They lasted less than 90 seconds without loss of consciousness. They consisted of bilateral sensations of chill, associated with coloured phosphenes in the right hemi-field, nausea, thoracic compression, followed a few seconds later by piloerection involving both arms and legs. Results of neurological and general examination were normal.



#### Correspondence: P. Masnou Service de Neurologie, CHU de Bicêtre, 78 rue du Général Leclerc, 94275 Kremlin-Bicêtre Cedex <pascal.masnou@bct.aphp.fr>

Routine laboratory studies, including white cell count, showed normal results.

Standard EEG recording showed slow waves on the left temporal region. MRI performed one week after the first seizure, showed increased signal intensity of the left hippocampus in T2 and FLAIR-weighted images (*figure 1*).

She received antiepileptic drugs, which decreased the severity and the frequency of seizures. The number of seizures decreased from 30 to 10 per day.

Three and six months after the onset of epilepsy, follow-up MRI showed a slight regression of the signal abnormality in the left hippocampus (*figure 2A*, B).

Video-EEG performed six months after the start of the disease recorded six seizures, each lasting less than one minute. These seizures consisted of sensations of chill ascending from the feet to the whole of the body, associated with piloerection on the left arm. Ictal EEG showed diffuse flattening of the electrical activity followed by a rhythmic slow activity with a maximum amplitude on the left central and temporal area (*figure 3A*, B and *figure 4A*, B). Interictal video EEG was normal.

One month later, seizures ceased after adjustment of the antiepileptic medication. The patient became pregnant and had a healthy child.

Two years after the onset of epilepsy, MRI showed a clear decrease of the signal abnormality in the left hippocampus, however atrophy of the left hippocampus was observed (*figure 5*). The patient remains seizure-free with carbamazepine, 600 mg/d.

## Discussion

Pilomotor excitation as an ictal sign has been rarely well studied with video-EEG recordings. Several case reports and experimental findings confirm piloerection may be induced by epileptic discharges (Stefan *et al.* 2002). It could also be a secondary induced sensation occurring during seizures, in particular those associated with psychic symptoms such as feelings of fear. However, pilomotor excitation may be the first clinical symptom of seizure, as reported in five of out 25 patients with pilomotor seizures recorded at the Cleveland Clinic Foundation between 1994 and 2001 (Loddenkemper *et al.* 2004).

Ictal piloerection is usually associated with other symptoms. Most of them are autonomic signs: flushing, pallor, sweating, feeling of warmth or cold, shivering. In many cases, these autonomic signs are associated with other, non-autonomic ictal phenomena related to the onset and propagation of the ictal discharge: sensory hallucinations, feeling of fear, automatisms, loss of consciousness (Baumgartner *et al.* 2001, Loddenkemper *et al.* 2004).

Seizure consisting of piloerection as the principle ictal manifestation is very uncommon. Less than 10 cases have been reported in the literature (Roze *et al.* 2000, Lodden-kemper *et al.* 2004).

It most often occurs in patients with temporal lobe epilepsy (Stefan *et al.* 2003). However, the generator of ictal piloerection remains unclear. It has also been observed in seizures with frontal or parietal onset. The amygdala, anterior insula, anterior cingulate cortex and posterior orbitofrontal cortex are interconnected with the central



Figure 1. MRI performed seven days after the start of epilepsy: increase signal intensity of the left hippocampus in axial FLAIR-weighted images (hippocampic plan).



autonomic network: (Devinski *et al.* 2004). The autonomic network includes the hypothalamus, periaqueductal gray matter, parabrachial region in the pons, solitary tract nucleus and ventrolateral medulla with specific organization (Benarroch 1993). Electrostimulation or seizures spreading in the central autonomic network can modify autonomic functions. These autonomic changes can induce cardiovascular, respiratory, gastrointestinal, cutaneous, pupillary, urinary and genital, manifestations. Piloerection has been elicited by stimulation of multiple sites: insula, hippocampus, amygdala, hypothalamus, midbrain and medial prefrontal cortex in humans (Fish *et al.* 1993). Seizures originating in the mesial temporal area may spread to the insula inducing autonomic signs. In our observation, electroclinical and neuroimaging data analysis might suggest involvement of the left mesiotemporal and insula during the epileptic discharges. All these areas are interconnected with the central autonomic network. Piloerection may be localized, with the possibility of secondary spreading to another, homolateral or contralateral area of the body. It may also be generalized from the start of the seizures. Unilateral or initially unilateral piloerection is usually associated with an ipsilateral, epileptogenic focus (Loddenkemper *et al.* 2004).

A <sub>*</sub> ⊘	MTGI Ref	√ I	5µV/mm <b>∳</b> 0.1 s	<b>‡</b> 15 Hz		-0.0110/10/0
ED2 E4	8 9 10	11	12 13 14	15		m man a well to man har man
F4-C4	Mandanananananananana	- anter and an	mmuna		man man	and and an and and
C4-P4	mmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmm	Jann-manne	mandunder	and land an and an and an and an and an and an	And a	man was a second was and a second was
P4-O2	mannen	manum	Lunna and and	mandelahilanam	When	Magner and the second and the second
FP2-F8	mannan				and and a second like me	man man and with the man where we are the second
F8-T4	Munnahuman	hunnam	mmun	ana and a start and a start	man war war war war and	manun Manun war
T4-T6	Mannumment	month	mmunan	many	multiple and another Miller	warm allow man and and all and a weight war
T6-O2	mummmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmmm	manna	humana	. m MMMMMMMM	and a man and a man and a for the second states of	Maren and a start of the Market and the second and
Fz-Cz	month make programme	man	mannakan	WWW MARCH MAN MAN MARCH	~	
Cz-Pz	mannaman	mann	manna	~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~	when when when	menon have have more hard
FP1-F3	mannen	mon	Many Marian	may many and	man white and the second	home and the second of a second
F3-C3	-	mmm	moundsharmon	pypersurveyees and the second	an marker of the server	mana man and the share and a support
C3-P3	www.www.www.	Manna	monter	paardhamperayinneasidhade	and the second states and the second	warman who was have a server and
P3-O1	manner	manne	manning	mmmhhhhhhh	m when a star	and more and a second and as
FP1-F7	hammon	mon	month Manual Manual	~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~	mun and part	more thank the second of the second second second
F7-T3	mannon	mounter	~ mandal mar	and a second second second	Marine and a state of the second	an a
T3-T5	wwwwwwwwww	how	mannan	water and	many and many and many and	www.commission.commission
T5-O1	Mannenana	napproxim	moundation	manner and a sub-		and a superior and a
ECG	- manufr	- apr-ap	man	mon	manapal	monominaning
15:57:07	20 s	1000000	12 13 14	15 16 1.	7 18 19	20 21 22 23 24 25 26 27
	uus					
<sup>₿</sup> * Ø	MTGI Ref	V I	5µ∀/mm 🔹 0.1 s	15 Hz	🔹 A.ā 🎎 🎎	- Q O H D 2 E - 4
	28 29 30	31	32 33 34	35 36 3	7 38 39	40 41 42 43 44 45 46 47
FP2-F4	how we have a second	www		har were and		
F4-C4	har and a service	when a	and a second	Yman	and a second sec	an a
C4-P4	nete and the	a day de	and the second second	A meters	and the second sec	all for a second and a second and a second a fear and a fear a second a fear a second a fear a second a
FP2 F2	and a survey and a survey of the survey of t			al do a	detround and a second	WWWWwwwwwwwwwwwwwwwwwwwwwwwwwwwwwwwwww
F8 T4		www.ww	in the second se	in marine		and the second
T4-T6	and the second states	M. Mallinger	and a support of the second	A A A A A A A A A A A A A A A A A A A	and a second s	and management of the second
T6-O2	and the second s	aller , en	and a stand and a stand	and a second second	and the second	allenander her ander eine eine eine eine eine eine eine ei
Fz-Cz	hadrate and a start	- Arter Ma		d h	and a second sec	internet and the balance of the bala
Cz-Pz	man and and and and and and and and and a	when an	man and a survey of the	A reaction of the second	A samething a strange of	
FP1-F3	and advant	ty Am An	man and was shown of	When when the warden	- Arean	
F3-C3	mannennen	momente	man and a second second	Wet in the second with the second	and the fame and the second	An and a second second and a second
C3-P3	have marked	and Mary Anger	menter manually	monuture	and an and an and an and an and an and and	Marsham and the second and the secon
P3-O1	mannahur	many	mention	the man have not	mound	WWW.
FP1-F7	month	Marin	my many time,	1 Mary Man	warman	a manual and and a proposition of the second and
F7-T3	manunanter	harmon	warden warden the warden	mummer when	and the state of t	and promine the second and a second
T3-T5	mannaman	man how how	menneneppingunt	Margane hours	well a rettaining and the second	More many many many many many many many
T5-O1	entre manufactures	and marine parts	and a superior	mummer my my	have man por man parter	John marken and a start a start a start a start a start a start
ECG	mont	mpili	Junpurgung	mm	amanan	and a manufacture of the second
15:57:27	28 29 30 20 s	31	32 33 34	35 36 3	7 38 39	40 41 42 43 44 45 46 47
,					Coherence	

Figure 3. A, B). Ictal video-EEG recording showed diffuse flattening of the electrical activity followed by a rhythmic slow activity with maximum amplitude on the left central and temporal area.



Figure 4. A) Skin of left arm of the patient before seizure. B) Skin of left arm of the patient during the seizure; piloerection observed.

Previous case series found that left hemispheric epilepsy is most frequent than right hemispheric epilepsy in patients with ictal piloerection or cold shiver (Stefan *et al.* 2002). This finding is still a subject of debate. Piloerection has



**Figure 5.** MRI performed two years after the onset of epilepsy. The patient is seizure-free. Clear decrease in the hyperintensity signal of left hippocampus in axial and coronal T2 and FLAIR-weighted images (perpendicular to hippocampic plan). The left hippocampus is atrophic.

also been observed in patients with right temporal epilepsy (Devinski *et al.* 2004).

No specific etiology has been found. Of the previous reported cases, etiology included tumor, post-traumatic contusion, hippocampal sclerosis or atrophy, tuberous sclerosis, cavernous angioma, temporal malformation, radionecrosis (Roze *et al.* 2000, Loddenkemper *et al.* 2004).

From our observation, the etiology of the epilepsy is unclear. Magnetic resonance imaging performed seven days after the onset of the disease showed increase signal intensity in the left hippocampus, in both T2 and FLAIRweighted images. Follow-up MRI showed regression of this signal abnormality. Two years after the first seizure, while the patient became seizure-free, atrophy and a slightly increased signal intensity of the left hippocampus in T2 and FLAIR-weighted images were found. These abnormalities are not suggestive of an ischemic lesion. Also, a neoplastic lesion appears to be unlikely because of the spontaneous regression of the signal abnormalities with subsequent atrophy of the left hippocampus. By contrast, this finding might suggest neuronal loss (Lansberg et al. 1999, Meierkord et al. 1997) or changes in the hippocampus associated with frequent and daily seizures (Van Paesschen et al. 1998, Bernasconi et al. 2005). However we can't exclude infection or an inflammatory process because unfortunately CSF analysis was not performed at the onset of the disease (Suzuki et al. 1999).

Voltage-gated potassium channels antibodies were not assayed. These have been recently reported in cases of limbic encephalitis and other seizure-associated disorders. Nevertheless, our patient did not present with any clinical features of paraneoplastic or non-paraneoplastic limbic encephalitis (McKnight *et al.* 2005, Wieser *et al.* 2005, Vincent *et al.* 2004).

### References

Baumgartner C, Lurger S, Leutmezer F. Autonomic symptoms during epileptic seizures. *Epileptic Disord* 2001; 3: 103-16.

Benarroch EE. The central autonomic network: functional organisation, dysfunction, and perspective. *Mayo Clin Proc* 1993; 68: 988-1001.

Bernasconi N, Natsume J, Bernasconi A. Progression in temporal lobe epilepsy. Differential atrophy in mesial temporal structures. *Neurology* 2005; 65: 223-8.

Devinski O. D'esposito M. *Neurology of cognitive and behavior disorders*. New York: Owford University Press, 2004: (336–51).

Fish DR, Gloor P, Quesney FL, *et al.* Stimulation of the temporal and frontal lobes in patients with epilepsy. *Brain* 1993; 116: 397-414.

Lansberg MG, O'Brien MW, Norbash AM, *et al.* MRI abnormalities associated with partial status epilepticus. *Neurology* 1999; 52: 1021-7.

Loddenkemper T, Kellinghaus C, Gandjour J, *et al.* Localising and lateralising value of ictal piloerection. *J Neurol Neurosurg Psychiatry* 2004; 75: 879-83.

McKnight K, Jiang Y, Hart Y, et al. Serum antibodies in epilepsy and seizure-associated disorders. *Neurology* 2005; 65: 1730-6.

Meierkord H, Wieshmann U, Niehaus L, *et al.* Structural consequences of status epilepticus demonstrated with serial resonance imaging. *Acta Neurol Scand* 1997; 96: 127-32. Roze E, Oubary P, Chedru F. Status-like recurrent pilomotor seizures: case report and review of the literature. *J Neurol Neurosurg Psychiatry* 2000; 68: 647-9.

Stefan H, Pauli E, Kerling F, *et al.* Autonomic auras: left hemispheric predominance of epileptic generators of cold shivers and goose bumps?. *Epilepsia* 2002; 43: 41-5.

Stefan H, Feichtinger M, Black A. Autonomic phenomena of temperature regulation in temporal lobe epilepsy. *Epilepsy Behav* 2003; 4: 65-9.

Suzuki K, Jimi T, Wakayama Y, *et al.* A case of non-herpetic acute encephalitis presenting high intensity lesion at unilateral temporal cortex on MR FLAIR image. *Rinsho Shinkeigaku* 1999; 39: 750-6.

Van Paesschen W, Duncan JS, Stevens JM, *et al.* Longitudinal quantitative hippocampal magnetic resonance imaging study of adults with newly diagnosed partial seizures: one-year follow-up results. *Epilepsia* 1998; 39: 633-9.

Vincent A, Buckley C, Schott JM, *et al.* Potassium channel antibody-associated encephalopathy: a potentially immuno-therapy-responsive form of limbic encephalitis. *Brain* 2004; 127: 701-12.

Wieser S, Kelemen A, Barsi P, *et al.* Pilomotor seizures and status in non-paraneoplastic limbic encephalitis. *Epileptic Disord* 2005; 7: 205-11.